Case Report / Olgu Sunumu

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A Rare Cause of Nasal Septal Mass: B Cell Lymphoma

Nadir Bir Nazal Septal Kitle Nedeni: B Hücreli Lenfoma

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Abstract / Özet

Lymphomas comprise 3% to 5% of all malignancies, with non-Hodgkin's lymphoma accounting for 60% of cases. The nasal cavities and paranasal sinuses are rarely affected by primary NHL. There are three subtypes of NHL on the basis of their immunohistochemical findings: B-cell lymphoma, T-cell lymphoma and natural killer (NK)/T-cell lymphoma. A 77 year-old woman presented at our clinic with a one month history of a nasal obstruction and unilateral epistaxis of the left nasal cavity. Punch biopsy reported a lymphoplasmocytic diffuse malignant infiltrate. Immunohistochemistry was strongly positive for the B cell associated markers CD20 and it was diagnosed as nasal septal B cell lymphoma.

Key Words: Non-Hodgkin's lymphoma, B cell lymphoma, nasal septum

Lenfomalar tüm malignitelerin %3-5' ini oluştururken non-Hodgkin lenfomalar (NHL) lenfoma vakalarının %60'ını oluşturur. Primer NHL nazal kavite ve paranazal sinüslerde nadiren görülür. İmmünohistokimyasal özelliklerine göre NHL'lerin 3 alt tipi vardır: B hücreli lenfoma, T hücreli lenfoma ve natural killer (NK)/T hücreli lenfoma. Yetmiş yedi yaşında kadın hasta kliniğimize bir aydır olan burun tıkanıklığı ve sol nazal kaviteden kanama ile başvurdu. Punch biyopsi sonucu lenfoplazmositik diffüz malign infiltrasyon olarak rapor edildi. Immunohistokimyasal incelemede B lenfositler ile ilişkili marker olan CD20 güçlü pozitif olarak bulundu ve nazal septal B hücreli lenfoma tanısı kondu.

Anahtar Kelimeler: Non-Hodgkin lenfoma, B hücreli lenfoma, nazal septum

Introduction

Lymphomas comprise 3% to 5% of all malignancies, with non-Hodgkin's Lymphoma (NHL) accounting for 60% of cases (1). The nasal cavities and paranasal sinuses are rarely affected by primary NHL. Nasal lymphomas differ from lymphomas that arise in the paranasal sinus and Waldeyer's ring. Primary nasal lymphomas are rare in the Western population but occur frequently in Asia, South-America and Mexico, where they constitute up to 10% of non-Hodgkin's lymphoma (2, 3).

There are three subtypes of NHL on the basis of their immunohistochemical findings: B-cell lymphoma, T-cell lymphoma and natural killer (NK)/T-cell lymphoma (4). Among subtypes seen in nasal lymphomas, the most common in the Asian population is the NK/T-cell lymphoma. The B cell lymphoma subtype is the most common in the Western population (5).

The initial symptom in most cases is nasal obstruction; hemo-purulent rhinorrhea is the second most common sign (6, 7). It is well recognized that most patients with B-cell lymphoma have a prolonged history of sinonasal symptoms due to involvement of adjacent structures such as the nasopharynx, orbit, cheek, anterior cranial fossa, and the opposite antrum as in carcinomas of the nasal cavity and paranasal sinuses (8).

Prognostic factors include tumor volume, immunophenotype and extent of lesion. Recognition from the outset of this poor prognosis subtype of lymphoma is important for more effective treatment, such as chemotherapy combined with radiation(9). With B-cell lymphomas, a better understanding of the biology of NK/T-cell tumours using cDNA arrays might help divide patients into prognostic groups or uncover novel targets and strategies for improving this rare but very aggressive cancer (10). Nasal B cell NHL is a distinct clinical and pathological entity and is an aggressive rare disorder.

Case Report

A 77 year-old woman presented with a recent onset of nasal obstruction and unilateral epistaxis of the left nasal cavity of one month's duration. She denied any nasal pain or numbness. The nasal obstruction and unilateral epistaxis had gradually increased during the last days. Her medical and family histories were unremarkable. Laboratory examination revealed an erythrocyte sedimentation rate of 113 mm/h, C-reactive protein levels 83 mg/L, white blood cell count of 11,500/mL and hematocrit of 37%.

