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Multiple Myeloma In A 25- Year-Old Man With Pericardial Involvement And Secondary Extramedullary Plasmacytomas

Dr. A. Naim KESKİN (1), Prof Dr. Gülten AKTUĞLU (2), Dr. Özlem SEZGİN (3), Dr. Hüseyin GÜNDÜZ (4), Dr. Yüksel BARUT (5).

ÖZET

Konjestif kalp yetersizlığı bulgularıyla hastanemize başvuran 25 yaşındaki erkek hastanın ileri tetkikinde perikardial efüzyon, litik kemik lezyonları ve her iki 6. kosta ile komşu yumuşak doku kitleleri saptandı. Hastanın laboratuvar tetkikleri sonucunda multipl myelom (MM) tanısı kondu. Bu vaka multipl myelomun modern tanı kriterlerine uyan 30 yaş altındaki nadir vakalardan birisidir. Ayrıca kalp tutulumu ve (ekstarmeduller) plazmositomalar da multipl myelomun çok seyrek görulen bulgularıdır.

Anahtar Kelimeler : *Multipl Myelom, Perikardial efuzyon, sekonder ekstramedullar plasmasitoma.*

SUMMARY

A case of multiple myeloma with pericardial effusion and seconder extamedullary plasmacytoma.

A 25- year- old young man with pericardial effusion, congestive heart failure, lytic bone lesions and multiple secondary extramedullary plasmacytomas on thoracic costae was diagnosed as having multiple myeloma (MM). This case is one of the very occassional patients younger than aged 30 years to meet modern diagnostic criteria for MM. Besides, cardiac involvement and extramedullary plasmacytomas are both rare disorders of MM?

Key Word: *Multiple myeloma, pericardial effusion, secondary extramedullary plasma cytomas.*

CASE REPORT

A 25- year- old young man was admitted to our hospital on the 25 th of July, 1994, with the complaints of shortness of breath, pain on the various parts of the body. Pain, especially on the ribs, appeared 6 months ago and he was treated with the diagnosis of myalgia. Dyspnea and orthopnea started 15 days ago and rapidly progressed.

On physical examination, he was extremely pale and in moderate respiratory distress. Blood pressure was 130/60 mm Hg, the heart rate was 120/min, and he was afebrile. CVS examination revealed 2/6 systolic and 2/6 diastolic murmurs on the left sternal side. He had jugular venous distention. The liver was palpable 4 cm below the righ costal margin. The spleen was nonpalpable. On the left axillary area, microlymphadenopathies were palpated. Other system investigations were in normal limits.

The ECG showed sinus tachycardia with negative precordial T waves. An echocardiography revealed a little pericardial effusion, dilatation of left ventricle and minimal tricuspid regurgitation (1+).

On laboratory investigation; WBC was 13.000 /mm3, Hb. 5.9 g/dl, Htc 17.9 % RBC count 223x10^{4}mm3, MCV 80fL and PLT count

(1) Director of Ist Internal Medicine Clinic in SSK Istanbul Hospital.
(2) Department of Haematology in Cerrahpaşa Medical Faculty in University of Istanbul.
(3) Resident in Ist Internal Medicine Clinic in SSK Istanbul Hospital.
(4) Resident in Internal Medicine Department of Cerrahpaşa Medical School in University of Istanbul.
(5) Director of Radiodiagnostic clinic in SSK Istanbul Hospital.

15.3x10⁴/mm3. The white cell diferrential indicated 45% granulocytes, 53 % lenfocytes, 1% monocytes and 1 % atypical mononuclear cells. Erythrocyt sedimentation rate was 150 mm/hour. Blood glucose, liver functiontests and lipid levels were in normal range.

Total serum protein was 9.4 gr/dl, albumin 4.6 gr/dl, globulin 4.8 gr/dl. The serum immunoglobulin profile was charactarized by a marked increase in Ig G fraction 2960 mg/dl (Normal range 504-1765 mg/dl), and a significant decrease in Ig A and Ig M fractions- 9 mg/dl (N: 85-385 mg/dl) and 43 mg/dl (N: 45-250 mg/dl), respectively. Serum protein electrophoresis revealed a monoclonal gammopathy on the gamma band. (4.43 gr - N: 0.7-1.5 gr). Bence-Jones protein in urine was negative.

On the chest X-ray, there was marked cardiomegaly and bilateral soft tissue masses with destructive lesions on the neighboring costae. A CAT scan revealed bilateral soft tissue masses with destructive lesions on the 6th costae. A bone survey showed lytic lesions on thoracic costae, and frontal and parietal portions of the skull.

On bone marrow aspirates, there was 9 % lenfocytes, 41 % granulocytes and 50 % plasma cells.

With the diagnosis of multiple myeloma, the patient was transferred to the Department of Haematology of Cerrahpaşa Medical School in University of Istanbul.

The clinical and laboratory investigations performed in that center supported our data and diagnosis. Spesific therapy with 15 mg Alceran and 96 mg Prednol daily was started.

At the end of the first week, there was a rapid improvement in cardiac symptoms. By repeated echocardiographies the regression of pericarditis was verified. There was a moderate increase in Hb and Htc levels.

