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Review Article	5000	250	50	6	10 or total of 20 images
Case Report	1000	200	15	No tables	10 or total of 20 images
Letter to the Editor	500	No abstract	5	No tables	No media

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Tables should be included in the main document, presented after the reference list, and they should be numbered consecutively in the order they are referred to within the main text. A descriptive title must be placed above the tables. Abbreviations used in the tables should be defined below the tables by footnotes (even if they are defined within the main text). Tables should be created using the "insert table" command of the word processing software and they should be arranged clearly to provide easy reading. Data presented in the tables should not be a repetition of the data presented within the main text but should be supporting the main text.

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Book Section: Suh KN, Keystone JS. Malaria and babesiosis. Gorbach SL, Barlett JG, Blacklow NR, editors. Infectious Diseases. Philadelphia: Lippincott Williams; 2004.p.2290-308.

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Books with a Single Author: Sweetman SC. Martindale the Complete Drug Reference. 34th ed. London: Pharmaceutical Press; 2005.

Editor(s) as Author: Huizing EH, de Groot JAM, editors. Functional reconstructive nasal surgery. Stuttgart-New York: Thieme; 2003.

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Thesis: Yılmaz B. Ankara Üniversitesindeki Öğrencilerin Beslenme Durumları, Fiziksel Aktiviteleri ve Beden Kitle İndeksleri Kan Lipidleri Arasındaki Ilişkiler. H.Ü. Sağlık Bilimleri Enstitüsü, Doktora Tezi. 2007.

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Manuscripts Published in Electronic Format: Morse SS. Factors in the emergence of infectious diseases. Emerg Infect Dis (serial online) 1995 Jan-Mar (cited 1996 June 5): 1(1): (24 screens). Available from: URL: http:/ www.cdc.gov/ncidodlElD/cid.htm.

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Case Reports

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Treatment of Antegrade Left Main Coronary Artery Dissection Due to A Iatrogenic Catheter Using the Reverse Crush Bifurcation Technique

İatrojenik Katetere Bağlı Antegrad Sol Ana Koroner Arter Diseksiyonunun Reverse Crush Bifurkasyon Tekniği ile Tedavi Edilmesi

Ahmet Öz, Süleyman Çağan Efe, Mehmet Sait Altıntaş, Turgut Karabağ, Burak Ayça
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ABSTRACT

Iatrogenic left main coronary artery (LMCA) dissection is a rare but potentially life-threatening complication of invasive coronary procedures. The technique for managing LMCA dissection varies and depends on the patient's comorbidities and degree of hemodynamic stability. Here, we report the case of a 51-year-old patient with iatrogenic LMCA dissection who was successfully treated using the reverse crush bifurcation technique.

Keywords: Iatrogenic dissection, left main coronary artery, reverse crush bifurcation technique

ÖZ

İatrojenik sol ana koroner arter (LMCA) diseksiyonu, invaziv koroner prosedürlerin nadir ancak potansiyel olarak yaşamı tehdit edici bir komplikasyonudur. LMCA diseksiyonunu yönetme stratejisi değişkendir ve hastanın komorbiditelerine ve hemodinamik stabilite derecesine bağlıdır. Burada; 51 yaşında erkek iatrojenik LMCA diseksiyonu olan hastanın reverse crush bifurkasyon tekniği ile başarılı bir şekilde tedavi edildiği bir olgu sunulmuştur.

Anahtar Kelimeler: İatrojenik diseksiyon, sol ana koroner arter, reverse crush bifurkasyon tekniği

Introduction

Coronary angiography is widely used worldwide (1). The overall complication rate in diagnostic coronary angiography is very low. Despite the experience of advanced catheter technologies and physicians, stroke and dissection/occlusion of peripheral or coronary arteries still remain a major problem in cardiac catheterization (2). The left main coronary artery (LMCA) is a significant and rare complication. LMCA dissection may occur due to iatrogenic, spontaneous, or ascending aortic dissection complication (3). Iatrogenic LMCA dissection can be seen as antegrade or retrograde dissection (4). Here; a case of iatrogenically catheter-induced antegrade LMCA dissection and successfully treated using reverse crush bifurcation technique was presented.

Case Report

A 51-year-old male patient was evaluated in an outpatient clinic with a typical complaint of exertion dyspnea and angina that occurred with minimal exertion over the last 20 days. On physical examination, the heart sounds were rhythmic and no additional sound/murmur was heard. Other system examinations were normal. His resume included hyperlipidemia and smoking. He had no history of constant drug use. His pedigree had no obvious features. His electrocardiogram was in sinus rhythm. No change in segment ST-T was observed, and systolic function was normal in echocardiography (EF 60%). The patient was admitted to the coronary intensive care unit with the diagnosis of unstabil angina pectoris. Blood tests showed fasting blood glucose: 101 mg/dL, creatinine: 0.7 mg/dL, low-density lipoprotein: 220 mg/dL and HbA1c: 5.5 mmoL/moL. Selective coronary angiography was planned for the patient and written informed consent was obtained. In coronary angiography; There was plaque in the distal of the LMCA, subtotal stenosis in the osteal of the left anterior descending (LAD) artery and distal TIMI-3 flow. The circumflex (CX) artery was detected with plaques in the first poses, and the dissection line extending from the LMCA to the distal of the CX artery was observed in the caudal poses. The right coronary artery was found

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with plaques (Figure 1). After the patient had severe angina during the coronary imaging process and monitoring ST depressions on the monitor, percutaneous coronary intervention (PCI) was planned for the patient's lesions. The left system was cannulated with a 7-F EBU 3.75 guiding catheter. 8,000 units of heparin and intravenous tirofiban were administered. Soft wires were directed to the LAD artery and CX artery distal. LAD artery osteal lesion was predilated using a 2.0x15 mm balloon. Then, a 3.0x18 mm drug-eluting stent (DES) was implanted from the LMCA at 16 atm to the LAD artery osteal region, Proximal optimization technique was performed with a 4.0x9 mm non-compliant balloon. Soft wire was advanced distally by performing CX artery rewiring. Kissing dilatation was applied using LAD artery 3.0x15 mm and CX artery 2.0x20 mm balloons. When 3.0x33 mm DES could not be advanced to the CX artery lesion, kissing dilatation was applied again using LAD artery 3.0x15 mm and CX artery 3.0x12 mm balloons. Subsequently, the proximal 3.0x33 mm DES was implanted in the CX artery at 16 atm, with the LMCA stent enclosed. Since dissection was limited in the distal of the stent and it did not disturb the distal flow, he was followed up medically. The stent was crushed with a 3.0x15 mm balloon, the proximal part of which was parked in the LAD artery. CX artery rewiring was done again and kissing dilatation was applied with 3.0x15 mm balloons. Due to dissection line monitoring in LMCA proximal, 4.5x9 mm bare metal stent was implanted at 16 atm. The patient's angina complaints regressed, ST depression improved, and the procedure was terminated. After 4 days of follow-up in the coronary intensive care unit, the patient was discharged with acetyl salicylic acid 100 mg 1x1, clopidogrel 75 mg 1x1, metoprolol 50 mg 1x1, ramipril 2.5 mg 1x1, pantoprozole 40 mg 1x1 and atorvastatin 40 mg 1x1 and control coronary angiography was planned. In the control coronary angiography performed one month later; The LMCA stent was open, minimal dissection area at the proximal stent, LAD artery stent open, CX artery stent open, dissection area distal to the stent that limited itself and did not disrupt the flow was detected (Figure 2). Since the patient did not have angina and exertional dyspnea, medical follow-up was decided.

Discussion

Iatrogenic LMCA dissection is a rare complication during coronary angiography or angioplasty. Incidence has been reported at approximately 0.07% (5). Iatrogenic LMCA dissection risk factors may depend on the patient and the procedure. Patient-related risk factors can be listed as; coronary artery abnormalities, connective tissue disorders (Marfan's syndrome), atherosclerotic changes (LMCA stenosis), arterial hypertension, bicuspid aortic valve, aortic root calcification and age (6). Procedure-related risk factors include catheter manipulation, use of >6F catheters, stiff catheter tips, strong contrast agent injection, balloon dilation, and stenting (7,8). Medical follow-up (if there is no hemodynamic disorder), PCI and coronary bypass operation are available as treatment. PCI is preferred in hemodynamically unstable patients in terms of time and technique (9). In an observational study involving 38 patients with iatrogenic LMCA dissection, 6 patients were treated conservatively, 14 patients were treated percutaneously, and 17 patients surgically. No difference was found between percutaneous intervention and surgery during the 5-year follow-up (5). If the hemodynamics of patients with iatrogenic LMCA dissection are not stable, it is very important for a positive result to be controlled quickly with percutaneous intervention. Our case was also successfully treated with reverse crush bifurcation technique due to unexpected catheter-induced LMCA dissection disrupting the patient's hemodynamics.

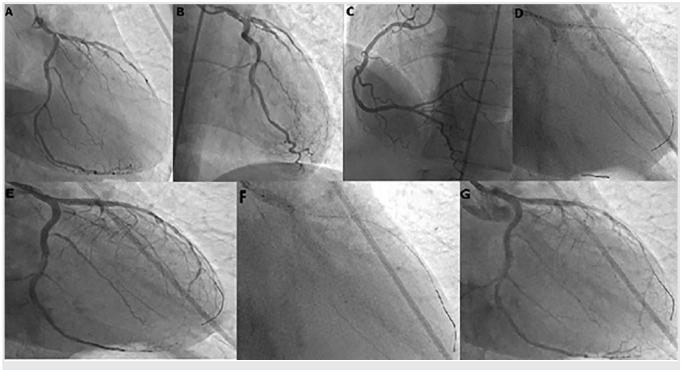

Figure 1. A: Subtotal stenosis in the left anterior descending artery osteal, **B:** Dissection extending from left main coronary artery to distal to circumflex artery, **C:** Right coronary artery plated, **D:** Kissing ballooning, **E:** dissection at proximal left main coronary artery, **F:** Left anterior descending artery proximal stent implantation, **G:** Successful reverse crush bifurcation stenting

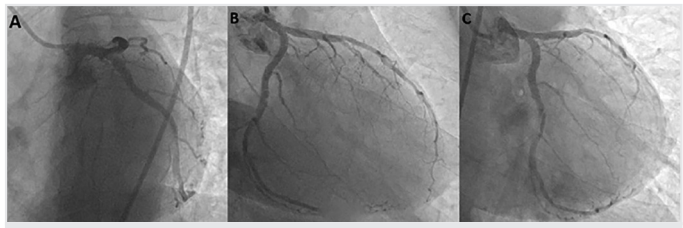

Figure 2. A, B: Open left main coronary artery stent, minimal dissection area proximal to stent, **C:** Circumflex artery stent is open; self-limiting dissection line distal to the stent

As a result, the best treatment strategy of choice in patients with iatrogenic LMCA dissection is immediate recognition of this complication, determination of the hemodynamic state of the patient and the need for surgery. Stenting strategy by percutaneous intervention is acceptable in hemodynamically unstable patients and should be carried out as soon as possible.

Ethics

Informed Consent: The patient and written informed consent was obtained.

Peer-review: Internally peer-reviewed.

Authorship Contributions: Surgical and Medical Practices - A.Ö., B.A.; Concept - A.Ö.; Design - A.Ö., B.A., T.K.; Data Collection or Processing - S.Ç.E.; Literature Search - M.S.A., S.Ç.E.; Writing - A.Ö., T.K.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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Intramuscular Myxoma of the Right Cruris: A Case Report

Sağ Bacakta İntramusküler Miksoma: Olgu Sunumu

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ABSTRACT

Intramuscular myxoma is one of the rare soft tissue benign tumours. The most common localisation of the myxoma is the heart muscle. It has been reported less frequently in the hip and around the shoulder, trunk, neck and cruris. We aimed to report this rare case of the cruris.

Keywords: Benign tumour, cruris, intramuscular myxoma

ÖZ

İntramusküler miksoma yumuşak dokunun iyi huylu nadir tümörlerinden biridir. Miksomanın tipik yerleşim yeri kalp kasıdır. Kalça, omuz çevresi, gövde, boyun ve bacakta daha az rastlanmaktadır. Bacakta görülen bu nadir olguyu bildirmeyi amaçladık.

Anahtar Kelimeler: İyi huylu tümör, bacak, intramusküler miksoma

Introduction

Intramuscular myxoma is a rare benign soft tissue tumour of mesenchymal origin surrounded by muscular tissue (1,2). Myxomas are slow-growing tumours. They are most commonly localised in the heart muscle, while other rare cases were detected within the muscle tissue of the extremities. It has been reported less frequently in the hip and around the shoulder, trunk, neck and cruris (3). It is observed more frequently among women in the fourth to sixth decades (4,5). Therefore, we aimed to report this rare case.

Case Report

Informed consent was obtained from the patient for the case presentation below. A 56-year-old female patient was evaluated for pain and swelling in the right cruris. The patient had been suffering from this complaint of swelling for six months, with the addition of pain for the last month. On physical examination, the lesion was found to be firm, mobile and painful. The patient was evaluated by ultrasonography and magnetic resonance imaging. The mass was found as well-circumscribed with homogenous signal intensity, located on the proximal cruris anterior-medial side in the subcutaneous fat tissue and having a size of 24x64 mm The lesion was demonstrated by T1 weighted sequences low signal intensity and T2 fat suppressed sequences high signal intensity in the magnetic resonance imaging. The mass, which was distinct from the surrounding tissues, was excised in the form of an en-block with a narrow margin using a longitudinal incision (Figure 1). Histopathology demonstrated a tumour consisting of spindle- and stellate-shaped cells embedded within loose fibrous and myxoid stroma. It was found to be moderately well-circumscribed. Atypical cytologic features and mitotic figures were not detected. The lesion contained fatty tissue areas separated by fibrous bands in the fibroadipous tissues. This benign lesion was defined as a myxoma. The pain and cosmetic complaints

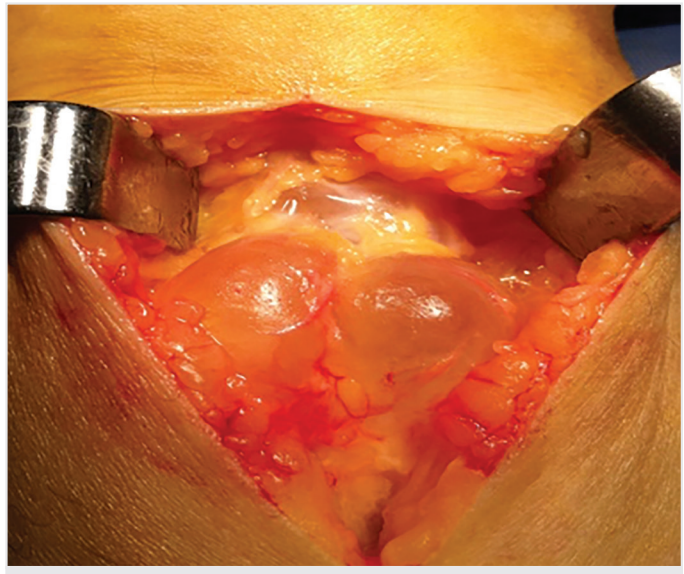

Figure 1. Intramuscular myxoma view during surgery

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were resolved after the surgery. It is thought that the pain complaint may have resulted from the local pressure effect of the mass. Physical examination did not observe any recurrences of the mass during the 2-year postoperative follow-up.

