

Pulmonary plasmacytoma in multiple myeloma: a rare case of extramedullary spread

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Multiple myeloma (MM) is a systemic disease primarily involving the bone marrow, constituting about 1% of all malignancies and 10% of hematological malignancies [1]. It is a neoplastic proliferation of monoclonal plasma cells that can result in renal impairment, osteolytic lesions, hypercalcemia, bone marrow failure, and the production of serum monoclonal proteins. Although usually restricted to the bone marrow, extraskeletal spread in the form of plasmacytoma, which represent localized extramedullary collections of malignant plasma cells, can occur in a significant number of patients. However, symptomatic pulmonary involvement during the course of MM has rarely been reported [2].

A 55-year-old, nonsmoker, woman presented with complaints of cough, chest pain, breathlessness, hoarseness of voice, and significant weight loss. Cough with breathlessness, which was present for 3 months, aggravated in the last 10 days before presentation, hampering her routine activities. Dull aching pain on the right side of the chest was noted, which increased on inspiration. She had been diagnosed with MM 5 years back and had received five cycles of chemotherapy, with no subsequent follow-up. On general physical examination, she was diagnosed to be hypertensive, with bilateral axillary lymphadenopathy with two to three mobile, nonmatted lymph nodes in the bilateral axilla. Spine tenderness was also noted. Dullness with reduced breath sounds and vocal resonance over the right mammary and infra-axillary areas during the respiratory system examination were noted. Hemogram was essentially normal (hemoglobin 10.3 g/dl, white blood cells 7100 cells/mm³, creatinine 1.2 mg/dl), except for erythrocyte sedimentation rate, which was elevated (140 mm/h), and unexplained thrombocytosis (332 000 cells/mm³).

Indirect laryngoscopy showed nonspecific vocal cord keratosis that was incidental and not associated with the primary disease. Ultrasonogram indicated bilateral

small axillary lymph nodes that were 5–8 mm in size, likely to be reactive and insignificant.

Radiography showed a collapse fracture in T12 vertebrae with osteolytic lesions in the skull. Chest radiograph and computed tomography topogram showed a homogeneous opacity in the right lung (Fig. 1a). Contrast-enhanced computed tomography chest showed a large enhancing homogenous soft tissue density lesion (Fig. 1b) in the right lung (8.6 × 10 × 1 cm). Lytic lesions were also noted in the posterior aspect of bilateral two to four ribs and expansile lytic lesions involving the right fifth and sixth ribs.

Serum protein electrophoresis showed a symmetrical spike in the β region suggestive of paraprotein (Fig. 2a). Bone marrow aspiration and biopsy showed marrow plasmacytosis of 30% with focal and diffuse interstitial infiltration of plasma cells (Fig. 2a–d), suggesting

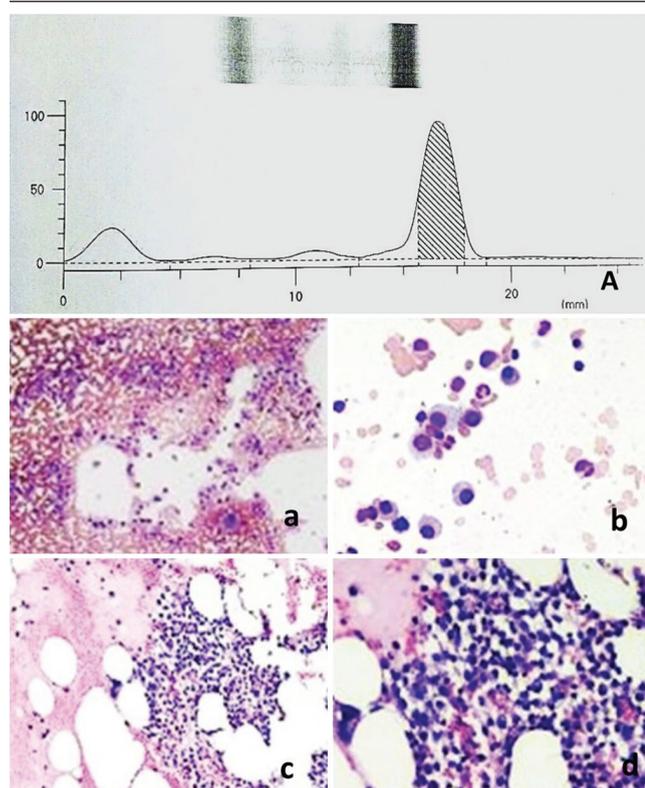
Fig. 1



Computed tomography topogram chest (a) showed a homogeneous opacity in the right lung (black arrow). Contrast enhanced computed tomography chest (b) showed a large enhancing homogenous soft tissue density lesion (Figure 1 B black arrow) in the right lung (8.6 × 10 × 11 cms). Lytic lesions were also noted in the posterior aspect of bilateral 2–4 ribs and expansile lytic lesions involving the right 5th and 6th ribs.