The patient was discharged on diuretic and uricolytic therapy after the spesific cure and he was called for monthly controls.

DISCUSSION

Multiple Myeloma (MM) is characterized by the neoplastic proliferation of a single clone of plasma cells, which results in bone pain, anemia and fatique. The clinical spectrum ranges from solitary plasmacytomas to extensive bone marrow infiltrations. It is generally a disease of older adults. The median age of onset is about 60 years. MM in young people and children is exceedingly rare.

Maida and all. (1) reported a case of a 13-year-old girl of MM of the IgA monoclonal gammopathy type in 1973 In 1976, three young men whose ages were ranging from 17 to 22 years were reported as having MM by Hewell and all. (2) There were a few more cases of MM in young population in previous literature but none of them met the modern diagnostic criteria described by De Vita and all. (3) in 1989.

TABLE 1. Diagnostic Criteria for Multiple Myeloma.

Major Criteria

I. Plasmocytoma on tissue biopsy.

II. Bone marrow plasmacytosis with > 30% plasma cells.

III. Monoclanal globulin spike on serum electrophoresis exceeding 3.5 gr./dl. for Ig G peak or 2.0 gr./dl. for Ig A peaks, > 1.0 g/24 hours of kappa or lambda light chain excretion on urine electrophoresis in the presence of amyloidosis.

Minor criteria

a. Bone marrow plasmacytosis 10% to 30% plasma cells.

b. Monoclonal globulin spike present, but less than the level defined above.

c. Lytic bone lesions.

d. Residual normal Ig M<50 mg/dl, Ig A<100 mg/dl, Ig G<600 mg/dl.

Diagnosis will be confirmed when any of the following features are documented in symptomatic patients with clearly progressive disease. The diagnosis of myloma requires a minimum of one major + one minor criterion or there minor criteria that must include a + b, i.e.,

1. I+b,I+c,I+d (I+a not sufficient)
2. II+b,II+c,II+d
3. III+b,III+c,III+d
4. a+b+c, a+b+d

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In 1992 Badwey and all. (3) described a 25-year-old woman with MM and they stated thas this case was one of the eight patients yunger than 30 years to meet these criteria.

Patients with MM must be distinguished from these with monoclonal gammopathy of undetermined significance (benign monoclonal gammopathy - BMG, and smoldering multiple myeloma - SMM). This differential diagnosis is important, because patients with BMG and SMM should not be treated until abnormalities progress or symptoms of MM develop, because they may remain stable for many years. (4)

Our patient showed bone marrow plasmacytosis of > 30 % plasma cells (11), monoclonal globulin spike on serum electophoresis was>3.5 g/dl for Ig G (III), he had Iytic bone lesions (c), and residual normal Ig M and Ig A values were < 50 mg/dl and <100 mg/dl respectively (d); so he met 2 major + 2 minor criteria.

Cardiac involvement is a rare disorder of MM. In one case reported by Goldberg and alt. (5) the patient was admitted with congestive heart failure and a diagnosis of MM was also made. She died four months-

TABLE 2. Differential diagnostic criteria between BMG, SMM and MM.

	BMG	SMM	MM
Protein value	<3 gr/dl	>3 g/dl	>3 g/dl
Bone marrow	<10% plasma cells	>10% plasma cells	>10-30% plama cells
Anemia	(-)	(-)	(+)
Skeletal lesions	(-)	(-)	(+)
Uninvolved serum Ig levels	N	()	()
Bence - Jones Proteinuria	(-)	(+/-)	(+/-)
Renal insufficiency	(-)	(-)	(+)

later and necropsys showed a large pericardial effusion associated with plasma cell infiltration of the pericardium. Piney and al. (6) reported a case of MM with heart muscle involvement. Another case of MM with a tumor of pericardium surrounding the great vessels at the cardiac base was described by Santana (7).

In our case, the patient was admitted to the hospital with typical signs of congestive heart failure and pericarditis. After the diagnosis and spesific therapy of MM, the symptoms regressed rapidly and by repeated echocardiographies we verified the cardiac improvement. Although we didn't perform pericardiosynthesis and showed myloma cell infiltration of the pericardium, in accordance with the drammatic response to the spesific therapy, we concluded that these clinical symtoms were results of MM involvement of the heart.

Extramedullary plasmacytoma is another rare disorder of proliferation of plasmacytoid cells, wich develops in various extramedullary sites, especially in oronasopharynx, gastrointestinal tract, spleen, lymph nodes and skin. Primary plasmacytomas mainly occur in the upper respiratory track. The bone marrow is usually spared, but some patiens with primary plasmacytomas subsequently develop MM. In contrast, patients, with myeloma have secondary lesions by direct extension from an underlying focus in the bone or as a result of distant metastasis (8,9).

Our patient had two soft tissue masses near both right and left 6th costae showing destruction on the bone. These were assumed to be secondary plasmacytomas for there was typical bone marrow infiltration of MM at the same time. After spesific therapy therapy the patient was free of pain, but the masses remained the same.

As with of the cases previously reported, the diagnosis of MM was not initially considered in this patient because of his young age. Besides, with the limited previous data of MM in young population, no accurate statement can be made regarding the prognosis.

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