Discussion

Benign and malignant soft tissue tumour, haematoma and abscess should be considered in the differential diagnosis. The intramuscular myxoma has not been observed in the presence of cellular atypia, increased mitotic activity and rapid growth (6). The separation of the borders from the surrounding muscle tissue in the radiological evaluation helps in the differential diagnosis (7). The relationship between intramuscular myxoma and fibrous dysplasia is known. Mazabraud's syndrome is a rare syndrome in which benign intramuscular myxoma occurs in association with a monostotic or polyostotic form of fibrous dysplasia on the bone (8,9). It may accompany the McCune Albright syndrome (10). Complete excision with narrow margins is advised. The researcher revealed the occurrence of local recurrence in association with the incompletely excised tumours. Metastases and a locally aggressive behaviour have not been observed. Neoadjuvant and adjuvant treatment were not recommended (11).

Intramuscular myxoma should be considered in the differential diagnosis of soft tissue lesions of the lower extremities.

Ethics

Informed Consent: Informed consent was obtained from the patient for the case presentation below.

Peer-review: Externally peer-reviewed.

Authorship Contributions: Surgical and Medical Practices - E.C.; Concept - A.B., E.C.; Design - A.B.; Data Collection or Processing - A.B., E.C.; Analysis or Interpretation - E.C.; Literature Search - A.B., E.C.; Writing - A.B.

Conflict of Interest: No conflict of interest was declared by the authors.

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Wellens Syndrome Presenting with Sudden Cardiac Arrest

Wellens Sendromuna Bağlı Kardiyak Arrest: Olgu Sunumu

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ABSTRACT

Wellens syndrome is characterised by changes in electrocardiogram precordial lead T-waves accompanied by proximal stenosis of the left anterior descending artery. Diagnostic criteria include T-wave changes plus a history of anginal chest pain without serum marker abnormalities, with significant ST-segment elevation and lack of Q waves. Wellens syndrome results in anterior wall acute myocardial infarction and cardiac death. We describe the case of a 33-year-old man who presented to the emergency clinic with cardiac arrest.

Keywords: Wellens syndrome, T-wave changes, cardiac arrest

ÖZ

Wellens sendromu prekordiyal T-dalga değişikliği ve sol ön inen arterin kritik darlığı ile karekterizedir. Tanı kriterleri T-dalgası değişiklikleri ve serum enzim yükselmesi olmaksızın anjinal göğüs ağrısı öyküsü, Q dalgası yokluğu, belirgin ST- segment elevasyonunu içerir. Wellens sendromu akut yaygın ön duvar enfarktüsü veya kardiyak ölüm ile sonuçlanabilmektedir. Burada acil kliniğe ani kardiyak arrestle getirilen 33 yaşında erkek hasta sunulacaktır.

Anahtar Kelimeler: Wellens sendromu, T-dalga değişikliği, kardiyak arrest

Introduction

Wellens syndrome was first described in 1982 and is a syndrome characterized by critical stenosis of the left anterior descending (LAD) artery and unique electrocardiogram (ECG) findings. It was first demonstrated by the detection of critical LAD artery stenosis in a group of patients with angina pectoris (1). There are two different ECG patterns. Type A pattern is less visible and more specific, and characterized by the presence of biphasic T-waves, which are mostly seen in precordial derivations, especially in V2-V3. Type B pattern is more common and is characterized by deep and symmetrical T-waves in the anterior derivation (1,2). In its etiology, there is a critical stenosis of LAD artery, and when undiagnosed, it may result in myocardial infarction or even death. Here, we present a young patient who was brought to the hospital with cardiac arrest and had Wellens syndrome pattern on ECG and 98% stenosis in the LAD artery on coronary angiography and underwent restenosis treatment.

Case Report

A 33-year-old male patient was brought to our emergency clinic with a picture of respiratory and cardiac arrest. In his history, it was learned that there was a feeling of pressure in the chest for the last few months whose frequency and duration increased gradually in the last week, and that there were no additional suggestions after the ECG taken at the health institution he applied a week ago. It was stated by his relatives that he suddenly collapsed in the morning, could not breathe and became bruised and was brought to the emergency room quickly. It was learned that he had a history of smoking 20 packs/year, occasional alcohol and infrequent substance use. Cardiovascular resuscitation was performed in the patient, whose respiration was not observed and did not have a heart beat at admission. After the intervention, the patient who had a peak heartbeat but had insufficient spontaneous respiration was taken to the intensive care unit. The patient's ECG at admission also showed high widespread ST in V1-V6 derivations. In the ECG performed 1 week ago, aV2-V4 biphasic T-wave was observed (Figure 1). Emergency cardiac catheterization was performed with the diagnosis of Wellens syndrome and widespread anterior wall myocardial infarction. In coronary angiography, critical stenosis (98%) was detected in the proximal LAD artery and it was revascularized (Figure 2, 3). He was extubated on the second day because he had spontaneous breathing. Antibiotherapy was started when pneumonic consolidation was observed in the chest radiography of the patient whose fever increased in the follow-up. On the seventh day, the patient, whose infective parameters regressed, had no additional problems, was discharged.

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Discussion

T-wave changes seen in the ECG in Wellens syndrome can often be overlooked in the symptom-free period or are generally considered as non-specific. When not recognized, myocardial infarction can even result in death. The patient who is diagnosed with with intermittent chest pain or ambiguous complaints, a T-wave change in the ECG performed at the moment when there is no complaint of pain is characterized. In patients with a type A pattern on the ECG, the findings may turn into a type B pattern over time. Chest pain can be specific and non-specific.

de Zwaan et al. (2) first identified Wellens syndrome in 180 out of 1,260 patients with angina in the presence of typical ECG changes and stenosis

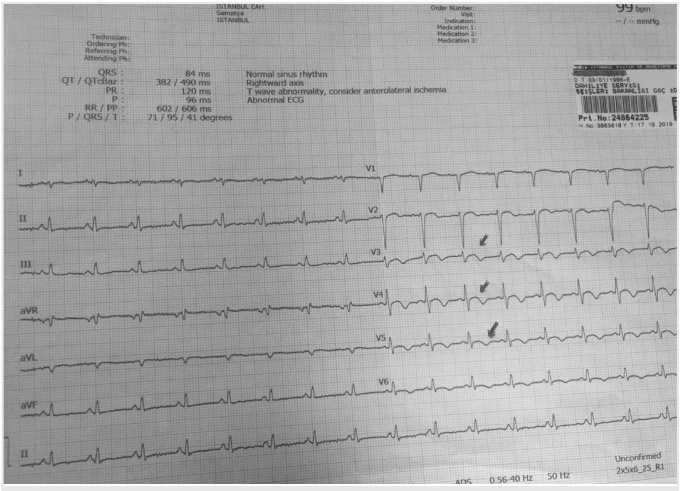

Figure 1. Negative T-waves in V3-5 derivations on electrocardiography at admission

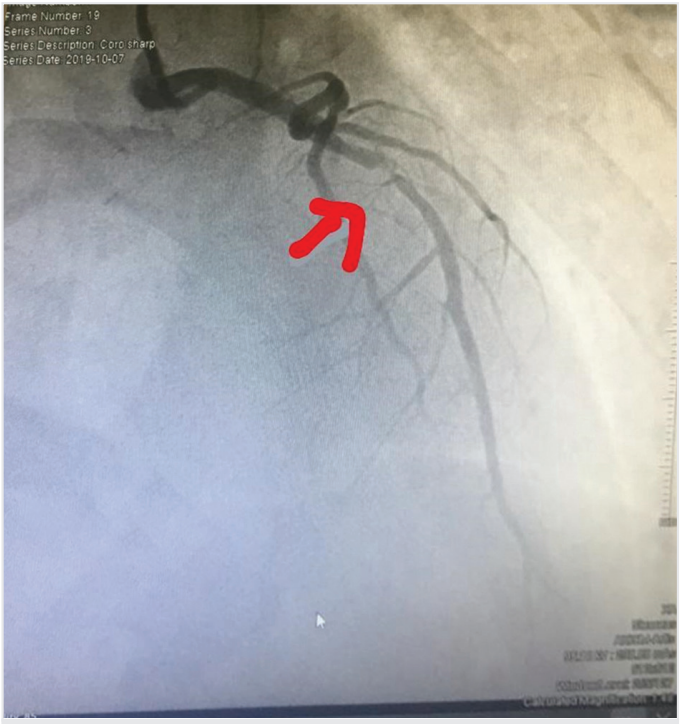

Figure 2. Coronary angiography showing 95% stenosis in the lumen of the proximal segment in the left anterior descending coronary artery

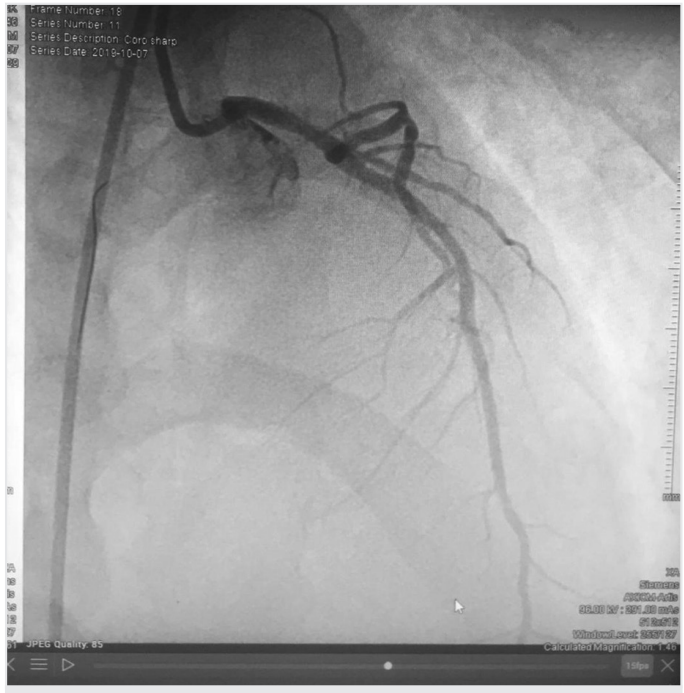

Figure 3. After successful revascularization, re-flow was enabled in left anterior descending

in the proximal LAD artery. While 108 patients had typical ECG findings at admission, it was detected later in 72 patients.

Diagnostic criteria of Wellens syndrome are intermittent chest pain, biphasic or symmetrical deep inverted T-waves primarily in V2 and V3, absence of pathological precordial Q waves, minimal increase or absence of cardiac enzymes (3). ECG findings in Wellens syndrome are typical before myocardial infarction develops, and recognition during this period is life-saving. When diagnosed, the first procedure to be performed is conventional angiography (4,5).

In some cases, ECG findings are seen without LAD stenosis, this situation is called pseudo-wellens. These are acute cholecystitis, congenital myocardial band, Takotsubo cardiomyopathy and substance abuse. In such cases, a good history and clinical examination should be evaluated. As a matter of fact, in the study conducted by Kobayashi et al. (6), the analysis of 424 patients with non-ST elevation myocardial infarction revealed a Wellens ECG pattern in 4.2% of the patients, but LAD stenosis in only 50%.

Although the cause of dynamic ECG changes is not fully known, LAD mimics vasospasm in the artery. It is thought that malnutrition and subsequent reperfusion may cause ECG changes.

This case resulted in sudden cardiovascular arrest due to the fact that the patient was not diagnosed despite the presence of non-specific chest pain prior to it. This reveals the importance of early diagnosis in Wellens syndrome. The incidence of myocardial infarction and sudden death has significantly increased in these patients, and revascularization is recommended when the diagnosis is made (3).

Ethics

Informed Consent: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions: Surgical and Medical Practices - S.Y., Y.B., H.U.A., M.E.P.; Concept - H.U.A., M.E.P.; Design - H.U.A.; Data Collection or Processing - S.Y.; Analysis or Interpretation - Y.B.; Literature Search - S.Y., Y.B., M.E.P.; Writing - S.Y., Y.B., H.U.A., M.E.P.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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Rheumatic Diseases Presenting with Guillain-Barré Syndrome: Sjögren's Syndrome and Systemic Lupus Erythematosus

Guillain-Barré Sendromu ile Prezente Olan Romatolojik Hastalıklar: Sjögren Sendromu ve Sistemik Lupus Eritematozus

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ABSTRACT

The coexistence of Guillain-Barré syndrome (GBS) with autoimmune rheumatic diseases is extremely rare. In addition to flu, infection, trauma, vaccination, blood transfusion, chemotherapy and surgical intervention, rheumatic diseases may be the underlying etiological cause of resistant and prolonged GBS cases. Therefore, appropriate treatments specific to rheumatic diseases may accelerate the healing process, reduce the possibility of neurological sequelae and greatly contribute to the rehabilitation process.

Keywords: Guillain-Barré syndrome, rehabilitation, rheumatic diseases, systemic lupus erythematosus, Sjögren syndrome

ÖZ

Guillain-Barré sendromu (GBS) ile otoimmün romatolojik hastalıkların birlikteliği oldukça nadirdir. Gribal enfeksiyon, travma, aşılama, kan transfüzyonu, kemoterapi ve cerrahi girişim yanında dirençli ve uzun süren GBS olgularında romatolojik hastalıklar altta yatan etiyolojik neden olabilir. Romatolojik hastalığa özgü spesifik tedavi iyileşme sürecini hızlandırarak nörolojik sekel kalma olasılığını azaltır ve rehabilitasyon sürecine büyük oranda katkı sağlar.

Anahtar Kelimeler: Guillain-Barré sendromu, rehabilitasyon, romatolojik hastalıklar, sistemik lupus eritematozus, Sjögren sendromu

Introduction

Guillain-Barré syndrome (GBS) is an acute inflammatory polyneuropathy characterized by rapidly progressive symmetrical muscle weakness and loss of deep tendon reflexes (DTR). Symptoms usually start in the lower extremities and progress to the trunk and upper extremities within days. The progression is usually symmetrical. Sensory loss, autonomic and cranial neuropathy, and neuropathic pain often accompany. Neurological signs develop over a period of several days to a month. If it is longer than one month, chronic inflammatory demyelinating polyradiculoneuropathy is mentioned (1). It is generally a middle-age disease and is more common in men (2). Influenza infection, trauma, vaccination, blood transfusion, chemotherapy, and surgical intervention that occurred weeks before the onset of symptoms are included in the etiology (3). Sjögren's syndrome (SS) is a systemic autoimmune disease characterized by lymphocytic infiltration of all exocrine glands, primarily tear and salivary glands. The incidence in women is 9 times higher, similar to other autoimmune diseases, and it peaks in the 4th-5th decades (4). It is more common for patients with SS in advanced age to manifest with neurological complications (2). Systemic lupus erythematosus (SLE) is a chronic connective tissue disease with unknown etiology and autoimmune character, which cause immunological disorders and involving many organs and systems. Central nervous system involvement constitutes 93.1% of the neurological involvement seen in SLE. Its association with GBS is quite low, such as 0.6-1.7% (5). Here, a female patient who developed GBS after previous gastrointestinal surgery and was diagnosed with SS afterwards, and a young male patient diagnosed with SLE after the development of GBS will be discussed in the light of the literature.

