

Multiple myeloma masquerading as a pulmonary mass: A rare presentation

Akciğerde bir kitle olarak maskelenen multiple miyeloma: Nadir bir ortaya çıkış

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ABSTRACT

Multiple myeloma represents malignant disorder of plasma cells. Tumour extension is primarily seen within the bone and bone marrow, despite widespread distribution of plasma cells in the body. Metastatic deposits outside bone marrow (extramedullary) are uncommon even in advanced multiple myeloma. Involvement of pulmonary parenchyma by myeloma cells either as plasmacytoma or as a pulmonary infiltrate is rare and is related to aggressive terminal phase of the disease. We are reporting a case of multiple myeloma with a pulmonary parenchymal mass as the initial presenting manifestation.

Key words: Pulmonary, multiple myeloma, extramedullary plasmacytoma (EMP).

INTRODUCTION

Multiple myeloma is a neoplasm of B cell lineage characterised by excessive proliferation of abnormal plasma cells involving primarily the bone marrow. These malignant plasma cells secrete an abnormal immunoglobulin causing a monoclonal gammopathy.¹ The disease process mainly involves the axial skeleton. The occurrence of extramedullary disease is uncommon in multiple myeloma.² Reported extramedullary sites include liver, spleen and lymph nodes.³ Lung parenchymal involvement in multiple myeloma is extremely rare.⁴ The prognosis of patient with pulmonary involvement is poor and is more commonly associated with aggressive terminal phase of myeloma.³ Here we are reporting an interesting case of multiple myeloma masquerading as a pulmonary mass with thoracic extension involving D4-D8 vertebrae and adjacent ribs.

ÖZET

Multiple miyeloma plazma hücrelerinin malign hastalığıdır. Plazma hücreleri vücutta yaygın olarak bulunmakla birlikte, tümör genellikle kemik ve kemik iliği içine yayılır. İlerlemiş vakalarda bile ilik dışına metasazla yayılımı nadirdir. Akciğer parankiminin miyelom hücreleri tarafından gerek plazmositoma olarak gerekse akciğer metastazı olarak tutulumu nadirdir ve hastalığın agresif terminal dönemi ile ilgilidir. Burada ilk olarak akciğer parankim kitlesi olarak belirti veren bir multiple miyeloma olgusunu sunuyoruz.

Anahtar kelimeler: Akciğer, multiple miyelom, ekstremitaller plazmositom.

CASE

A 50 years old male presented to the department of Chest and Tuberculosis at Post Graduate Institute of Medical Sciences, Rohtak, India with a vague complaint of chest pain, cough and mild breathlessness of 6 months duration. The pain was moderate in intensity and constant. He was a non-smoker and non-alcoholic. There was no history of fever or preceding trauma. Chest radiograph showed a peripheral shadow involving right upper and middle zone and extending beyond the thoracic cage. Physical examination of the patient revealed no significant abnormality except mild pallor. Routine investigations revealed Hb-9.6 g/dl, TLC-9800/cm³ with neutrophils-70%, lymphocytes-23%, eosinophils-2%, monocytes-5%. ESR was 60 mm 1st hour by Wintrobe method. The blood urea, serum creatinine, blood sugar, serum uric acid, serum bilirubin, SGPT, serum alkaline phosphatase, serum calcium and phosphorus were within normal limits. Percu-

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taneous fine needle aspiration cytology (FNAC) and biopsy from pulmonary lesion was done which showed cellular infiltrate comprising mature and immature plasma cells, including binucleate and multinucleate forms along with pulmonary paren-

chymal cells (Fig.1 a & b). Diagnosis of plasmacytoma was suggested and patient was advised to undergo computed tomography scan, skeletal survey and electrophoresis to rule out pulmonary dissemination of multiple myeloma.

