

Case Report

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A Case of Multiple Myeloma with Lung Involvement

Kaushik Saha[✉], Niranjan Kr. Sit, Arnab Maji, Debraj Jash

Pulmonary Medicine Department, N.R.S. Medical College, Kolkata, West Bengal, India

Abstract

Plasmacytoma are extramedullary accumulations of plasma cells. Most extramedullary plasmacytomas are associated with the upper respiratory tract. The lung is rarely involved. We report a rare case of lung plasmacytoma with multiple myeloma. The patient is a 60-year-old male who presented with chest pain and a lung mass visualized on CT scan. A preliminary diagnosis of occult lung cancer with widespread skeletal metastasis was made. The diagnosis of lung plasmacytoma with multiple myeloma was made after extensive investigations.

Keywords: Lung, Multiple myeloma, Plasmacytoma

Introduction

Multiple myeloma represents a malignant proliferation of plasma cells derived from a single clone. The tumor, its products, and the host response to it result in a number of organ dysfunctions and symptoms, including bone pain or fracture, renal failure, susceptibility to infection, anemia, hypercalcemia and occasionally clotting abnormalities, neurologic symptoms and manifestations of hyperviscosity. Primarily it is a disease of bone marrow but extramedullary plasmacytomas form a small percentage of plasma cell tumors, the majority of which occur in the head and neck. However, the occurrence of extramedullary disease

is very uncommon in multiple myeloma.¹ Here we report a case of multiple myeloma with lung plasmacytoma in a 60-year-old male smoker.

Case Report

A 60-year-old male presented with severe persistent progressive chest pain in both the right anterior and posterior chest wall for a duration of five months and pain in his neck that radiated towards his left upper arm for a period of two months. He had lost 10 kg of body weight in the previous one month. Chest pain was not relieved by medication and worsened at night. Neck pain was worse during movement. He had two episodes of high grade fever with

✉Corresponding Author:

Kaushik Saha, MD
Pulmonary Medicine
Department, Burdwan. Medical
College, Burdwan, West
Bengal, India
Mobile: +919433383080
Email:
doctorkaushiksaha@gmail.com



chills and rigor in the previous month. Physical examination revealed moderate pallor. Respiratory system examination revealed bilateral vesicular breath sounds with prolonged expiration. No tenderness was noted in the chest wall or the axial skeleton. Routine blood investigation showed the following results: hemoglobin (8 gm/dl), total leukocyte count of $8500/\text{mm}^3$ with neutrophils (65%) and lymphocytes (28%). The patient's ESR was 84 mm in the first hour and he had a platelet count of $255,000/\text{mm}^3$. ELISA for HIV 1 and 2 antigens was negative. Chest X-ray, PA view, revealed a lytic lesion over the right sixth rib laterally and a round opacity in the left side medially just above the aortic knuckle, continuous with the superior mediastinum (Figure 1). Whole abdominal ultrasonography revealed no abnormality. CT scan of the thorax revealed a left upper lobe mass with destruction of the transverse process and body of the T7 vertebra and an osteolytic lesion in posterior portion of the right sixth rib (Figures 2A and 2B). CT-guided FNAC from the lung mass revealed the presence of atypical plasma cells with pulmonary parenchymal cells (Figure 3A). Bone marrow aspiration was performed which was remarkable for decreased cellularity, a myeloid-erythroid ratio 3:1, depressed erythropoiesis, depressed leucopoiesis, normal megakaryocytes and 16% plasma cells of all nucleated cells (Figure 3B).



Figure 1. Chest X-ray (PA view) showing a lytic lesion over the lateral portion of the right sixth rib and a round opacity in the left side medially just above the aortic knuckle, continuous with the superior mediastinum.

Serum protein electrophoresis was performed which revealed the following: total protein (9.4 g/dl), albumin (3.37 g/dl), beta globulin (0.65 g/dl), gamma globulin (4.43g/dl), and M-spike (3.56 g/dl). Serum immunoglobulin determination revealed markedly elevated IgG (3200 g/dl). A diagnosis of multiple myeloma with lung plasmacytoma was established. Urinary examination of Bence Jones' proteins was negative in our patient. The patient was placed on oral melphalan (10 mg/day) along with prednisolone (40 mg/day) for four consecutive days, which was then repeated after two weeks. For management purposes we referred the patient to the Oncology Department. Currently, he is on the melphalan and prednisolone regimen.