Case Reports

Case 1

A 56-year-old female patient was hospitalized with the diagnosis of GBS for rehabilitation. In her history, she had a diagnosis of hiatal hernia, which was made in an external center where she applied with the

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complaint of shortness of breath 6 years ago. Nissen fundoplication operation was applied to the patient in May 2018 due to the ongoing and increasing complaints of shortness of breath and vomiting during this period. When the complaints of vomiting started 2-3 months after the operation, the patient was referred to the general surgery clinic of our hospital, and the mesh migration was seen in the gastroscopy performed in January 2019 and the operation decision was taken. Antibiotic treatment was arranged due to the development of subcutaneous infection during follow-up. On January 29, 2019, she was consulted with the neurology clinic due to numbness around the mouth and weakness in the arms and legs. No feature was detected in the brain magnetic resonance imaging of the patient. An electroneuromyography (ENMG) study showed acute sensorimotor polyneuropathy, in which motor axonal damage to the upper and lower sensory and motor fibers is at the forefront and the sural response is maintained. The patient was transferred to the neurology service with the pre-diagnosis of GBS. Protein was found to be high (134 mg/dL) in the cerebrospinal fluid (CSF) examination in lumbar puncture (LP). There was no reproduction in the CSF culture. In the neurology service, the patient was given intravenous immunoglobulin (IVIG) at a dose of 0.4 kg/day for 5 days. After one month, the patient was transferred to our service. In the patient's neurological examination, muscle strength was 3/5 in the right and left lower and upper extremities. DTR's were hypoactive. The patient's sitting balance, fine ability in the hands and grip strength were very poor. The patient was given joint range of motion (ROM), stretching, strengthening exercises, back and abdominal muscles strengthening, sitting balance training and neuromuscular electrical stimulation (NMES) (bilateral tibialis anterior, quadriceps, hamstring, wrist flexors and extensors, supraspinatus and deltoid) for four extremities in the bed. Afterwards, training was continued with standing with posterior shell and foot-ankle orthosis and walking with parallel bar.

Since the acute phase reactants were elevated during the patient's hospitalization in the physical therapy service, infection and malignancy were investigated in etiological aspect. Endoscopy, colonoscopy, abdominal and thorax computed tomography, breast ultrasonography and mammography were unremarkable. Her gynecological examination was normal. Protein electrophoresis, serum and urine immunofixation electrophoresis, tuberculosis culture was negative. Blood glucose levels were normal.

An extractable nuclear antigen profile was requested from the patient who had vague hand pain and dry mouth. The result of salivary gland biopsy taken with a pre-diagnosis of SS from the patient with anti-SSA positivity was negative.

A patient who was found to have dry eyes in the Schirmer test (right eye: 5 mm, left eye: 5 mm) was consulted with the rheumatology clinic, followed by pulse steroid (1000 mg/day, 3 days IV, then oral 40 mg prednisolone), rituximab was given as immunosuppressive and IVIG booster was given for 5 days. Sedimentation rate decreased from 103 mm/hour to 42 mm/hour. Steroid dose was reduced to 5 mg/day. Hydroxychloroquine was started at a dose of 200 mg/day as a disease modifier. Muscle strength increased to 4/5 level. At the end of the 4-week rehabilitation period, the patient reached the level of ambulation without the need for a device in the parallel bar. At 6th months after discharge, the patient could ambulate without support and was independent in daily living activities. Written informed consent form was obtained from the patient.

Case 2

A 39-year-old male patient applied to the emergency service of our hospital 3 months ago with complaints of weakness in the arms and legs and numbness in the hands after acute gastroenteritis lasting about a week. In the emergency examinations and cranial imaging, the patient, who had no neurological pathology explaining the acute picture, was transferred to the neurology service with the pre-diagnosis of GBS. In the CSF examination with LP, the protein level was found to be high and no cells were detected. In ENMG, acute demyelinating polyneuropathy was detected, where motor axonal damage and message block holding motor fibers at the top and bottom were at the forefront. The patient, who received IVIG treatment at a dose of 0.4 kg/day for 5 days with the diagnosis of GBS, was then transferred to our service for rehabilitation. In the patient's neurological examination, muscle strength was 1/5 in the right and left lower and upper extremities. The patient lacked balance of sitting, fine skill and grip in the hands. Muscle tone was hypotonic and DTRs could not be taken. In our service, training on in-bed positioning, ROM exercises, and sitting balance was started. Strengthening exercises were given to upper and lower extremities, abdominal and back muscles. NMES was applied to bilateral quadriceps, tibialis anterior, hamstring, finger flexor and extensor, wrist flexor and extensor, deltoid, supraspinatus muscles.

After the antinuclear antibody, anti-dsDNA and anti-Ro antibodies of the patient who was examined for vasculitis tests as part of the etiological examination resulted positive, he was consulted with the rheumatology clinic. The patient, who was diagnosed with SLE with SLE International Cooperation Classification Criteria, was continued with 60 mg/day prednisolone after pulse steroid for 7 days. Considering that SLE had neurological involvement, rituximab and hydroxychloroquine 200 mg 2x1/day was started. Antiviral therapy (entecavir) was started with the consultation of infectious diseases clinic to the patient with accompanying anti-HBcIgG positivity. In addition, plasmapheresis with 10 sessions of albumin was applied to the patient. With the response to the treatment, the steroid dose was reduced to 5 mg/day. Monthly IVIG booster application continued. After 3 months of rehabilitation, the patient reached the level of ambulation in the parallel bar. Written informed consent form was obtained from the patient.

Discussion

Neurological involvement is 20% in patients with SS. Interestingly; neurological signs may precede the symptoms of sicca. Various types of peripheral neuropathy have been identified in patients with SS [symmetrical sensorimotor polyneuropathy (most common), symmetrical sensory neuropathy, sensory neuronopathy, autonomic neuropathy, mononeuropathy, mononeuropathy multiplex, and cranial neuropathy (especially trigeminal neuropathy)]. The variety of clinical symptoms may cause delays in the diagnosis of SS (2). Association of SS and GBS is rare in the literature (6,7). In a study that tracked cases that developed GBS after surgery, the average age of patients was 55, and the

proportion of male patients was 56.5%. The average time between the surgical procedure and the development of GBS has been reported as 8 weeks. It has been observed that the risk of GBS is increased in patients with malignancy and comorbid autoimmune diseases. In the same study, association with autoimmune diseases was found in 10% of cases and a diagnosis of SS was found in 1.5% of cases. As a surgical procedure, it was most frequently (32.1%) associated with gastrointestinal surgery (3). The age of our patient was compatible with this case series. The diagnosis of SS and the gastrointestinal surgical procedure performed 2 months ago were considered as risk factors and indicators of poor prognosis.

1999 American College of Rheumatology identified 7 types of peripheral neuropathy associated with SLE: Acute inflammatory demyelinating polyradiculoneuropathy GBS, autonomic disorder, mononeuropathy, myasthenia gravis, cranial neuropathy, plexopathy, polyneuropathy. Coexistence of GBS and SLE is extremely rare among these. SLE is an autoimmune systemic disease with complex and varied clinical symptoms. Since GBS-like acute axonal neuropathies are rare in SLE patients, controlled clinical studies are insufficient. However, the relationship between SLE and GBS has been proven with the data in the literature (5).

In conclusion, it should be kept in mind in terms of early initiation of treatment and patient prognosis that there may be accompanying autoimmune rheumatologic disease in patients with GBS diagnosis. Anamnesis specific to autoimmune diseases, especially SS and SLE diseases, should be questioned and advanced laboratory examinations should be performed especially in patients with GBS, which are refractory and tend to become chronic at an advanced age.

Ethics

Informed Consent: Written informed consent form was obtained from the patient.

Peer-review: Internally peer-reviewed.

Authorship Contributions: Surgical and Medical Practices - E.A., Y.P.D., B.T.D., U.E.; Concept - E.A., Y.P.D., B.T.D., U.E.; Design - E.A., Y.P.D., Ö.B., U.E.; Data Collection or Processing - E.A., H.C., B.T.D., Ö.B.; Analysis or Interpretation - E.A., H.C., Y.P.D., U.E.; Literature Search - E.A., H.C., Ö.B.; Writing - E.A., H.C.

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Menometrorrhagia Due to Sertraline Treatment A Case Report

Sertralin Kullanımına Bağlı Menometroraji: Olgu Sunumu

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ABSTRACT

Selective serotonin reuptake inhibitors are antidepressant drugs commonly used in psychiatric diseases. The most common side effects of sertraline are diarrhea, constipation, sexual side effects, fatigue, dizziness, nausea, but sertraline-related bleeding disorder is a less common side effect. In our case, we aimed to present a case where we started sertraline, as a result of the diagnosis of depression and ıt seems abnormal bleeding other than menstrual time after the drug dose was increased called as menometrorrhagia and after the cessation of sertraline bleeding is ended.

Keywords: Menometrorrhagia, depression, sertraline

ÖZ

Seçici serotonin geri alım inhibitörleri psikiyatrik hastalıklarda yaygın olarak kullanılan antidepresan ilaçlardır. Sertralinin en sık görülen yan etkileri ishal, kabızlık, cinsel yan etkiler, yorgunluk, baş dönmesi, mide bulantısıdır ancak sertralin ile ilişkili kanama bozukluğu daha az görülen bir yan etkidir. Biz olgumuzda depresyon tanısı koyarak sertalin başladığımız, ilaç dozunun yükseltilmesi sonrasında adet zamanı dışında anormal kanama olarak tanımlanan menometroraji görülmesi ve sertralinin kesilmesi sonrası kanamanın gerilemesi sonucu sertraline bağlı olduğunu düşündüğümüz bir olgu sunmayı amaçladık.

Anahtar Kelimeler: Menometroraj, depresyon, sertralin

Introduction

Selective serotonin reuptake inhibitors (SSRI) are antidepressant drugs commonly used in psychiatric diseases (1). Sertraline is an SSRI molecule and is used in various psychiatric tables, primarily depression, due to its low side effects. The most common side effects of sertraline are diarrhea, constipation, sexual side effects, fatigue, dizziness, nausea, but bleeding disorder associated with sertraline is a less common side effect (2). Ecchymosis due to sertalin use was included in the cases (3). Here, we present a case with menometrorrhagia defined as abnormal bleeding after raising the drug dose from 50 mg/day to 100 mg/day, and whose bleeding regressed after discontinuation of sertraline, and we think that bleeding is due to sertraline.

Case Report

A 32-year-old housewife with two married children applied to our psychiatry outpatient clinic with complaints such as reluctance, loss of appetite, lack of enthusiasm, restlessness, and not enjoying life for the last 3 months. A psychiatric examination showed that her temperament was depressed and that she did not have a psychotic feature in her thought content. She didn't have a history of mental or physical illness, allergies, drug use on her resume. There was no history of mental illness in her family history. The Hamilton Depression scale applied to the patient resulted in 27 points. As a result, a diagnosis of major depression was made and sertraline 50 mg/day was started. The dose of sertraline was increased to 100 mg/day due to partial regression in her depressive complaints at the control three weeks later. When the history of the patient, who applied to our outpatient clinic again with the complaint of intense menstrual bleeding 3 weeks later, was questioned, it was learned that she presented to the obstetric and gynaecology clinic due to the excessive amount of bleeding that started ten days after her last menstruation, and no physical cause was found as a result of the examination and laboratory examinations. She was redirected to our outpatient clinic because it was thought to be due to sertraline use only. Blood tests were found within normal limits. In the laboratory examination; white blood cell: 5,750 cells/mm^3 (normal), red blood cell: 4,200,000 cells/mm^3 (normal), platelets: 220,000 cells/mm^3 (normal), prothrombin time: 11 seconds (normal), activated partial thromboplastin time: 29 seconds (normal), bleeding time: 4 minutes (normal), clotting time: 4 minutes (normal). There was no history of hematological disease in the patient's medical history and family history. It was recommended to discontinue sertraline due to the lack of features in blood tests

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suggestive of hematological disease and its onset after drug use. The patient's menstrual bleeding stopped three days later. It was learned that he had normal menstrual bleeding one month later. However, as the patient's depressive complaints increased, bupropion 150 mg/day was started. During her follow-up, her depressive complaints regressed. Off-cycle menstrual bleeding was not observed. Written consent was obtained from the patient.

Discussion

Excessive amount of menstrual bleeding before menstrual period in approximately the 3rd week of the use of sertalin without any other hematological reason, the patient's not using any other medication other than the certalin and the complaints after drug withdrawal disappear and since it was confirmed by the obstetrician, this was thought to be a side effect of drug use.

Although hematological side effects related to SSRI are quite rare, case reports are available in the literature on this subject. Hematological side effects associated with SSRI use are most commonly found in Australian drug side effect reports when examining the literature (4).

The most common hematological side effects are petechiae, purpura, ecchymosis and epistaxis. In addition to superficial bleeding, gastrointestinal bleeding and intracranial bleeding are also seen (5). In a case presented by Evans et al. (6) (1991), it was suggested that the impairment in platelet functions and prolonged bleeding time of a patient who used fluoxetine and died after a subdural hematoma may be related to fluoxetine use. Many hypotheses have been put forward and many studies have been made to explain the mechanism by which SSRIs cause bleeding disorders. Platelets contain a transporter similar to the serotonin reuptake transporter found in presynaptic nerve endings. Similar to their effect on presynaptic nerve endings, SSRI's inhibit serotonin reuptake transporter in platelets, reducing or depleting the serotonin reserves of platelets. Since the presence of serotonin is necessary for platelet clustering and hemostasis function, the activity of platelet aggregation and hemostasis is also reduced as a result of decreased or depletion of the amount of serotonin (7). In our case, there were no signs of ecchymosis or superficial bleeding.

When the literature is reviewed, it is seen that there is abnormal vaginal bleeding due to antidepressants in a small number of cases (8). There is no case in the literature about the effect of using sertalin to cause heavy bleeding, except when it increases the amount of menstrual bleeding. However, in one study, it was found that the increase in serotonin activity in the central nervous system after SSRI treatment increased prolactin level by stimulation of dopamine inhibition and prolactin releasing factors; and it has been reported that the probable cause of heavy breakthrough bleeding may be that it increases the levels of luteinizing hormone, prolactin, estradiol, progesterone and inhibits gonadal steroid metabolism (9). In our case, data could not be obtained because these hormone levels could not be looked at.

In our case, as a result of menometroragia that develops due to the use of sertraline, serotonin was discontinued and replaced by bupropion. Bupropion was recommended as SSRI without bleeding side effects when examining the literature (10) but in a meta-analysis, 3,981 studies were examined and no significant difference was found between SSRIs in terms of bleeding risk (11). However, in our case, vaginal bleeding did not recur with bupropion.

As a result, SSRI and sertraline increase the risk of bleeding. Sertraline and other SSRIs inhibit serotonin reuptake into platelets, affecting normal platelet clustering, which can result in bleeding, purpura, petechiae, ecchymosis. It is believed that this effect depends on the dose of the drug (12). Therefore, in cases where high doses are required, possible hematological side effects should be considered. We recommend that it be used more carefully in the selection of antidepressants in patients with low platelet count and/or suspected platelet dysfunction.

Ethics

Informed Consent: Written consent was obtained from the patient.

Peer-review: Externally peer-reviewed.

Financial Disclosure: The author declared that this study received no financial support.

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Obesity Treatment Can Also Cure Type 2 Diabetes Mellitus: A Case Report

Tip 2 Diabetes Mellitusu Olan Obezite Hastasında Obezite Tedavisi Diyabetide Tedavi Edebilir: Bir Olgu Örneği

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ABSTRACT

Obesity is a leading risk factor for type 2 diabetes mellitus (DM). Lifestyle modification should be the first step of diabetes treatment regardless of the medications prescribed upon diagnosis. An obese patient who was diagnosed with type 2 DM during visit at his obesity outpatient clinic was prescribed medication and lifestyle modification. The patient achieved remission of diabetes after losing 25% of weight during the follow-up period. This case emphasises the importance of lifestyle modification in diabetes treatment.

Keywords: Obesity, type 2 diabetes mellitus, lifestyle modification

ÖZ

Obezite, tip 2 diabetes mellitus (DM) için önde gelen risk faktörüdür. Tanı konduğunda hastaya önerilen medikal diyabet tedavisi ne olursa olsun, yaşam tarzı değişikliği tedavinin birinci basamağını oluşturmalıdır. Burada; obezite polikliniğine başvurusunda tip 2 DM tanısı konan, yaşam tarzı değişikliği ve ilaç tedavisi başlanıp %25 kilo kaybı sonrasında ilaçsız izlemde diyabet kliniği bulunmayan bir olgu ele alınarak, diyabet tedavisinde yaşam tarzı değişikliğinin öneminin bir kez daha vurgulanması amaçlanmıştır.