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Fig. 2

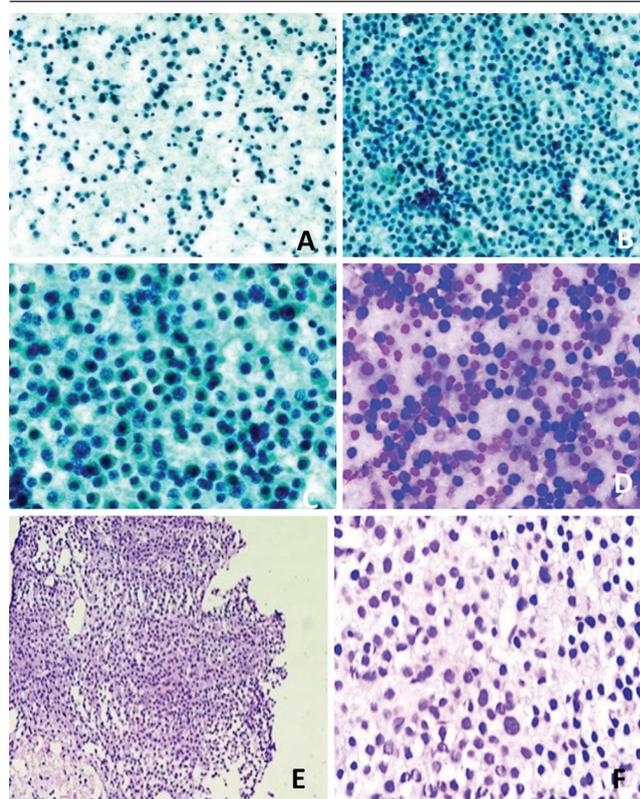


Serum electrophoresis (A) shows a symmetrical spike in the β region suggestive of paraprotein. Bone marrow aspiration and biopsy (a–d) show a plasmacytosis of 30% with focal and diffuse interstitial infiltration of plasma cells — myeloma relapse.

myeloma relapse. In this case, the lesion was peripheral and there was no role for fiber optic bronchoscopy-guided transbronchial lung biopsy.

Computed tomography-guided fine needle aspiration and biopsy of the lesion in the lung was performed. Fine needle aspiration cytology showed plasma cells with eccentrically placed nuclei and some with prominent nucleoli (Fig. 3a–d). Histopathology indicated monoclonal neoplastic plasma cells and plasmablasts with pleomorphism and prominent nucleoli (Fig. 3e and f). The lung lesion was concluded to be pulmonary plasmacytoma as a manifestation of extramedullary spread of myeloma. The differential diagnosis to be ruled out by immunohistochemistry was mucosa associated lymphoid tissue (MALT) lymphoma with plasma cell differentiation, which could be CD20+ and negative for CD138 and CD79a [3]. CD138 and CD79a will be positive in pulmonary plasmacytoma. Pulmonary plasmacytoma can be distinguished from plasma cell granulomas in which there will be an admixture of inflammatory cells such as lymphocytes and macrophages within a fibrous stroma. Immunohistochemistry can be required rarely to differentiate between both the conditions.

Fig. 3



(a–d) Fine needle aspiration cytology of the lung mass. (a) Cellular smears showing plasma cells (Pap stain; $\times 100$). (b) Sheets of plasma cells (Pap stain; $\times 200$). (c) Plasma cells with eccentrically placed nuclei and some showing prominent nucleoli (Pap stain; $\times 400$). (d) Numerous plasmablasts (May grunwald giemsa (MGG) stain; $\times 400$). (e, f) Lung biopsy. (e) Fragment of tumor tissue showing monoclonal neoplastic plasma cells (H&E stain; $\times 40$). (f) Plasmablasts showing pleomorphism and prominent nucleoli (H&E stain; $\times 400$).

Extramedullary plasmacytoma is a monoclonal proliferation of plasma cells in soft tissues or an organ [4]. The sites of extramedullary dissemination reported in the literature are the spleen, liver, lymph node, kidneys, thyroid gland, adrenal gland, ovary, testis, lung, pleura, pericardium, intestinal tract, and skin [5]. They account for about 3% of plasma cell malignancies, ~80% of which occur in the upper respiratory tract, namely, oropharynx and paranasal sinuses. In the lower airway, plasmacytoma settles in the tracheobronchial tree, structures of the hilum, or rarely in the lung parenchyma. The association of MM with lung plasmacytoma is extremely rare – only 5% of patients with extramedullary plasmacytomas have coexistent MM [3]. Second, plasmacytoma presenting as a mass lesion in the lung mimics commonly occurring lung malignancies. The differential diagnoses are metastatic carcinoma and lymphoma [1]. Such a clinicoradiological presentation is rare.

Other case reports of pulmonary manifestations of pulmonary myeloma include homogenous

opacification, multiple pulmonary nodules and mediastinal lymphadenopathy, reticulonodular infiltrates, and intrapulmonary calcification.

Extramedullary plasmacytoma in patients with MM carries a poor prognosis and treatment that includes chemotherapy and autologous stem cell transplantation [6].

Despite advances in the diagnosis of MM, it remains an incurable disease because the disease follows a relapsing course in the majority of patients, irrespective of the treatment regimen or initial response to treatment.

Although pulmonary plasmacytomas are rare, they must be considered in the differential diagnosis of patients presenting with pulmonary manifestations in MM as the management is different for both types of plasma cell dyscrasias, with primary pulmonary plasmacytomas having a better prognosis than pulmonary MM.

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Conflicts of interest

There are no conflicts of interest.

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