Discussion

In multiple myeloma the bone marrow is infiltrated with aggregates of abnormal plasma cells that lead to multifocal lytic bone lesions. Extramedullary plasmacytoma is a monoclonal proliferation of plasma cells in soft tissues or an organ. The relationship between multiple myeloma, solitary plasmacytoma of the bone, and extramedullary plasmacytoma is not well understood. For some authors these three entities represent different aspects of the same disease spectrum. Others regard solitary plasmacytoma of the bone as a rare manifestation of multiple

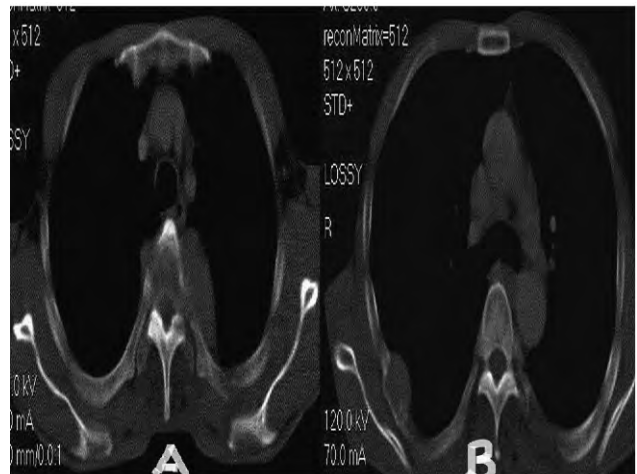


Figure 2. Computed tomographic (CT) scan of the thorax showing a left upper lobe mass with destruction of the transverse process and body of the T7 vertebra (A). An osteolytic lesion seen in the posterior portion of the right sixth rib (B).

Table 1. Comparison of pulmonary manifestations in multiple myeloma as described in various case reports.

	Weber et al. ¹⁰	Shin et al. ¹³	Ramnik et al. ⁷	Sullivan et al. ⁶	Koss et al. ⁹	Present case
Cases (n)	1	2	1	1	5	1
Age (yrs)/sex	52/F	71/M 68/F	60/M	51/M	50–79 4 M/1 F	60/M Left upper lobe mass with osteolytic lesion in right 6th rib and body of T7 vertebra.
Chest radiology	Pulmonary calcification	Mass multiple nodules	Homogenous opacity in upper + mid zone of right lung and erosion of right 6th rib.	Bilateral hilar lymph node + lung nodule	Hilar mass in 2 cases. Intraparenchymal mass in 3 cases.	

myeloma. Extramedullary plasmacytoma should, however, be regarded differently and the diagnosis restricted to tumors that occur outside the bone marrow, may infiltrate nearby lymph nodes or cause distant metastasis.²

Extramedullary plasmacytoma accounts for about 3% of plasma cell malignancies and approximately 80% of which, in the upper respiratory tract (commonly in oropharynx and paranasal sinuses). 1, 3 In the lower airway, plasmacytoma settles in the tracheobronchial tree, structures of the hilum or, rarely in the lung parenchyma.^{4,5} However association of multiple

myeloma with lung plasmacytoma is extremely rare.^{6,7} Only 5% of patients with extramedullary plasmacytomas have coexistent multiple myeloma.⁸

The most typical thoracic manifestation of multiple myeloma is bony involvement of the thoracic cage. Other manifestations include pneumonia, intra-parenchymal mass lesions, mediastinal lymphadenopathy, interstitial pattern like reticulonodular shadows, and intrapulmonary calcification. Pulmonary manifestations in multiple myeloma as reported by various authors are given in Table 1.^{9,10}