Anahtar Kelimeler: Obezite, tip 2 diabetes mellitus, yaşam tarzı değişikliği

Introduction

Obesity is the leading risk factor for type 2 diabetes mellitus (DM). Regardless of the medical diabetes treatment suggested to the patient when diagnosed, lifestyle change should be the first step of treatment. In this study, it is aimed to emphasize once again the importance of lifestyle change in the treatment of diabetes by addressing a case who was diagnosed with type 2 DM at the admission to the obesity outpatient clinic, who did not have a diabetes clinic in drug-free follow-up after a 25% weight loss with lifestyle change and medication.

Case Report

A 34-year-old male patient applied to the obesity outpatient clinic for the first time in February 2016. The patient was a high school graduate, working in the textile industry, and was married. His childhood weight was normal; she started to gain weight after marriage and reached his highest weight in the last 5 years. He applied to our outpatient clinic because he was at the highest weight in his life and started to have difficulty in movements. His medical history was unremarkable except for tonsillectomy operation. He did not have a diagnosis or a history of drug use that could cause weight gain. He had never attempted to lose weight before. In his family history, there was obesity and type 2 DM in the mother. He did not smoke or drink alcohol.

Eating habits: He ate 3 main meals a day, but his biggest meal was dinner. He continued to eat every night at home until he fell asleep, and during daylight hours at work, between meals, and these were usually simple carbohydrates (such as wafers, sweets, cakes, biscuits). There were no signs of eating disorders.

Physical activity habits: He was not doing exercises. He was using public transport on his way to work and walked between transport points and home/work.

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Psychological status: There were no signs of depression or mood disorder.

Motivational status: The patient wanted to voluntarily participate in a weight loss program and lose weight only for his health, he was motivated.

Physical examination: Height: 174 cm, weight: 112 kg, body mass index (BMI): 37 kg/m^2, waist circumference: 116 cm, hip circumference: 120 cm. Other physical examination findings were normal except arterial blood pressure with 140/100 mmHg.

Hemogram, routine biochemistry, hemoglobinA1c (HbA1c), 2nd hour postprandial blood glucose, hormone profile, then 1 mg dexamethasone suppression test, spot urine protein/creatinine were checked, electrocardiography, echocardiography, 24-hour ambulatory blood pressure monitoring and abdominal ultrasonography were performed.

After the results were evaluated, he was diagnosed with type 2 DM, hypertension (HT) and hyperlipidemia (HL) according to current guidelines (1-3). Drug treatment was started with metformin and olmesartan/thiazide. The patient refused to receive HL therapy. Simultaneously, the patient was given medical nutrition therapy, exercise prescription, and training for behavior change.

Patient made great adjustment to recommendations and came to the doctor and dietician controls regularly. He dropped from 112 kg to 84 kg, giving a total of 28 kg in the 3-year follow-up. When he had 2nd degree obesity, BMI decreased to 28 kg/m^2 and this switched him to overweight group (Table 1). HbA1c was 6.2 in May 2016 and was 5.4 in November 2016, and blood pressure was low. Drug treatments were gradually discontinued when the patient was being monitored. With continued lifestyle change, there was no increase in blood sugar (BS), A1c and blood pressure (Table 2). Scheduled last application date: July 2019 and the patient is still in the control program.

Informed consent was obtained from the patient.

Discussion

Obesity is the main determinant of poor metabolic control in type 2 diabetes. Therefore, prevention and treatment of obesity is of great importance in the management of patients with type 2 diabetes. (4)

5-10% weight loss can lower HbA1c levels, improve cardiovascular risk factors, reduce the use of antihyperglycemic, antihypertensive and lipid-lowering drugs within 1 year, and bring type 2 diabetes into remission within the first 5 years (5,6).

Table 2. Metabolic changes

Parameter	First application	Last control
FBS (mg/dL)	126	79
HbA1c (%)	7.5	5.2
TG (mg/dL)	208	67
LDL (mg/dL)	136	92
HDL (mg/dL)	31	47
Insulin (iu/mL)	43.93	4.33
GGT (mg/dL)	59	33

FBS: Fasting blood sugar, HbA1c: hemoglobinA1c, TG: triglyceride, LDL: low-density lipoprotein, HDL: high-density lipoprotein, GGT: gamma-glutamyl transferase

Many studies have shown that the combination of energy-restricted diets and exercise has an additive effect on weight loss. The beneficial metabolic effects of the Mediterranean diet have been identified and may delay the need for antihyperglycemic drug therapy in newly diagnosed type 2 diabetes patients (5,6).

In our patient, after weight loss, the amount of medication used for diabetes and HT was first reduced and then stopped without the need for medication. BS and blood pressure arterial values were normal in follow-up without medication; diabetes and HT went into remission with the continuation of lifestyle changes.

Medical nutrition therapy, physical activity and behavior change constitute the basis of obesity treatment. The first step of type 2 DM treatment is to make these changes and to provide weight loss in patients with obesity/overweight. Permanent lifestyle change is the greatest success in both diabetes and obesity treatment.

Ethics

Informed Consent: Informed consent was obtained from the patient.

Peer-review: Externally peer-reviewed.

Authorship Contributions: Surgical and Medical Practices - F.A.; Concept - F.A.; Design - F.A.; Data Collection or Processing - F.A., H.U.A.; Analysis or Interpretation - F.A., H.U.A.; Literature Search - F.A., H.U.A.; Writing - F.A., H.U.A.

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Table 1. Weight controls of the patient

Date	02.2016	05.2016	8.2016	11.2016	01.2017	02.2017	04.2017	06.2017
Weight (kg)	112	96	91	88	90	90	91	91
Date	**07.2017**	**10.2017**	**12.2017**	**02.2018**	**05.2018**	**8.2018**	**01.2019**	**07.2019**
Weight (kg)	90	94	86	82	82	85	84	84

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A Rare Disease in the Differential Diagnosis of Chylothorax: Waldenström's Macroglobulinaemia

Şilotoraks Ayırıcı Tanısında Nadir Bir Hastalık: Waldenström Makroglobulinemisi

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ABSTRACT

Waldenström's macroglobulinaemia is a disease in the B lymphoproliferative diseases group with immunoglobin M monoclonality and may present with different clinical manifestations. A 52-year-old male patient presented with complaints of weight loss and shortness of breath. Computed tomography (CT) results revealed a mass with a malignant soft tissue density in the abdomen. In the thoracic images, free fluid of up to 130 mm was found between the pleural leaves on both sides. His biopsies from the abdominal mass and bone marrow were reported to be compatible with a lymphoplasmocytic lymphoma in the presence of plasmoid differentiation B cell neoplasia and a Waldenström clinic. Cyclophosphamide -adriamycin -vincristine -prednisolone regimen was used in the first cycle chemotherapy. Positron emission tomography/CT imaging, which was performed on the 15th day after the second cycle of chemotherapy, showed that the patient's tumour was stable in size, but metabolic partial regression was observed. Bortezomib-dexamethasone-rituximab regimen was planned because of the resistant chylothorax. The patient had a dramatic clinical response and the chylothorax regressed completely after the first cure treatment. Chylothorax is a rare clinical presentation with chyle in the pleural area. It has traumatic and non-traumatic causes. Non-traumatic causes are most frequently seen due to malignancies. Waldenström's macroglobulinaemia has a very rare incidence, and the clinical association between Waldenström's macroglobulinaemia and chylothorax is very interesting. Standard treatment regimens vary, but the response to treatment also varies. Our case was resistant to the first line treatment, but had a dramatic response to the bortezomib and rituximab-based treatment.

Keywords: Waldenström's macroglobulinaemia, chylothorax, bortezomib, hyperviscosity

ÖZ

Waldenström makroglobulinemisi, immünoglobulin M monoklonalitesi gösteren B lenfoproliferatif hastalıklar grubunda bulunan bir malignitedir ve farklı klinik belirtilerle ortaya çıkabilir. Elli iki yaşında erkek hasta, kilo kaybı ve nefes darlığı yakınmalarıyla kliniğimize başvurdu. Bilgisayarlı tomografi (BT) ile yapılan görüntülemelerinde; batında geniş yer kaplayan malign görünümlü bir kitle tespit edilirken, toraks görüntülerinde, bilateral plevral yapraklar arasında 130 mm'ye varan serbest sıvı saptandı. Abdominal kitle ve kemik iliğinden alınan biyopsilerinin, plazmoid farklılaşma gösteren B hücre neoplazisi ve Waldenström kliniği ile birlikte değerlendirildiğinde lenfoplazmositik lenfoma ile uyumlu olduğu bildirildi. İlk kürde siklofosfamid -adriamisin-vinkristin -prednizolon rejimi tercih edildi. İkinci siklus kemoterapi sonrasındaki 15. günde yapılan pozitron emisyon tomografisi/BT görüntülemesinde, hastanın tümör boyutu stabil iken metabolik olarak parsiyal regresyon gözlendi. Dirençli silotoraks kliniği nedeniyle bortezomib-deksametazon-rituksimab rejimi planlandı. İkinci seri bu tedavi ile dramatik klinik yanıt elde edildi ve ilk tedavi sonrası şilotoraks tamamen geriledi. Şilotoraks, plevra yaprakları arasında şilöz materyalin birikimi ile görülen nadir bir klinik tablodur. Travmatik ve travmatik olmayan nedenleri vardır. Travmatik olmayan nedenleri en sık malignitelerin oluşturduğunu görmekteyiz. İki ayrı nadir antite olarak, şilotoraks ve Waldenström makroglobulinemisinin görülmesi oldukça ilgi çekicidir. Standart tedavi rejimleri değiştiği gibi, alınan yanıtların da farklı olduğu gözlenmektedir. Olgumuzda birinci basamak tedaviye direnç gözlenmiş olup, bortezomib ve rituksimab bazlı tedaviden ise dramatik yanıt alındı.

Anahtar Kelimeler: Waldenström makroglobulinemisi, şilotoraks, bortezomib, hiperviskozite

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Introduction

Chylothorax is the accumulation of chyle in the pleural area. While short and medium chain fatty acids are converted to free fatty acids by intestinal lipases and absorbed into the portal circulation, larger molecules are absorbed into the thoracic duct system in the form of chylomicrons together with the lower limb lymphatic drainage. Traumatic or non-traumatic damage of this lymphatic system results in the accumulation of chylous material in the pleural cavity. Traumatic damage is most frequently caused by surgery, while non-traumatic damage is most frequently caused by malignancies (1).

Waldenström's macroglobulinemia is a disease in the B lymphoproliferative diseases group with immunoglobin M (IgM) monoclonality and the production of IgM heavy chain and light chain (2). The clinical presentation of the disease varies, including fever, weight loss-like constitutional symptoms, symptomatic anaemia, thrombocytopenia, amyloidosis and related organ involvement symptoms, hyperviscosity, lymphadenopathy, organomegaly, etc, it may present with different clinical manifestations. The cure expectancy for Waldenström's macroglobulinaemia is still not expectable (2). In this case report, we present a patient with a clinic of Waldenström macroglobulinaemia who presented with an aggressive lymphoblastic lymphoma associated with an aggressive and refractory clinic of a chylothorax.

Case Report

A 52-year-old male patient applied to the general surgery outpatient clinic in June 2019 with complaints of weight loss and shortness of breath. Computed tomography (CT) results revealed a mass with a malignant soft tissue density, extending from the thoracic level along the paravertebral area to the inferior area, surrounding the aorta and its branches in the retroperitoneum and also surrounding the iliac arteries. In the upper abdomen, a mass lesion extending towards the portal hilar area and along the superior mesenteric artery axis was extended to the midline of the abdomen. In the thoracic images, a free fluid of up to 130 mm was found between the pleural leaves on both sides, and a view of atelectasis was observed at the level of the adjacent lower lobe segment.

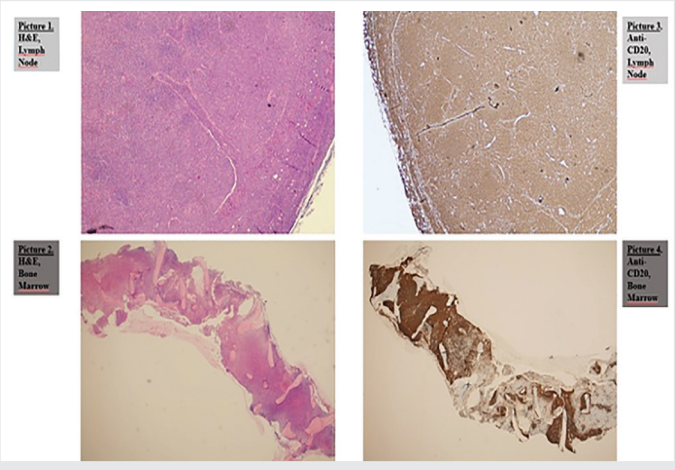

Figure 1. Images of lymph node and bone marrow biopsy sections with hematoxylin-eosin and CD20 with immunochemistry

The patient was referred to the haematology clinic after a biopsy of the abdominal mass and bone marrow was performed. Biopsy from the mass was reported to be compatible with a lymphoplasmocytic lymphoma in the presence of plasmoid differentiation B cell neoplasia and Waldenström clinic. Bone marrow biopsy showed a low-grade CD20 (+) lymphoid infiltration in the bone marrow and was reported to be in favour of Waldenström's macroglobulinaemia involvement (Figure 1). In addition to anaemia and the sedimentation rate, the serum IgM level was 11 g/L and the free kappa level was 137 mg/L. Furthermore, total protein was 9.2 g/dL, albumin was 3.5 g/dL, globulin was measured as 5.7 g/dL and aBeta 2 microglobulin was 8.8 mg/L. Bilateral evacuating thoracentesis was performed because of the bilateral massive pleural effusion causing respiratory distress and desaturation (Figure 2). A catheter was inserted and the pleural effusion was evaluated to be compatible with a chylothorax (Figure 3). A nephrostomy catheter was inserted because of bilateral grade 3 hydronephrosis, predominantly on the left side due to compression from the abdominal mass.

Cyclophosphamide -adriamycin -vincristine -prednisolone (CHOP) regimen (cyclophosphamide 750 mg/m²/day intravenous (iv). 1st day, adriamycin 50 mg/m²/day iv.1st day, vincristine 1.4 mg/m² /day iv. 1st day (maximum 2 mg) prednisolone 100 mg/day iv (days 1, 2, 3, 4, 5) was used in the first cycle. Positron emission tomography (PET)/CT imaging, which was performed on the 15th day after the second cycle of chemotherapy, showed that the patient's tumour was stable in size, but metabolic partial regression was observed. Bortezomib-dexamethasone-rituximab (BDR) regimen (bortezomib 1.3 mg/m², dexamethasone 40 mg iv day 1, 4, 8 and 11; rituximab 375 mg/m² iv day 11) was planned because of the therapy-resistant chylothorax originating from the thoracic tube. His mass was the only metabolic responsible and did not diminish in size. The patient had a dramatic clinical response with a complete regression of the chylothorax after the first cure treatment. Talc plorodesis was performed by pulling the chest tube, and the nephrostomy catheter was also removed. The patient was discharged to continue treatment as an outpatient. A written informed consent was obtained from our patient.