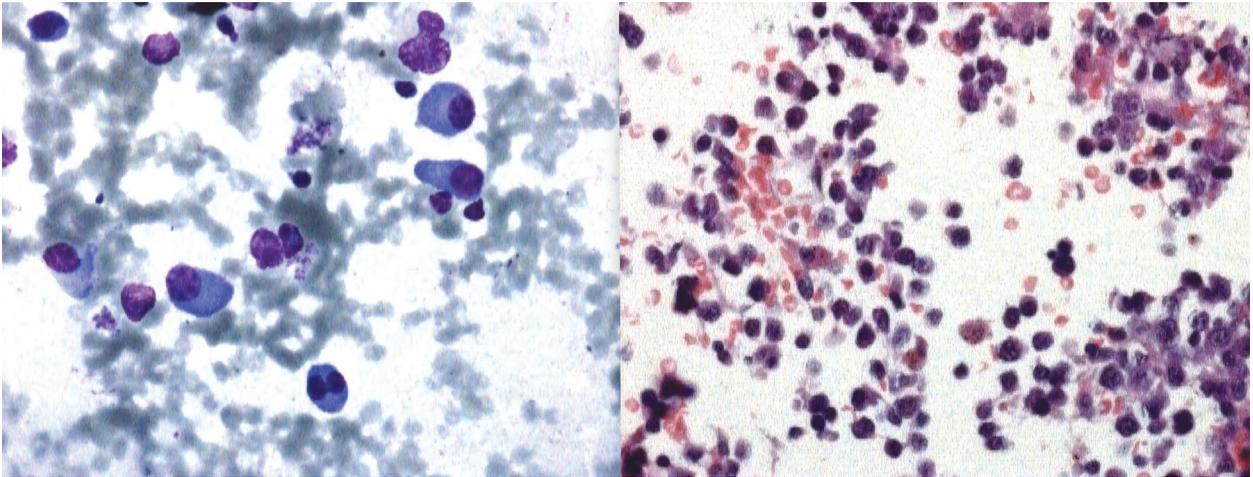


Figure 1 a&b. FNAC and biopsy lung: Photomicrograph revealing plasma cell infiltrate (Giemsa 400X, H & E 400X)

A subsequent skull radiograph revealed multiple punched out lytic lesions (Fig. 2) Contrast tomography (CT) examination of chest revealed large peripheral lung mass with smooth margins in lateral segment of right middle lobe showing homogenous

contrast enhancement. The mass was extending in the costovertebral space of D-4 to D-8 vertebrae alongwith erosion of the adjacent vertebra and ribs (Fig. 3).

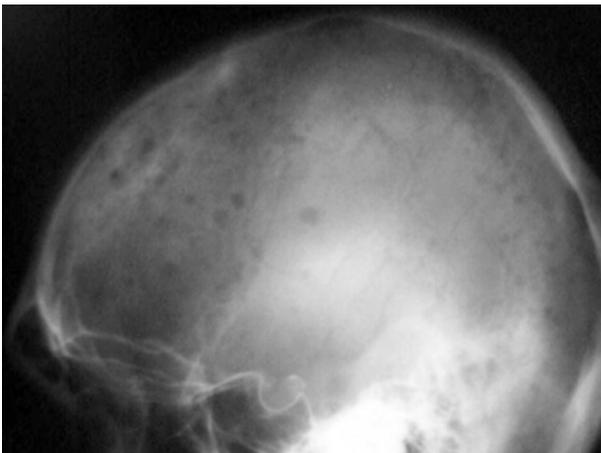


Figure 2. X-ray skull: showing multiple punched out lytic lesions

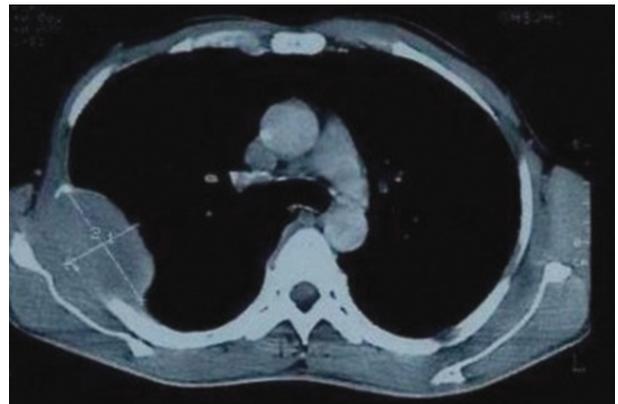


Figure 3. CT scan showing mass in lung extending in thoracic cage and lytic lesion in D-4 to D-8 Vertebra

Bone marrow aspiration revealed about 10% plasma cells. Serum protein electrophoresis revealed normal total serum proteins (7.8gm/dl), mildly raised gamma globulin -1.84g/dl, (normal 0.6-1.6g/dl) and normal serum albumin, alpha 1, alpha 2 and beta globulin. Bence Jones proteinuria was absent. Immunofixation showed A/G -1.42, serum IgG -17.1g/dl (normal 6.94-16.2 g/dl), serum IgA-2.38g/l, serum IgM -0.82 g/l, serum free Kappa (light chain) 2.50mg/dl (normal 0.33-1.94 mg/dl), serum free lambda (light chain) 1.03mg/dl, serum free kappa/lambda ratio-2.42 (normal 0.26-1.65). It showed monoclonal band in serum protein electrophoresis lane, corresponding to monoclonal band seen in IgG and Kappa lanes suggestive of IgG kappa monoclonal gammopathy. A final diagnosis of multiple myeloma with dissemination in pulmonary parenchyma and adjoining soft tissues was made.