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Endoscopic examination showed a large hemorrhagic, friable, granular, vegetative mass covered with a crust along the septum of the nose that obstructed the left nasal vault (Figure 1). Punch biopsy from the nasal tumor revealed a diffuse lymphoplasmacytic malignant infiltration of collogen bundles and angioinvasion (Figure 2). Immunohistochemistry was strongly positive for the B cell associated surface markers (Figure 3). Flow cytometric analysis of peripheral blood revealed a predominant population of B cells. Computerized Tomography (CT) imaging revealed an invasive septal mass in the left nasal cavity (Figure 4). No cervical lymphadenomegaly was observed. The patient was treated with 5000 cGy of external-beam radiation followed by four cycles of R-CHOP (rituximab, cyclophosphamide, doxorubicin hydrochloride, vincristine and prednisolone). Clinical diagnosis of this nasal septal mass revealed a B cell Lymphoma. Our patient has exhibited an aggressive course and died within six months following the diagnosis.

Discussion

The nasal cavity is the predominant site of involvement in T-cell and NK/T-cell lymphomas, whereas sinus involvement without nasal disease is more common in B-cell lymphomas (4). In our case, the nasal septum was involved. Usually, the median age at presentation is 50 years and there is a male to female ratio of 2:1 (11, 12).

Primary nasal cavity lymphoma usually presents as an ulcerative and necrotic mass at the nasal or midline facial structures and palate, often causing destruction of adjacent soft tissues, cartilage and bony structures. The nasal mucosa is usually pale ,friable and granular and is often accompanied by purulence or crusting. Oroantral fistulas can frequently occur as a result of mucosal ulceration and osseous necrosis (2). Our case showed mucosal ulceration, with a pale, friable and granular appearance. No oroantral fistula or osseus necrosis was identified.

Definitive diagnosis is based on identification of characteristic histopathologic features in surgical pathology specimens. Flow cytometry revealed a predominant population of B cells. Recently, the immunophenotype has been shown to significantly affect the outcome of this disease. Differential diagnosis includes entities that depend more upon clinical and laboratory studies (13). Histopathologic findings include diffuse lymphoplasmacytic malignant

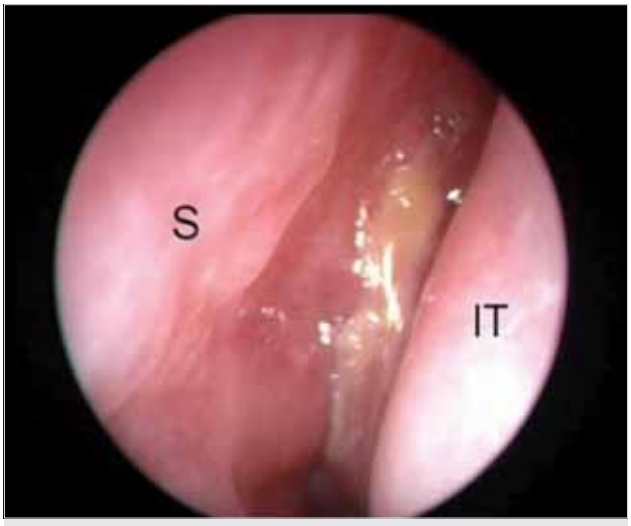

Figure 1. Extensive amount of friable, granular tissue along the septum of the nose obstructing the left nasal vault; a hemorrhagic vegetating mass covered with crust
S: Nasal septum, IT: Inferior turbinate

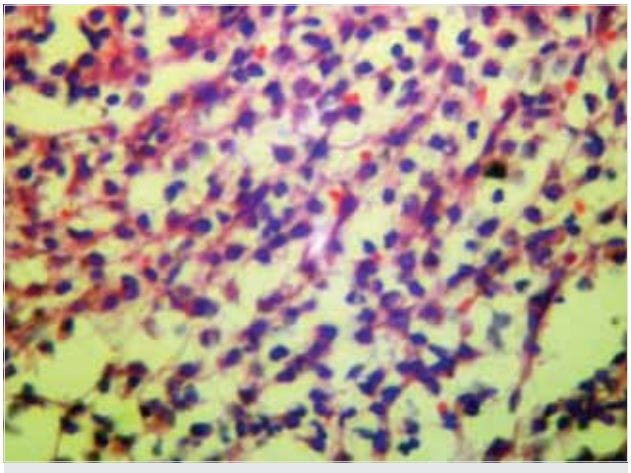

Figure 2. Diffuse lymphoplasmacytic malignant infiltration (HEX200)