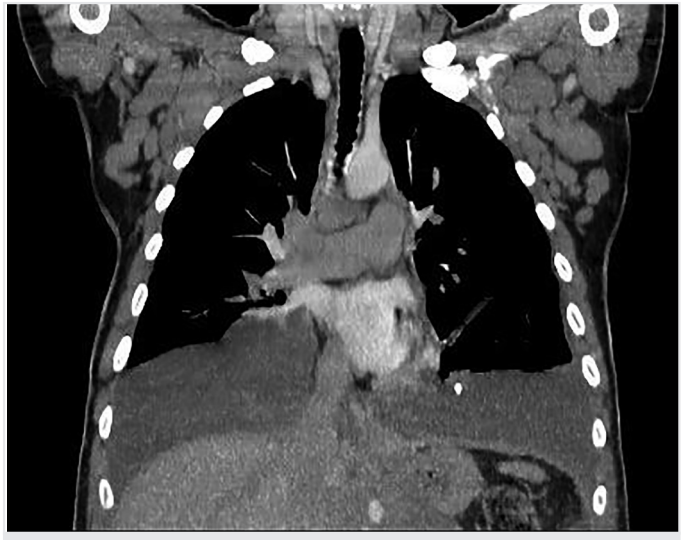

Figure 2. Computed tomography image during diagnosis: bilateral chylothorax

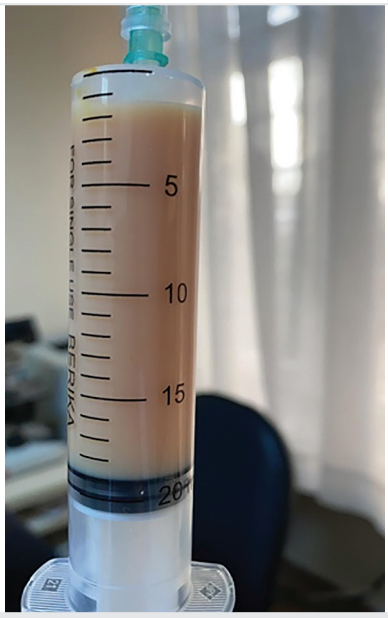

Figure 3. The appearance of chylothorax after a diagnostic percutaneous thoracic puncture

Discussion

Chylothorax with chyle in pleural area is a rare clinical presentation. It is also a rare cause of pleural effusion with chyle leakage into the pleural space. It has both traumatic and non-traumatic causes. Among surgical factors, thoracic surgical procedures are the most common traumatic factors (3,4). Non-traumatic causes are most frequently seen due to malignancies. Given that Waldenström's macroglobulinaemia also has a very rare incidence with 0.38/100,000 (5), the clinical association between Waldenström's macroglobulinaemia and chylothorax is very interesting.

The second important point to highlight in our case is the response to the preferred treatment regimen. For this case with Waldenström's macroglobulinaemia, CHOP therapy was chosen as the preferred treatment since it does not contain rituximab, due to the risk of developing hyperviscosity in line with the recommendations of current treatment guidelines, followed by rituximab CHOP in the second cure. Due to the persistence of the pleural effusion and chylothorax, the patient was started on a BDR chemotherapy regimen recommended as the second-line chemotherapy treatment (6-8). We had a good clinical response with the BDR regimen, which is consistent with the literature. In the literature, similar rare cases presenting with chylothorax have been. In these case reports, it is noted that regimens based on bortezomib and its combinations are recommended.

Chylothorax is a very rare clinical presentation, and traumatic factors play a major role in its aetiology. Although malignancies lead to non-traumatic causes, Waldenström's macroglobulinaemia is a rare cause. Standard treatment regimens vary, and responses to treatment also vary. Our case was resistant to the first line treatment, but showed a good clinical response with a bortezomib and rituximab-based treatment.

Ethics

Informed Consent: A written informed consent was obtained from our patient.

Peer-review: Internally peer-reviewed.

Authorship Contributions: Concept - İ.S., M.H.D., H.G., H.M., O.Y.; Design - İ.S., M.H.D., H.G., H.M.; Data Collection or Processing - İ.S., H.M.; Analysis or Interpretation - İ.S.; Literature Search - İ.S., H.M.; Writing - İ.S., M.H.D., H.G., H.M., O.Y.

Conflict of Interest: No conflict of interest was declared by the authors.

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CADASIL Syndrome Presenting as Adjustment Disorder

Uyum Bozukluğu Tablosu ile Prezente Olan CADASIL Sendromu Olgusu

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ABSTRACT

Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL), usually referred to as CADASIL, is an inherited autosomal dominant condition that can cause stroke, dementia and other neurological impairments. Clinically, it presents most frequently as neurogical disturbances, although psychiatric symptoms may also be seen in about 20%-41% of cases. Psychiatric disturbances encountered most often in CADASIL include mood and adjustment disorders, particularly depression.

In this case report, we highlight a patient with CADASIL syndrome, who had adjustment disorder with neurological signs appearing only later, leading to diagnosis of CADASIL. Treatment of the condition and patient follow-up are also discussed below.

Keywords: CADASIL, adjustment disorder, depression

ÖZ

Subkortikal enfarkt ve lökoensefalopati ile giden otozomal dominant serebral arteriyopati (CADASIL) yetişkinlerde inme ve vasküler demansın en sık ailesel nedenidir. CADASIL tanısı almış hastalarda nörolojik bozukluklar daha ön planda olsalar da psikiyatrik bozukluklara rastlanma oranı %20-%41 arasında değişmektedir. Duygudurum bozuklukları ve uyum bozuklukları en sık eşlik eden psikiyatrik bozukluklardır. Bu yazıda depresif mizaçlı uyum bozukluğu tanısı ile yatışı yapılan hastada ortaya çıkan nadir rastlanan bir nörolojik hastalık olan CADASIL olgusu ve tedavi ve izlem süreci tartışılacaktır.

Anahtar Kelimeler: CADASIL, uyum bozukluğu, depresyon

Introductıon

Autosomal dominant cerebral arteriopathy and a small vascular disease, leading to subcortical infarction and leukoencephalopathy cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is the most common familial cause of stroke and vascular dementia that develops due to small vessels in adults (1). Clinically; it is characterized by migraine, lacunar stroke or transient ischemic attack, cognitive disorders and psychiatric disorders (2). CADASIL is caused by a mutation in the *NOTCH3* gene located on the 19th chromosome (3). Although CADASIL is clinically observed in the fourth and fifth decade, migraine attacks with aura are observed in the second and third decade. It has also been suggested that sporadic hemiplegic migraine cases that occur without brain imaging findings may be an initial sign of CADASIL (4). Cognitive impairments seen in CADASIL occur especially in executive functions and attention deficit (5). Widespread hyperintense areas in the white matter are observed in T2-weighted magnetic resonance (MR) imaging of 90% of the patients. The incidence of hyperintense areas increases dramatically with advancing age (6). In our case, the inability to access T2-weighted MR images constituted the limitation of our article. Pathological examinations showed that it is a non-atherosclerotic disease that does not contain amyloid and affects the walls of small arteries (7). The pathogonomic finding for CADASIL is the demonstration of granular osmophilic material in the tunica media base of the cerebral artery under an electron microscope (8). Although neurological disorders are at the forefront in patients diagnosed with CADASIL, the incidence of psychiatric disorders ranges from 20% to 41%, and psychiatric findings constitute one of the cardinal characteristics of the disease. The most common of these are mood disorders and adjustment disorders with a rate of 9-41%. A smaller proportion are also accompanied by psychotic symptoms (9). In this article, a CADASIL case with depressive temperament adjustment disorder will be presented and psychiatric symptoms that can be observed in a rare neurological disease, as well as the diagnosis, treatment and follow-up process will be discussed.

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Case Report

Forty-year-old, married, without children, fifth of seven siblings, female patient was admitted to the psychiatric training and research hospital inpatient clinic with complaints such as malaise, despair, suicidal thoughts, inability to leave the house, inability to do household chores that have continued for the last 3-4 months.

In his psychiatric evaluation, it was observed that the patient, who looked at her age and was respectful to the interviewer, was conscious and oriented, and her self-care was adequate and at her age. During the interview, affect was evaluated as tearful and her mood as dysphoric. Although the speed and amount of speech decreased, the patient, whose associations were regular, could turn towards the goal. In her thought content, there were themes of psychosocial stressors that developed due to the relationship of her husband with another person and the threat of the other party's family to her and her sisters. She didn't describe delusions or hallucinations. When the patient's life history was deepened, it was learned that she had been admitted to the outpatient clinic with existing complaints a few months ago and was taking sertraline 100 mg/day and mirtazapine 15 mg/day. The patient was diagnosed with depressive temperament adjustment disorder, as it was observed that the events she experienced had affected her life for the last few months, she remained between her family and her marriage due to the events, her problems increased with the thought of separation from her spouse and her functionality decreased significantly, these depressive symptoms did not occur on the basis of a personality pathology and did not result from the grief process.

In the history of the patient, it was learned that there were headache attacks that started since the age of 18 and gradually increased, preceded by nausea and flies, and that she could not write intermittently during these attacks, intermittent speech, occasional gazing on and staring. In her family history, her mother was treated for migraine-type headache with aura, and her uncle had numbness in the tongue, speech disorder, and migraine-type headache; one of her siblings occasionally complained of not being able to speak, another sibling 4-5 years ago numbness in the face, weakness in the hands; in the other sibling, it was learned that complaints such as forgetfulness, loss of strength in the arm, numbness in the lips and inability to speak were recovered from time to time.

As a result of neurology consultation examination, brain MR imaging report performed in different centers, hyperintensities in cerebral hemispheres; on the observation of hyperintense signal changes in the temporal lobes, subcortical and periventricular white matter areas, external capsules, adjacencies of the temporal, occipital and frontal horns of the lateral ventricle, and in a patchy form with diffuse confluency in the corona radiata, periventricular and subcortical white matter areas at the centrum semiovale level, it was stated that the diagnosis of CADASIL was considered. We think that the limitation of our article is that we could not access T2 flair images of our case because they were performed at a different center (Figure 1). In addition, our patient was subjected to a mini mental status examination test and it was evaluated as a mild cognitive impairment with a 24/30 result. It was learned that the diagnosis of CADASIL was confirmed by detecting *NOTCH3* gene mutation in genetic tests. After the diagnosis of CADASIL was made, it was learned that the gene analysis was performed in all 4 siblings, and the other siblings had *NOTCH3* gene mutation and the other two siblings did not admit to the hospital voluntarily.

The patient was followed up in our inpatient clinic. The patient, who was clarified to be CADASIL after neurological consultation, was referred to the neurology outpatient clinic after the completion of her acute psychiatric treatment. Our patient still continues to follow up regularly and is in remission.

Informed consent was obtained from the patient for a case report.

Discussion

CADASIL is an extremely rare genetic disease that most commonly occurs as transient ischemic attacks. It has been reported that cognitive impairment is seen in 48% of CADASIL cases, dementia seen in 28% of cases is accompanied by 90% gait disturbances, 86% urinary insufficiency, and 52% pseudolbulbar palsy. Eigthty seven percent of migraines seen in 38% of cases are migraine with aura, but migraine without aura can also be accompanied. In our case, a slight level of cognitive impairment was detected in a psychometric examination. In the family history of our case, migraine type headache present in all sisters and grandmother is consistent with CADASIL, an autosomal dominant cerebral arteriopathy. The diagnosis of CADASIL is made clinically, at a relatively young age, in the presence of lacunar infarcts that develop in the absence of any risk factors such as hypertension or arteriosclerosis, and when other members of the family have a similar situation (10). CADASIL is often accompanied by psychiatric disorders. In a study conducted by Peters et al. (11) with 80 CADASIL patients, the rate of major depression was found to be 10% (11). Bipolar mood disorder (12), psychotic disorders (13) accompanying CADASIL are included in the literature (Table 1). In our case, depressive temperament adaptation disorder is at the forefront.

In our case, the emergence of symptoms due to the marital problems he experienced in the last months and the partial benefit from the treatment made us think that she could not cope with the stress present in the diagnosis rather than the mood disorder and that the problems of adaptation were more prominent.

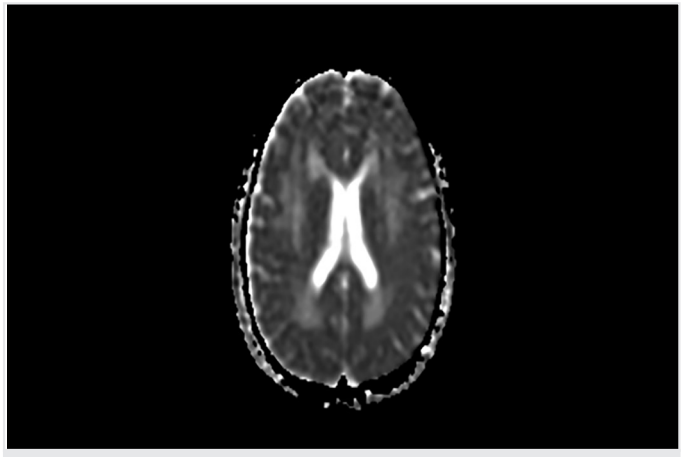

Figure 1. Cranial magnetic resonance image

Table 1. Psychiatric tables accompanying CADASIL

	Number of patients	Psychiatric disorder
Peters et al. (11) (2005)	80	Major depression (10%)
Dichgans et al. (14) (1998)	102	Adjustment disorder (23%)
Mishra et al. (13) (2018)	Case report	Psychosis
Kesebir et al. (12) (2010)	Case report	Bipolar disorder

In the study of Dichgans et al. (14), 102 CADASIL patients were followed and adjustment disorder was found in 24. We think that the reason for the adjustment disorder that developed in our case was the difficulty in coping with the stress in her life due to this disease. In one study, it was shown that the depressive complaints accompanying CADASIL negatively affect the quality of life. Quality of life is also very low in our case (15).

In conclusion, accompanying headaches in patients presenting with adjustment disorder and a similar history in family members should bring to mind the possibility of CADASIL, which is rarely encountered, and it is recommended to follow the disease with neurological follow-up and treatment and additional supportive treatments for the crisis given to the patient.

Ethics

Informed Consent: Informed consent was obtained from the patient for a case report.

Peer-review: Externally peer-reviewed.

Authorship Contributions: Concept - P.Ö.; Design - S.B.; Data Collection or Processing - S.B.; Analysis or Interpretation - E.E.; Literature Search - P.Ö.; Writing - S.B.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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Fever of Unknown Origin: A Patient with Intermittent Idiopathic Fever for Three Years

Nedeni Bilinmeyen Ateş: Üç Yıldır Ateşi Olan Bir Hasta

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ABSTRACT

Fever of unknown origin (FUO) is defined as a fever ≥38.3 ºC on at least three occasions over a period of at least three weeks, with no obvious diagnosis despite one week of inpatient investigations. Over 200 infections, neoplasm and inflammatory diseases can lead to an FUO. The definition of FUO is difficult, even though more sophisticated diagnostic tools such as positron emitted tomography and many biochemical and serological assays have become routinely available. However, it is surprising that despite the improvement in diagnostic modalities, some FUO still remain undiagnosed. When the diagnosis of FUO was not defined, invasive tests such as biopsies may be useful. Bone marrow biopsy is an accepted method in FUO to uncover haematological malignancies and certain infections. We report a case of prolonged fever 4-5 times each year for 3 years which has presented FUO. She had bone morrow biopsy and was diagnosed with T-cell large granular lymphocytic leukaemia.