DISCUSSION

Multiple myeloma is a haematological malignancy characterised by malignant clonal proliferation of plasma cells in the bone marrow. It is associated with serum monoclonal protein, skeletal destruction with osteolytic lesions, pathological fractures, bone pains, hypercalcemia, renal failure, and anemia. The disease spans a spectrum from localised, smoldering or indolent to aggressive, disseminated forms with plasma cell infiltration of various organs, plasma cell leukemia, and disorders due to deposition of abnormal immunoglobulin chains in tissues. Generalised bone marrow involvement in multiple myeloma is typically present.⁵ Rarely in advanced multiple myeloma, metastatic deposits outside the bone marrow (extramedullary) are seen.¹ Myeloma cells found at extramedullary site may either be due to extramedullary plasmacytoma (EMP) or due to extramedullary dissemination of multiple myeloma.⁶ EMP is uncommon and is characterised by discrete solitary masses of neoplastic monoclonal plasma cells outside bone marrow.² Most common sites for solitary extramedullary plasmacytoma include mainly upper respiratory tract such as nasal cavities, paranasal sinuses and nasopharynx without the involvement of bone marrow.⁵ Extramedullary dissemination of multiple myeloma is also uncommon and reported sites include spleen, liver, lymph nodes, kidney, thyroid gland, adrenals, ovary, testes,

lung, pleura, pericardium, intestinal tract and skin.⁶ Multiple myeloma masquerading as pulmonary nodule is extremely rare. Pulmonary involvement in myeloma is so rare that there is no mention of its occurrence in several large series. Kintzer et al found that 46% of patients in a series of 958 cases had thoracic involvement by myeloma. Most of them showed bone involvement or pulmonary infiltrate secondary to an infectious process. Only 11 patients developed extramedullary plasmacytoma in the thorax and four patients had pulmonary infiltrate suggestive of myeloma cell infiltrate (with only one proven case).⁷ In another study 19 (4.4%) out of 432 patients of multiple myeloma were identified as having extramedullary disease, common sites being lymph node, pleura and soft tissues with only 3 cases (6.2%) occurring within lung parenchyma.² Pulmonary involvement seems to be more commonly associated with aggressive terminal phase of myeloma.³ We report a case in which pulmonary parenchymal lesion was the initial presentation of the disease and the diagnosis of multiple myeloma was confirmed subsequently on investigations.

In diagnostic criteria of multiple myeloma, major criterias include plasmacytosis on tissue biopsy, bone marrow plasmacytosis > 30% plasma cells, monoclonal globulin spike on serum electrophoresis (> 3g/dl for IgG, 2 g/dl for IgA) or on urine electrophoresis (>1g/24hr of kappa or lambda light chain) while minor criterias include bone marrow plasmacytosis of 10-30% plasma cells, monoclonal globulin spike less than the level defined above, lytic bone lesions and residual normal IgM<0.05g/dl, IgA<0.1g/dl, IgG<0.6g/dl. The diagnosis of multiple myeloma requires a minimum of two major or one major and one minor criteria each or three minor criteria (always including 1 and 2).⁸ In our case, the major criterias included extramedullary pulmonary parenchymal plasmacytoma on tissue biopsy and percutaneous FNAC of lung; and in minor criteria patient's bone marrow aspiration showed approximately 10% plasma cells, multiple lytic lesions on skull radiography and abnormal monoclonal globulin spike showing mild increase in serum IgG and serum kappa levels. These findings were suggestive of the diagnosis of multiple myeloma.

The pulmonary involvement in multiple myeloma needs to be differentiated from solitary extramedullary pulmonary plasmacytoma, the treatment

and prognosis of the two conditions is vastly different. The diagnostic criterias for solitary extramedullary pulmonary plasmacytoma are monoclonal plasma cell histology on tissue biopsy, bone marrow plasma cell infiltration not exceeding 5% of all nucleated cells, absence of osteolytic bone lesions or other tissue involvement, absence of hypercalcemia or renal failure, low serum M protein concentration, if present.⁹

The most typical thoracic manifestations of multiple myeloma are bony involvement of thoracic cage or pulmonary infiltrate secondary to infection. Other described manifestations of myeloma in the lungs include multiple nodular lesions, diffuse reticulonodular pattern, pulmonary calcification or amyloidosis.²

Multiple myeloma masquerading as pulmonary mass at its first presentation prompted us to put forward this case report.

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