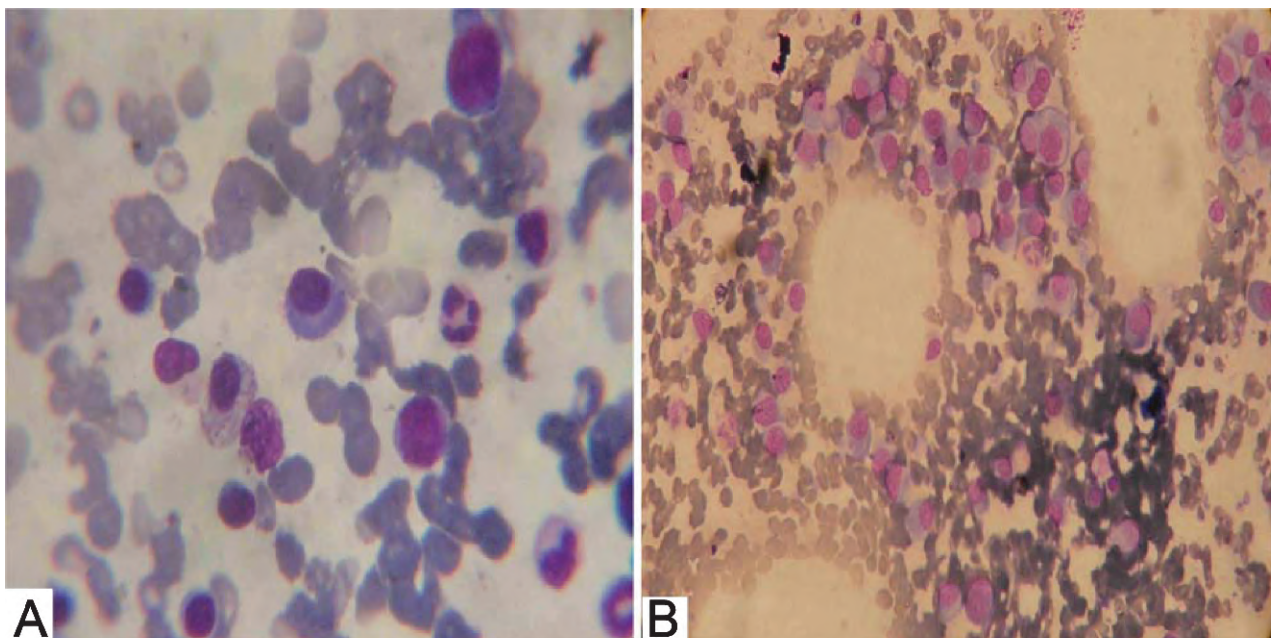


Figure 3. (A) CT-guided FNAC from the lung mass showing the presence of atypical plasma cells with pulmonary parenchymal cells (H&E, 200x). (B) Bone marrow aspirate showing decreased cellularity with plasma cells (H&E, 200x).

We have made the diagnosis of plasmacytoma in our patient based on the following: i) FNAC finding of the lung mass and multiple myeloma, ii) M-protein in the serum, iii) bone marrow plasma cells $\geq 10\%$ (16% in our case), iv) myeloma-related organ or tissue impairment (osteolytic rib lesion in our patient) and v) the presence of monoclonal immunoglobulins in the serum.

After diagnosis of plasmacytoma aggressive search for multiple myeloma is vital as the management is entirely different for both types of plasma cell dyscrasias. Plasmacytomas are treated with radiotherapy, surgery or both.

There are two key stages in the treatment of newly diagnosed multiple myeloma patients. The first is induction therapy, which is the treatment regimen patients receive immediately following diagnosis. The second is maintenance therapy, a long-term therapy that patients often receive after induction therapy, and (usually) a stem cell transplant. Treatment for multiple myeloma is best categorized on the basis of the patient's age and prognostic factors. According to the 2011 NCCN Multiple Myeloma guidelines, therapy for multiple myeloma should consist of: i) a combination of bortezomib/cyclophosphamide/dexamethasone as primary induction therapy for transplant candidates, ii) a combination of bortezomib/dexamethasone (without cyclophosphamide) as primary induction therapy for patients who are not candidates for transplantation and iii) a combination of melphalan/prednisone/lenalidomide as primary induction therapy for nontransplant candidates. Patients who have a relapse after initial disease control may be treated with any of the agents not already utilized. If the multiple myeloma relapse occurs longer than six months after the initial therapy, then the initial regimen can be used again.¹¹

Extramedullary plasmacytoma in a patient with multiple myeloma carries a poor prognosis and treatment, which includes chemotherapy or autologous hematopoietic cell transplantation, is directed towards the underlying disease.¹²

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