Keywords: Fever of unknown origin, bone morrow biopsy, leukemia

ÖZ

Nedeni bilinmeyen ateş; ayaktan hastalarda en az üç hafta hastanede bir hafta süre ile tetkik edilmesine rağmen en az üç ölçümde ateşin ≥38,3 ºC'nin üzerinde olduğu durum olarak tanımlanır. Enfeksiyonlar, maliniteler, enflamatuvar hastalıklar olmak uzere 200 üzerinde hastalık nedeni bilinmeyen ateşin nedeni olabilir. Pozitron yayınlayıcı tomografi ve birçok biyokimyasal, serolojik ilerlemiş tanı metotlarının rutin olarak kullanılmasına rağmen nedeni bilinmeyen ateşin tanısını koymak zordur. Nedeni bilinmeyen ateşin ilerlemiş tanı metotlarına rağmen tanısı konulamadığında biyopsi gibi invazif girişimler yapılır. Kemik iliği biyopsisi ile hemotolojik maliniteler ve bazı enfeksiyonların tanısı konulabilir. Son 3 yılda, senede 4-5 kez ateşi olan bir nedeni bilinmeyen ateş olgusunu sunuyoruz. Hastada hematolojik bir malinite; T-hücre büyük granüllü lenfatik lösemi tanısı kemik iliği biyopsisi ile konuldu.

Anahtar Kelimeler: Nedeni bilinmeyen ateş, kemik iliği biyopsisi, lösemi

Introduction

Fever of unknown origin (FUO) was first described by Petersdorf and Beeson (1) in 1961.

It was defined as a fever greater than 38.3 ºC on several occasions, for a minimum duration of three weeks, without a diagnosis after intensive evaluation and diagnostic tests, and/or with an uncertain diagnosis after conducting investigations for one week in the hospital (2). This definition has been used to compare and contrast FUO in different eras, geographic locations and special patient populations (3). In addition, FUO was differentiated into four classes by Durak and Street (4) in 1991: classical FUO, nosocomial FUO, neutropenic FUO and human immunodeficiency virus (HIV)-associated FUO. Furthermore, they proposed the following definition of an FUO: a minimum diagnostic evaluation of three outpatient visits or three days of in-hospital investigations. Nevertheless, FUO has been used by clinicians to describe febrile illnesses of variable durations.

There are approximately 200 described causes of FUO, which are broadly categorised into infections, rheumatic-inflammatory diseases (such as connective tissue diseases) and malignancies (2,5). Despite extensive work-up and diagnostic advances, up to 51% of cases remain undiagnosed (2,5,6). The diagnosis of patients with FUO was made by the history and examination findings and laboratory testing, including complete blood count, blood chemistries, blood and urine cultures, C-reactive protein, erythrocyte sedimentation rate, HIV antibody, antinuclear antibody (ANA) and chest X-ray. Sometimes, bone marrow biopsy (BMB) may be needed for the investigation of haematological malignancy or certain infections such as tuberculosis (7,8).

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Here, we report the case of a 59-year-old female patient who had fever 3-4 times in each year for three years. We performed a BMB and her diagnosis was T-cell large granular lymphocytic leukaemia.

Case Report

The patient gave her informed consent.

A 59-year-old Caucasian woman with a recurrent history of chest infections was admitted to our clinic. She complained of worsening of fatigue, cough, chills and high-grade fever, which had been present for 2-3 weeks. Additionally, she had fever 3-4 times each year for three years. The complaints have been gradually increasing since last year. She was admitted to the hospital many times and treated with antibiotherapy and discharged. The patient did not report any other symptoms, including night sweats, weight loss and haemopthisis. She was not on any regular medication and had no history of exposure to ionising radiations or toxic chemicals.

She had been operated for chronic otitis media with effusion in 2010.

Her family history was unremarkable.

Her vital signs were stable. Physical examination at initial presentation revealed an awake, alert, well-nourished and fully oriented patient in no acute distress. There was no palpable lymph node and no joint deformity was observed. Fever: 39.1 ºC, blood pressure: 120/80 mmHg, heart rate: 84/minute rhythmic. On auscultation of lung heard normal breath sounds, and rare rales.

Laboratory results were as follows: complete blood count: Haemoglobin of 9.9 g/dL, total leukocyte count of 7.22×10^9/L and platelet 142,000/dL. Peripheral blood smear: 76.6% lymphocytes, 19.5% monocytes, 3.6% neutrophils, many atypical lymphocytes with azurophilic granules within their cytoplasm and few neutrophils were shown.

The absolute lymphocyte count was 5,553 and absolute neutrophil count was 260. The coagulation profile was normal. Biochemistry panel and the serologic tests for HIV antibody test and viral hepatitis C and B were unremarkable. Lactate dehydrogenase, liver and kidney function tests were normal. Rheumatoid factor, ANA and other autoimmune markers, antineutrophil cytoplasmic antibody profiles were all negative.

Thorax computed tomography (CT) imaging demonstrated lymphadenomegaly, which were not exceeding 11 mm in the mediastinum. Abdominal CT showed enlarged spleen and liver.

Positron emitted tomography (PET)/CT scan revealed multiple lymph nodes, which have increased [18F]-fluoro-2-deoxy-D-glucose in the axillary.

Flowcytometry of peripheral blood revealed a population of lymphoid cells that expressed CD2 (%) 92, CD3 (%) 90, CD5 (%) 85, CD8 (%) 78, CD (%) 45 72, TCRab (%) 94. CD4, CD56 and CD 57 were weakly positive.

The patient underwent bone marrow aspiration and biopsy and the flow cytometric study of the bone marrow aspirate. Trephine revealed slightly hypercellular bone marrow, erythroid series was regressive compared to the granulocytic series. Megakaryocytic and granulocytic populations were slightly hyperplasic. There was a noticeable increase in eosinophils. A lymphoid aggregate in the peritrabeculer area and interstitial lymphocytosis were seen. In antigenic investigation, the lymphoid population consisted of mainly CD3 (+) T lymphocytes and showed intrasinusoidal and interstitial spread. CD4 and CD56 were negative. Otherwise, CD8, perphorine, granzyme and TIA-1 were positive.

These findings were suggestive adult T-cell large granular lymphocytic leukaemia.

The patient consulted at the department of haematology and an antileukemic therapy was initiated.

Discussion

Despite the modern diagnostic tools and advanced therapeutic possibilities, FUO remain an unresolved challenge in medicine. FUO may be the symptoms of approximately 200 described causes (2,3). Aetiology may depend on the geography, because in developing countries, the percentage of infections is much higher than in developed countries, where neoplasms and inflammatory diseases are predominant (3). Although there is a common agreement that a detailed patient history and physical examination is crucial in patients presenting with unclear febrile illness. Fever pattern analysis may provide information on the diagnosis (5). The next step for diagnosing FUO is to determine the laboratory tests are that are necessary. Most authors propose multiple blood tests, urinalysis, stool tests, skin tests, cultures of different materials, chest X-ray and ultrasonography of the upper abdomen. If the diagnosis is not determined, another series of tests is proposed: ANA, rheumatoid factor, hepatitis serology, HIV serology, culture for mycobacteria, other bacteria and fungi and echocardiography. When no diagnosis if is found, CT of the neck, chest, abdomen and pelvis; and PET/CT if available, should be done. There is increasing evidence in the literature on the utility of CT and/or PET/CT in investigations for FUO (8). These imaging tests are effective in diagnosis infections, lymphoma and furthermore, the diagnosis confirmation or exclusion in 45-75% of cases. Despite the improvement of diagnostic technologies, especially imaging modalities, still are still undiagnosed FUO. If no diagnosis is achieved, some of investigators proposed a liver and BMB (8,9).

Wang et al. (10) proposed the bone marrow score to guide the need for BMB in FUO for haematological malignancy. The score included leucoerythroblastic changes in peripheral blood smear, thrombocytopenia, anaemia and neutropenia.

Previous studies have shown the presence of cytopenias to be more predictive of BMB findings for haematological malignancies (11,12). Recently, BMB was shown to be useful in several conditions including infection, malignancy and autoimmune diseases, which are all in the differential diagnosis of patients with FUO (8,13).

In the present case, adult T-cell large granular lymphocytic leukaemia was diagnosed with BMB. The diagnosis probably was delayed. She had fever 3 and 4 times each year for 3 years. In the past, she had been administered wide spectrum antibiotics and antipyretics each time it occurred. Therefore, it camouflages the presenting fever and symptom, thereby delaying the diagnosis.

It is important to search for causes of diseases in patients who had deteriorated conditions. Biopsies must be performed in difficult cases, especially when malignant diseases are suspected.

There is no single standard FUO management protocol. Empiric antibiotics are not indicated unless the patients with FUO are neutropenic. Antibiotics may delay the diagnosis of some occult infections. Empiric glucocorticoids are also not indicated unless there is a strong clinical suspicion for rheumatologic disease (5).

The most critical feature of the diagnosis of patients with FUO is to take a careful history, physical examination and to reassess the patient frequently. A wide variety of diagnostic laboratory tests and imaging studies may be useful in FUO. Cytopenic patients should also undergo BMB.

Ethics

Informed Consent: The patient gave her informed consent.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions: Surgical and Medical Practices - E.A., E.Ö., M.H.D.; Concept - E.A., D.C.Ç.; Design - E.A.; Data Collection or Processing - D.C.Ç.; Analysis or Interpretation - E.A.; Literature Search - D.C.Ç., S.H.; Writing - E.A.

Conflict of Interest: No conflict of interest was declared by the authors.

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Giant Size Foreign Body Reaction at the First Web Space in the Hand: A Case Report

El Birinci Web Aralığında Yabancı Cisim Reaksiyonuna Bağlı Büyük Kitle: Olgu Sunumu

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ABSTRACT

Mass lesions in the hand are common soft tissue problems. This case study aims to examine the giant size foreign body reaction showing atypical features at the first web space in the right hand and to emphasise different diagnosis.

Keywords: Foreign body reaction, hand, mass

ÖZ

Kitle oluşturan lezyonlar elin en yaygın yumuşak doku problemlerinden biridir. Bu olguda sağ el birinci web aralığında yabancı cisim reaksiyonuna bağlı oluşan atipik devasa yumuşak doku kitlesi sunulmuştur. Ayrıca el yumuşak doku patolojilerindeki ayırıcı tanısı vurgulanmıştır.

Anahtar Kelimeler: Yabancı cisim reaksiyonu, el, kitle

Introduction

Mass lesions in the hand are common soft tissue problems caused by benign skin and soft tissue tumours, foreign body reaction, vascular lesions, granulomatous reactions due to infection, cysts, nerve tumours, premalignant tumours, malignant skin and soft tissue tumours, soft tissue sarcomas, malignant peripheral nerve tumours and metastatic masses (1,2). However, most soft tissue lesions in the hand are benign (3,4). Furthermore, foreign body reactions are reactive traumatic masses. These lesions are the reaction of the organism to the implanted foreign body. The tissue reaction to the foreign body varies depending on the type of foreign body, contact time and anatomic location. Hence, this type of reaction is the isolation method of foreign body (5-7). This case study aims to examine the giant size foreign body reaction showing atypical features in the first web space in the right hand and to emphasise different diagnosis.

Case Report

Informed consent is taken from the patient for the case presentation below. A 58-year-old male patient with a 1-year mass at the first web space in the right hand (Figures 1, 2) that grew in size over time was examined. The physical examination of the mass was painless and palpable. During the evaluation, no signs of infection were found, but the patient's penetrating trauma story was not available. The radiopaque appearance was also not observed during a direct radiographic evaluation. However, ultrasonography examination revealed a 3x2 cm mass surrounded by hypoechoic halo.

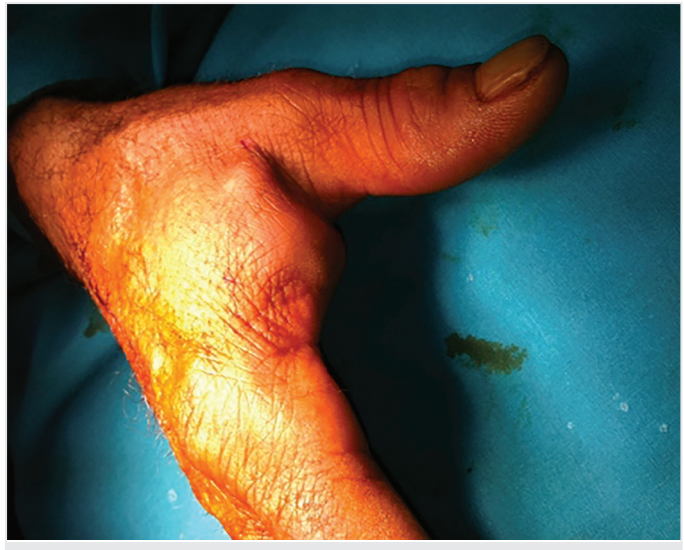

Figure 1. Growing palpable mass at the first web space in the right hand

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Thus, an excisional biopsy was planned. Under regional anaesthesia, the mass was reached with an incision parallel to the skin's stria. The encapsulated hard nodule was easily excluded from the surrounding soft tissue (Figure 3). Histopathologic examination showed the inert nature of numerous giant cells surrounding the foreign body.

Pain and cosmetic problems were resolved after surgery. Excision of the mass allowed for prompt resolution. The recurrence of the mass during the 3-year postoperative follow-up was not seen during the physical examination.

Discussion

In identifying the aetiology, it is important to demonstrate the presence of a foreign body in a radiological evaluation or penetrating trauma story (8,9). The absence of a penetrating trauma story in this case, and the fact that the dimensions are large enough, did not primarily suggest a preoperative foreign body reaction, but it may not be possible to distinguish giant size foreign body reaction from soft tissue benign tumour preoperatively (10). Excisional biopsy of the lesion allows pathological evaluation and diagnosis to be confirmed and is therapeutic. Simple excision is curative.

Although the 58-year-old male patient has no history of penetrating trauma or prior surgery, we managed to get a giant size foreign body reaction. If the mass detected is independent of the aetiology, the foreign body should be considered for the differential diagnosis. Thus, it can be successfully treated with complete excision of the mass.

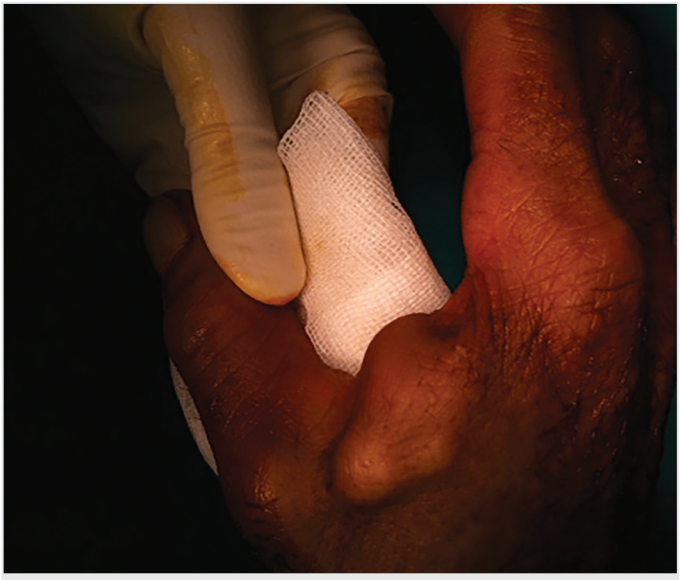

Figure 2. Physical examination of the mass located at the first web space in the right hand

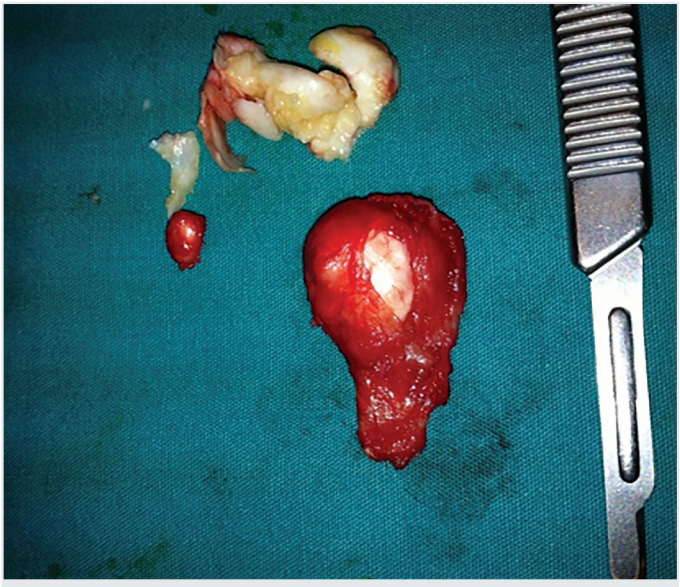

Figure 3. Macroscopic view of the excised mass

Ethics

Informed Consent: Informed consent is taken from the patient for the case presentation below.