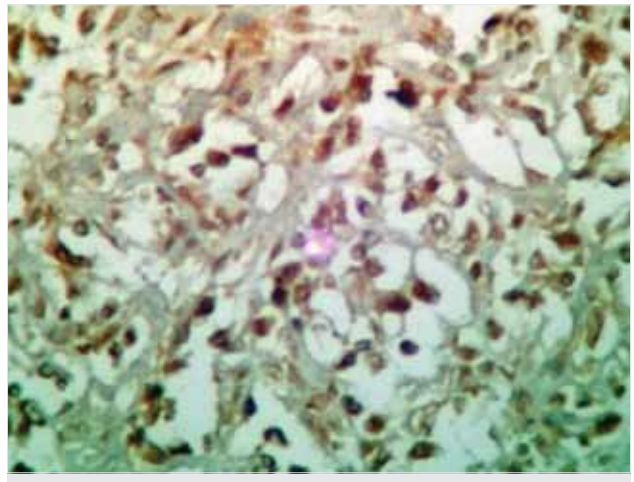

Figure 3. Immunohistochemistry was strongly positive for the specific B cell associated marker CD20

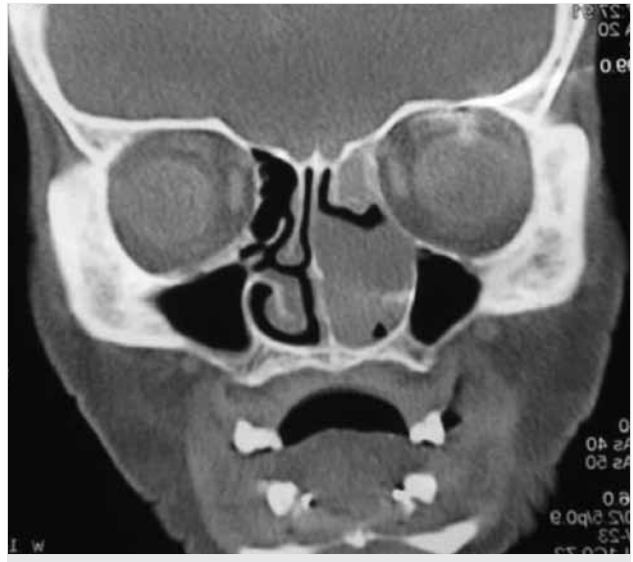

Figure 4. Computerized Tomography imaging revealed an invasive septal mass in the leftnasal cavity

infiltration and dissection of collogen bundles and angioinvasion. Immunohistochemistry was strongly positive for the B cell associated markers.

The prognosis of the sino-nasal Non-Hodgkin's lymphoma depends on the type and stage of disease, the number of sites of extra nodal spread, invasion of the central nervous system and the patient's general condition. Patients with lymphomas of high histopathologic grade and recurrent or disseminated disease have the worst prognosis. Two-third of the patients remain in the remission phase after initial therapy. In one-third of the patients, the disease relapses and three-quarters of these patients die of the disease (14). The cumulative five-year survival rates are about 30% for all types, 55% for diffuse large B-cell lymphoma, 33% for peripheral T-cell lymphoma, and 20% for angiocentric lymphoma (15). Five-year overall and event-free survival rates after treatment are 52% and 50%, respectively (16). Patients with T-cell lymphoma have a better survival rate than patients with B-cell lymphoma. NK/T-cell lymphomas exhibit a more aggressive clinical course, frequent treatment failure to radiotherapy and/or chemotherapy and are significantly associated with an extremely poor prognosis when compared with B-cell and T-cell lymphomas (4). In B-cell lymphomas, a better understanding of the biology of NK/T-cell tumours using cDNA arrays might help divide patients into prognostic groups or uncover novel targets and strategies for improving this rare but very aggressive cancer (10). A series from Japan had a 5-year survival of 25% for the nasal cavity and 85% for the paranasal sinuses, with 88% of the patients having a large B-cell subtype NHL (17).

Nasal lymphomas often present with soft tissue masses and/or local destruction/erosion of bony structures of the nasal cavity (most common sites: maxillary sinus wall, lamina papyracea, and nasal turbinate), resulting in the obliteration of the nasal passage and the adjacent paranasal sinuses in 75% of cases. Nasal B cell lymphomas constitute an uncommon type of malignant lymphoma, differing from other lymphomas that arise in the paranasal sinus and Waldeyer's ring (2, 3).

Radiotherapy and chemotherapy have been used as primary treatment of this disease. Surgery is only required in the case of life-threatening or function- threatening conditions to decompress the upper respiratory system, open the paranasal sinus, or to relieve pressure in the orbits (8).

In the present case, a surgical approach was performed for biopsy and relief of nasal obstruction and after 6 months of follow-up, the patient died due to systemic complications.

Conclusion

In conclusion, nasal B cell NHL is a distinct clinical and pathological entity classified as an aggressive extranodal malignant lymphoma. It is important for otolaryngologists and other clinicians to be familiar with the head and neck manifestations of lymphomas and include them in the differential diagnosis.

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

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