Peer-review: Externally peer-reviewed.

Authorship Contributions: Authorship Contributions: Surgical and Medical Practices - E.C., A.B.; Concept - E.C.; Design - E.C., A.B.; Data Collection or Processing - E.C.; Analysis or Interpretation - E.C., A.B.; Literature Search - E.C., A.B.; Writing - E.C., A.B.

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Poland Syndrome with Unilateral Vocal Fold Paralysis

Unilateral Vokal Kord Paralizisinin Eşlik Ettiği Poland Sendromu

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ABSTRACT

Poland syndrome (PS) is an unusual congenital syndrome characterised the one-sided non-existence of the pectoralis major muscle, upper extremity and, rib anomaly. It is observed more frequently in men and is occasionally right-sided. Physical examination and radiological imaging are very important in diagnosing the condition. Constructive surgery is primarily the preferred in treatment. We aimed to discuss a case of PS with unilateral vocal fold paralysis for the first time in the literature.

Keywords: Poland syndrome, vocal fold paralysis, laryngeal electromyography

ÖZ

Poland sendromu (PS), unilateral pektoralis majör kasının yokluğu, aynı taraf göğüs duvarında kosta anomalileri ve üst ekstremite deformiteleri ile karakterize nadir görülen konjenital bir sendromdur. Erkeklerde daha sık görülür ve genellikle sağ tarafı tutar. Tanıda fizik muayene bulguları ve radyolojik görüntüleme yöntemleri önemlidir. Tedavisinde konstrüktif cerrahi ön plandadır. Literatürde ilk kez bildirilen vokal kord paralizisinin eşlik ettiği PS olgusunu tartışmayı amaçladık.

Anahtar Kelimeler: Poland sendromu, vokal kord paralizisi, laringeal elektromiyografi

Introduction

Poland syndrome (PS) is an uncommon constitutional abnormality, which is typically described as the unilateral non-existence of the pectoralis major muscle; non-existence of the breast and nipple; lack of the pectoralis minor muscle; anomalies of the anterior ribs, scapula and clavicle; alopecia of the axillary and mammary regions; brachydactyly and syndactyly (1,2). It occurs more frequently in men and on the right side (3). Physical examination and radiological imaging are important in making the diagnosis. In this paper, we present a rare case of PS with unilateral vocal fold paralysis and also discuss the results in the literature.

Case Report

A 58-year-old male patient suffering from dysphonia presented to our clinic. In the meantime, a right-sided chest depression was present. He had been suffering from long-term dysphonia since childhood, and had no history of surgery. On physical examination, we observed the right hemithorax depression, the non-existence of the pectoral muscles (Figure 1) and that the right nipple was upper-situated than the left nipple. Other clinical and laboratory results were within the normal range. There was no family history of laryngeal diseases. A chest radiograph showed hyperlucency of the right hemithorax, and the heart substituted left hemithorax was observed; upon this knowledge, a thorax computed tomography (CT) was planned. In thorax CT, the non-existence of the pectoralis major muscle, 3rd costal hypoplasia, and decreased attenuation of the right lung were observed (Figure 2). The right lattisimus dorsi and serratus anterior muscles were found to be normal. Cranial and neck CT performed to investigate the aetiology of the vocal fold paralysis were normal and did not yield any pathological result. Abdominal ultrasonography, breath function test, and cardiac electrocardiography-echography were also normal. On stroboscopic examination for the vocal fold paralysis, it was observed that the left-side vocal fold found in the paramedian position while the left arytenoid was displaced towards the inferomedial position (Figure 3). In laryngeal electromyography (EMG), no motor unit potentials were seen in the thyroarytenoid and cricothyroid muscles. The neural damage and denervation potentials were recorded in the laryngeal EMG. Due to the patient's request, reconstructive surgery was not performed; the patient was informed about his vocal fold pathology and recommended follow-ups with breath and voice exercise programmes. Approval (informed consent) was obtained from the patient to present this case report.

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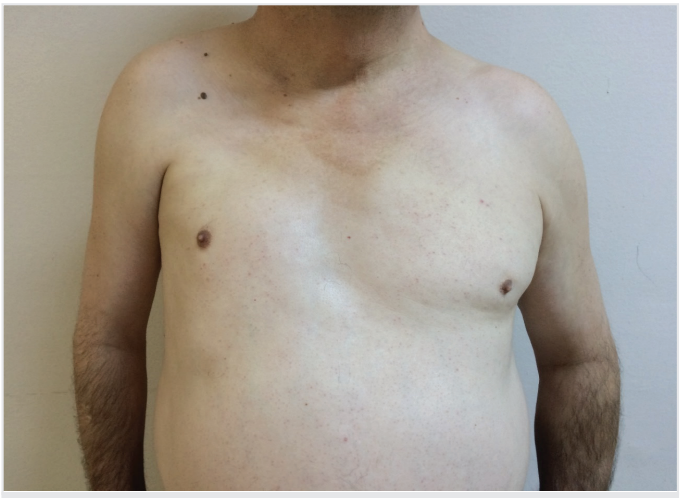

Figure 1. Chest asymmetry with right anterior chest wall depression and an absent pectoralis muscle

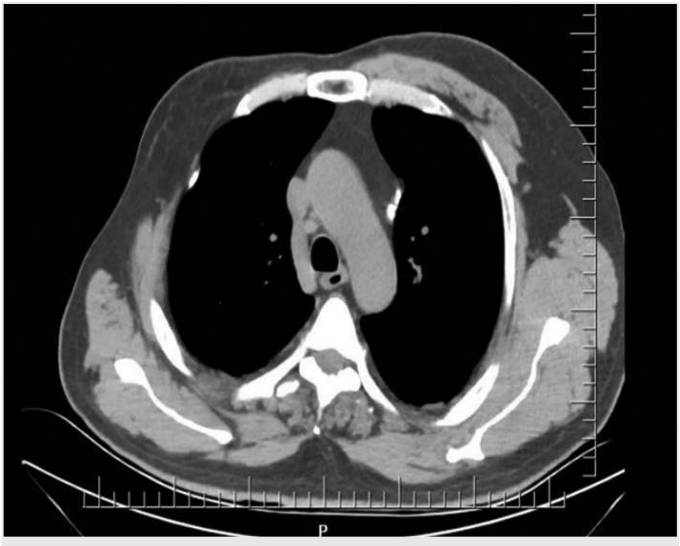

Figure 2. Thorax computed tomography of the axial plane shows the absence of the pectoral muscles in the right side

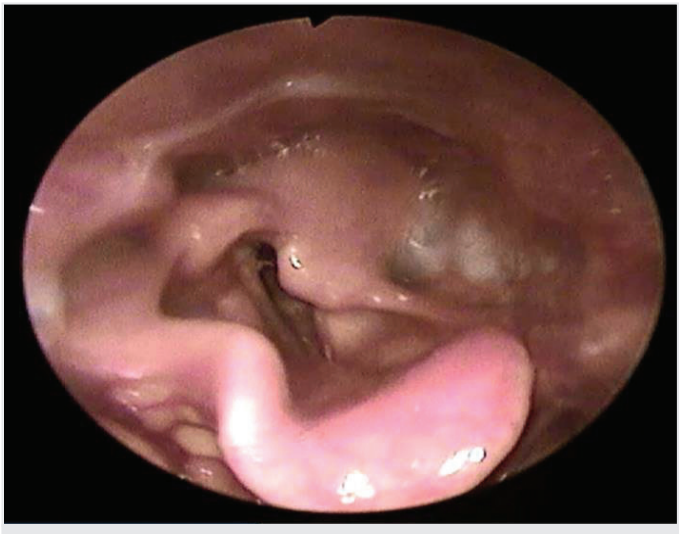

Figure 3. Endoscopic image of the left vocal cord paralysis

Discussion

PS is an unusual congenital abnormality that is defined as a one-sided thoracic development disorder (4). The non-existence of the unilateral pectoralis major muscle is the main pathological condition, and underdevelopment or non-existence of the breast and nipples often accompanies this anomaly. In addition, this syndrome may be accompanied by non-existence of the pectoralis minor muscle; deformities of the scapula, clavicle and rib; and alopecia of the axillary and mammary region. Less frequently, the latissimus dorsi, serratus anterior and external oblique muscles may also be involved (5,6). The hypoplasia of the forearm, wrist, or hand, brachydactyly and non-existence of the phalanges or digits can be seen as extremity abnormalities (7). Breast abnormalities may range from breast absence to simple breast hypoplasia (8). Our patient has non-existence of the right pectoralis major-minor muscles and third rib hypoplasia.

The incidence of PS ranges from 1/7,000 to 1/100,000 (9). PS is more common in men. It mostly affects the right side of the body and bilateral involvement is extremely rare (10). The most accepted theory is that in the sixth-seventh week of embryonic life, blood flow decreases on the affected side as a result of the abnormality of the subclavian or vertebral arteries or their branches, and this situation causes partial tissue loss in the affected region (11). Another hypothesis is that various factors like congenital vascular maldevelopment, injuries and exposure to medications in the intrauterine period may lead to hypoxia in the chest wall and adjacent extremity bud of the foetus and finally result in this syndrome (12,13). PS is generally considered to be a sporadic congenital anomaly, but hereditary cases have been reported (14). In our case, there was no hereditary factor and a right side involvement in accordance with the existing literature.

PS may be accompanied by various visceral abnormalities and neoplasms; therefore, it is important to be diagnosed. Diagnosis is made by physical examination and radiological imaging. Chest deformity, organ growth retardation and limb anomalies can be detected during the physical examination. Chest radiography may show that, radiolucency is observed to increase, and displaced to the other side of the heart. The extremity radiography may demonstrate hand anomalies. CT imaging can show muscle and bone anomalies in detail, thus providing useful data for planning a reconstructive surgery (15). In our case, the non-existence of the right pectoralis major muscle, 3rd costal hypoplasia and decreased attenuation of the right lung were observed in thorax CT.

PS is less often accompanied by other abnormalities such as renal anomalies, endocrine anomalies, dextrocardia, Sprengel deformity, Myasthenia gravis, Mobius syndrome and Parry Romberg syndrome (16). Interrelation was found with leukaemia, lymphoma, leiomyosarcoma, breast tumour and neuroblastoma, demanding heightened oncological awareness in these settings (17-19). In our case, we did not detect any symptoms related to those diseases or syndrome.

Vocal fold paralysis is frequently detected in otolaryngology practice. Although the aetiology of paralysis is variable, iatrogenic trauma, malignant tumours and infections have been commonly reported as being the cause (20,21). However, a cause may not be detected in approximately 20% of patients. The most common cause of paralysis

has been reported as surgical trauma (22). Additional findings of our patients were not available to explain the aetiology of the vocal fold paralysis.

Videolaryngoscopy is widely used as a diagnostic tool in the management of vocal fold paralysis (23). Laryngeal EMG is often used to predict the diagnosis, pathogenesis and prognosis of vocal fold paralysis (24). In patients whose paralysis has not improved, compensation mechanisms are developed to maintain the natural structure and physiology of the larynx (25). Apart from compensation, after vocal fold paralysis occurs in patients who cannot achieve complete glottal closure, some structural changes also occur. Among these changes are an asymmetric bending larynx, changes in the location of the arytenoid cartilage, changes in the vocal fold tension, granuloma formation and ventricular fold hypertrophy (26,27). In our patient, dysphonia emerged at a very early period. The patient had a left vocal fold paralysis and a shortened and altered inferomedial position of the fold in video laryngoscopic examination. Laryngeal EMG also performed on the patient, and a reduced neuronal activity was determined according to the counterparty. The patient did not want an invasive procedure and was followed-up with breath and voice exercise programmes.

Treatment options should be individualised according to the demographic characteristics of the patient and the severity of the deformity due to the variable clinical picture. Surgery is recommended in patients with advanced thoracic collapse, paradoxical thoracic movement, unprotected mediastinum, non-existence of breast tissue and cosmetic defects (28). In patients with a depressed thorax, the concave structure should be corrected by surgical repair, thereby ending the paradoxical movement of the chest wall. Functional problems in most patients may not bother patients very much, and patients may want to undergo surgery for cosmetic reasons (29). Surgical treatment may not be performed only in patients whose pectoral muscles are affected and with minimal abnormality. Flaps (lattisimus dorsi and rectus abdominus) can be used to repair the structural abnormality in the chest wall and provide functionality, or repairs can be made using only custom-made silicone prosthesis for cosmetic purposes (29). Latissimus dorsi muscle is frequently used in soft tissue repairs and successful results have been obtained (30). In our patient with the diagnosis of PS, no reconstructive surgery was performed because the patient's deformity was minimal and did not cause any functional impairment.

PS is a rare anomaly and other components of the syndrome need to be investigated in patients with thoracic abnormalities. This case report is the first study in the literature of PS accompanied by vocal fold paralysis. In our patient, the pathology associated with unilateral involvement is important in terms of showing that it may be associated with abnormalities of the contralateral side. Therefore, bilateral examinations performed in the investigation of anomalies in patients with PS would be useful.

Ethics

Informed Consent: Approval (informed consent) was obtained from the patient to present this case report.

Peer-review: Externally peer-reviewed.

Authorship Contributions: Surgical and Medical Practices - M.Y.; Concept - Z.A., B.M.Ş., Ö.Y.; Design - M.Y., B.M.Ş., Ö.Y.; Analysis or Interpretation - M.Y., Z.A., Ö.Y.; Literature Search - B.M.Ş.,; Writing - M.Y., Z.A., Ö.Y.

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Primary Spinal Epidural Lymphoma: A Case Report

Primer Spinal Epidural Lenfoma: Olgu Sunumu

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ABSTRACT

Primary spinal epidural lymphomas are the disorders that are very rarely seen. Spinal cord compression requires surgical decompression after which chemotherapy is initiated. A 46-year-old man presented with back and leg pain with progressive weakness of the lower extremities. Magnetic resonance imaging showed an extradural mass lesion compressing the cord anterolaterally and posteriorly at T-11. Surgical decompression with tumoural resection was performed. The pathological examination was reported as diffuse large B-cell lymphoma with germinal centre phenotype. Chemotherapy was initiated by the haematology clinic, and the patient was referred to physical therapy unit for his minimal paraparesis. After the operation, no pathologic involvement was observed on positron emission tomography. Patients presenting with back and leg pain should be considered for primary spinal epidural lymphoma, and the treatment should be planned accordingly since patients have good prognosis with early treatment.

Keywords: Primary spinal epidural lymphoma, thoracic spine, spinal cord compression, surgery, chemotherapy

ÖZ

Primer spinal epidural lenfomalar çok nadir görülen lezyonlardır. Omurilik basısı olması halinde cerrahi dekompresyon ve sonrasında kemoterapi başlanması gereklidir. Kırk altı yaşında erkek hasta sırt ve bacak ağrısı, bacaklarda giderek artan güç kaybı ve yürüme güçlüğü yakınmalarıyla başvurdu. Manyetik rezonans görüntülemede T-11 seviyesinde korda anterolateral ve posterior olarak basan ekstradural kitle lezyonu izlendi. Tümöral rezeksiyonla birlikte cerrahi dekompresyon yapıldı. Patoloji sonucu germinal merkez fenotipli B-hücreli lenfoma infiltrasyonu olarak geldi. Hematoloji kliniği tarafından kemoterapi başlandı. Hasta minimal parapareziyle fizik tedaviye sevk edildi. Ameliyat sonrası pozitron emisyon tomografisinde patolojik tutulum gözlenmedi. Sırt ve bacak ağrısıyla birlikte bacaklarda güç kaybı ve yürüme güçlüğü olan hastalarda primer spinal epidural lenfoma tanısı göz önünde bulundurulmalı, hastaların tedaviye iyi yanıt vermesi nedeniyle erken teşhis ile tedavi iyi planlanmalıdır.

Anahtar Kelimeler: Primer spinal epidural lenfoma, thoracic vertebra, spinal kord basısı, cerrahi, kemoterapi

Introduction

Primary spinal epidural lymphomas are rarely seen. They are reported as 0.1%-6.5% of the lymphoma cases and 9% of spinal epidural tumours (1). Primary spinal epidural lymphomas are defined as type of lymphomas with no other disease site at the time of diagnosis (2). It is thought to arise from paraspinal lymphoid tissue with secondary involvement of the spinal cord (3). Thoracic spine is mostly involved with male predominance. Clinical features may include backpain, lower limb weakness, sensory deficit and urinary or anal sphincter function disturbance (4,5).

Here, we present the case of a 46-year-old man with lower limb weakness because of a spinal extradural cord lesion which was diagnosed as germinal centre type of diffuse large B-cell lymphoma.

Case Report

A 46-year-old man was admitted to neurosurgery outpatient clinic with intense thoracic pain and difficulty in walking due to loss of strength in both legs that started a month ago. His medical history and family history were both unremarkable, except the proton pump inhibitor he was taking for gastritis.

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His neurologic examination demonstrated paraparesis of 4/5. He had a sensory level of hypoesthesia at T-10. No abnormal reflex was detected and urinary and anal sphincter tonus was normal. The other systemic findings and laboratory results were found to be normal.

A magnetic resonance imaging (MRI) was ordered as the patient was thought to have a thoracic or thoracolumbar spinal involvement because of primary spinal tumour or metastasis. MRI showed an intraspinal extradural mass lesion iso-hypointense on T1-weighted imaging (T1WI) and iso-hyperintense on T2WI at T-11 level. The lesion was homogeneously enhanced and compressed medulla especially on the anterior aspect and extended posterolaterally (Figure 1 A-D) (Figure 1 A, B: Pre-operative MRI, Figure 1 C, D: Postoperative MRI).

Weakness of lower limbs progressed in a 2 weeks' period during his diagnostic investigation, and he could not walk without assistance.

The patient was hospitalised in our neurosurgery clinic, and surgery was planned to take a biopsy both for the diagnosis and the urgent need for decompression spinal surgery. He was operated, and medullary decompression was accomplished with T-11 laminectomy and tumoural resection on the right side, where the tumoural tissue was abundant, paying special attention not to pull the spinal cord. A grey-pink, firm lesion with the bone involvement was extracted and sent for pathological examination, which was reported as a diffuse large B-cell lymphoma (Figures 2-4). Immunochemistry report showed CD20 (+), CD23 (+), CD45 (+), BCL-2 (-), BCL-6 (+), CD3 (-), CD5 (-), CD10 focal (+), CD56 (-), MUM-1 (-), C-MYC (-) of neoplastic cells. Positivity of Ki-67 in nearly all large tumour cells showed high proliferation index. The diagnosis was B-cell lymphoma infiltration with germinal centre phenotype.

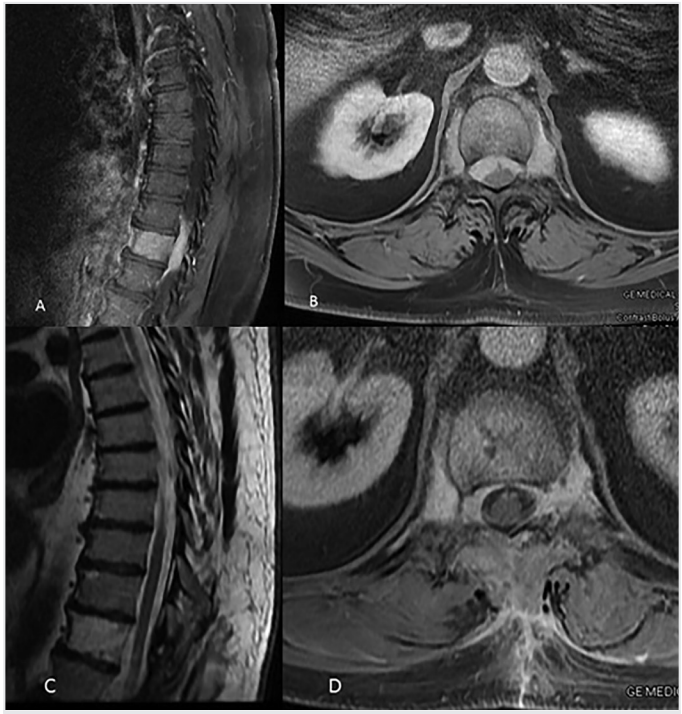

Figure 1. A, B: Sagittal T1-weighted (T1W) contrast magnetic resonance imaging (MRI) shows epidural mass compressing the spinal cord at T-11 level. Axial T1W contrast MRI shows lesion extending from anterolateral region posteriorly **C, D:** Postoperative sagittal and axial T1W contrast MRI showing the spinal cord decompression after tumour resection and chemotherapy

The patient was investigated further for other possible sites of lymphoma. No other primary site of lymphoma was revealed from the reports of whole body bone scintigraphy scan, single photon emission computed tomography (CT) and the positron emission tomography (PET), and the final diagnosis was a primary spine epidural lymphoma.

The patient quickly mobilised with little pain and was need of assistance during the early postoperative period. The patient was consulted with haematology clinic, and bone marrow biopsy was performed. The pathological result showed mild small B-cell proliferation and rare CD20 (+) large B-cells, normocellular bone marrow suboptimal for evaluation. Chemotherapy including rituximab, cyclophosphamide, doxorubicin and prednisolone (R-CHOP protocol) was initiated with the

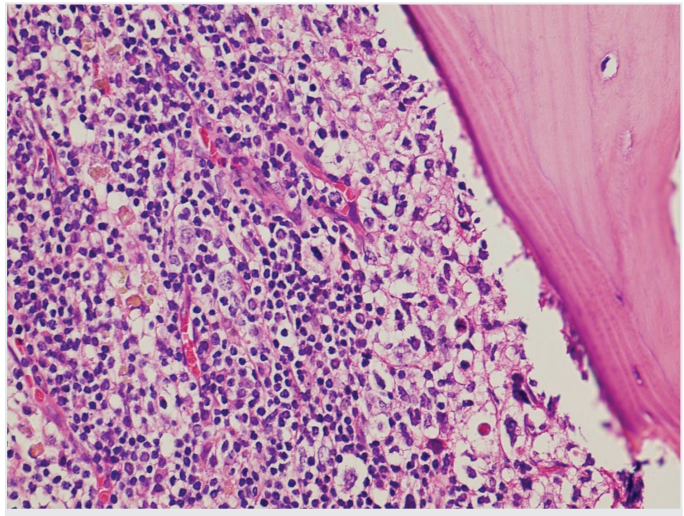

Figure 2. On the right side, bone trabeculae and at the adjacent area infiltration of the atypical lymphoid cells is observed. The infiltrative cells are irregularly bordered or lobulated lymphoid cells with clear cytoplasm and big nuclei. Lymphocytes are also observed at the distant areas of trabeculae (HEx200)

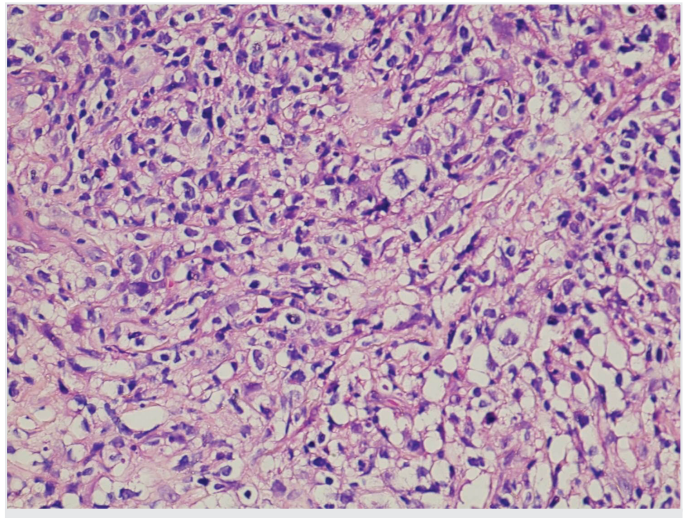

Figure 3. The area with large atypical lymphoid cells and diffuse infiltration (HEx200)

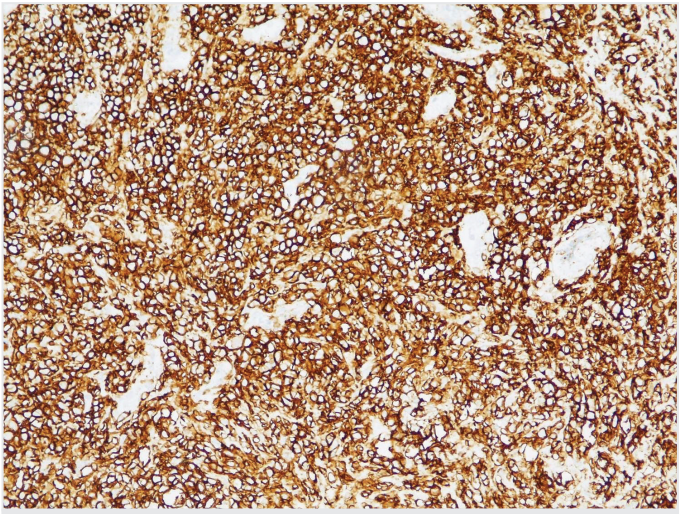

Figure 4. Lymphoid infiltration shows strong and diffuse staining with CD20 immunohistochemically (DABx200)

definitive diagnosis of B-cell lymphoma. Chemotherapy was completed to six cycles. The patient was administered high-dose methotrexate for the purpose of central nervous system (CNS) prophylaxis. PET-CT was observed to be fully responsive at the end of the treatment. After chemotherapy, he started to walk without assistance and MRI showed clearance of the tumoural tissue in the epidural region. After 2 years of follow-up period, presently, he is mobile, and has only complaint of minimal gonarthralgia, which relieves with physiotherapy.

The voluntary informed consent form was obtained from the patient.

Discussion

Extranodal non-Hodgkin lymphoma (NHL) comprises 24%-48% of all NHLs. Spinal extradural lymphomas account for 9% of all spinal epidural tumours and 0.9% of all extranodal NHLs (5). NHL of spinal epidural region is rarely seen and reported as 9% of all spinal epidural tumours (5-7). Subdural or intramedullary involvement has also been reported to be rare (8).

It is found to be more common in male than female patients with a ratio of 7:2. Age of presentation is in the fourth to seventh decade. Most common involvement is in the mid-thoracic spine (69%), followed by lumbar (27%) and cervical spine (4%) (2,5,9-11). Differential diagnosis includes metastasis, primary bone tumours, multiple myeloma and infections (4).

Primary spinal epidural NHL diagnosis is made when all other system pathologies are excluded (3). We also excluded all other etiological possibilities in our case, and the diagnosis was made after the operation and biopsy evaluation.

The pathogenesis of primary spinal epidural lymphoma was explained by the role of chronic inflammatory process, chronic infection, autoimmune disease and the meningoepithelial component (5,10). The theories about the origin are controversial. The lymphoma cells are thought to originate from lymphatic tissue along the venous plexus in the epidural space or the paravertebral lymphoid rests (2,11). Antigenic stimulation with a transformation cascade was theorised as epidural tissue contains lymphoid cells. Other suggestions include the origination of these epidural tumours from paraspinal, vertebral and retroperitoneal tissues after entering through the intervertebral foramina (9,12).

Histopathological examination of primary spinal epidural lymphoma shows atypical lymphoid cell proliferation. B-cell lymphoma is the most common type. Immunohistochemistry demonstrates tumour cell positivity for leukocyte common antigen and CD20 (13).

Spinal epidural lymphomas are generally diagnosed as B-cell lymphomas as in our case. Diffuse large B-cell lymphoma is the most common type (2), and it represents 30% of all NHLs. Spinal epidural lymphoma of this type is classified as germinal centre B-cell type or non-germinal centre B-cell type. First one is CD10 (+) or CD10 (-)/BCL-6 (+)/MUM-1 (-), and the second one is CD10 (-)/BCL-6 (-) or CD10 (-)/BCL-6 (+)/MUM-1 (-). Germinal centre B-cell type is more common in the spine compared to CNS (5,14).

Presentation with a spinal cord compression is rarely seen among NHL patients, and it is reported as 5%. Primary spinal lymphoma patients first complain about back or leg pain and difficulty in walking. Neurologic deficits as paraparesis, ataxic gait, sensory disturbance with possible urinary and bowel involvement occur later. Progression to lower limb weakness due to cord compression may be observed in a 2-week to 2-month period (2,5,15).

MRI is a useful diagnostic tool to detect the cord compression and for surgical planning. However, first, systemic primary lymphoma should be excluded. Lesion is usually hypo-isointense on T1WI and iso-hyperintense on T2WI, however, T2WI may show low signal due to the variable density of tumour cells (2,4). MRI appearance of the lesion was isointense on T1-weighted images and iso-hyperintense on T2WI with homogeneous enhancement in our case in accordance with the literature.

As an adjuvant technique, PET shows the tumour as an area with intense hyper-metabolic activity (16,17).

Since NHL is very sensitive to radiotherapy and chemotherapy, surgery is considered in patients with one site involvement, in order to confirm the diagnosis and decide for the treatment options accordingly. Surgical decompression and biopsy provide the highest possible volume reduction. Thus, it is considered as the best modality in patients with spinal cord compression. Rapid progression of motor weakness, especially with loss of urinary anal sphincter control requires emergency decompression. Patients with spinal instability may require spinal stabilisation (5,10). This approach also allows for histopathological diagnosis in a patient with no other known diagnosis related to the epidural lesion and expedites the planning of the chemotherapy and radiotherapy if needed.

Adjuvant therapy increases the disease-free survival period (18,19). Intrathecal chemotherapy may be considered in individualised treatment (2). Chemotherapy, including cyclophosphamide, vincristine and prednisone, is recommended after the surgical decompression and pathological diagnosis. Radiotherapy at doses of 25 Gy may be given as an adjuvant treatment.

Primary spinal epidural lymphoma should be considered in patients with spinal cord compression of extradural soft tissue lesion with or without bone involvement. The prognosis and functional recovery are relatively better for patients with spinal cord compression because of NHL, and it is important to make a timely diagnosis before paraplegia occurs.

Ethics

Informed Consent: The voluntary informed consent form was obtained from the patient.

Peer-review: Externally peer-reviewed.

Authorship Contributions: Surgical and Medical Practices - N.G.Ö., Ö.B., Ö.F.Ş., F.A., H.T.K.; Concept - N.G.Ö.; Design - N.G.Ö., F.A.; Data Collection or Processing - N.G.Ö., Ö.B., Ö.F.Ş., G.E.H.; Analysis or Interpretation - N.G.Ö., F.A.; Literature Search - N.G.Ö., Ö.B., Ö.F.Ş.; Writing - N.G.Ö., F.A.

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