Multiple myeloma presenting as thoracic plasmacytoma – Two rare cases

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Abstract
Multiple myeloma (MM) is a clonal proliferation of plasma cells with multiple osteolytic lesions. Multiple myeloma in the thorax is relatively uncommon. Hereby, we present 2 cases of multiple myeloma involving the thorax (1) in a 55 years old nonsmoker, female (2) and in a 45-years-old, smoker, male.

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Introduction
Whereas the most common thoracic disorders associated with myeloma are bone involvement or pulmonary infiltrates secondary to a complicating infectious process, primary thoracic involvement by myeloma occurs in less than 1% of cases [1,2].

Six reported patterns of EMP (extramedullary plasmacytoma) of the thorax include lung mass [3], multiple pulmonary nodules [4], diffuse reticulonodular infiltration by myeloma cells with amyloid deposition [2], lymphadenopathy and mediastinal mass [4,5] pleural effusion with nodular pleural thickening [4] and tracheobronchial infiltration [2].

The diagnosis of extramedullary thoracic plasmacytoma is particularly difficult when there is no thoracic vertebral or rib involvement. The radiologic appearance is nonspecific, with findings on a CT scan or MRI mimicking those of primary or metastatic carcinoma, sarcoma, neuroendocrine or neuroectodermal tumour and lymphoma [3,6].

In a study of 958 cases of multiple myeloma, 6 patients presented with an extramedullary plasmacytoma in the lung [7]. In another study, 19 (4.4%) out of 432 patients of multiple myeloma were identified as having extramedullary disease, common sites being the lymph node, pleura and soft tissues with only 3 cases occurring within the lung parenchyma [8].

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We report 2 cases of multiple myeloma of which in case-1 pulmonary parenchymal lesion was the initial presentation of the disease and the diagnosis of multiple myeloma was confirmed subsequently on investigations and case-2 in which the patient presented with expansile mass in the left lower chest wall.

Case-report

Case-1

A 55-year-old female presented with fever with cough, weakness and shortness of breath for 2 weeks. There was h/o spontaneous fracture clavicle 2 months ago. Past h/o similar respiratory illness-5 years back followed by ATD intake then for 2 months.

Past Investigations revealed Hb-9.9 g/dl, ESR-68 mm/h. Chest X-ray showed homogeneous opacity in the left lung apex (Fig. 1) and FNAC was inconclusive at that time. Repeat FNAC done after a few weeks showed epitheloid cells. Patient was advised for a follow-up but she did not turn up in between as she was apparently well.

Recent Investigations showed Hb-8 g/dl, ESR-125 mm/h, RBC-mildly hypochromic. Chest X-ray 2 weeks before showed the same findings with nondelienated left rib (Fig. 2). Again when she attended the clinic, chest X-ray was repeated which revealed the same findings along with fracture of left clavicle (Fig. 3) giving rise to suspicion of metastasis/plasma cell disorders. Subsequent X-ray of the humerus and skull showed multiple osteolytic lesions (Figs. 4 and 5). FNAC from the nodular swelling of lung showed the presence of plasmacytoid cells-both mononucleate and multinucleate (Fig. 6). Bone marrow aspiration revealed predominance of plasma cell (more than 10%) along with abnormal plasma cells-binucleate and trinucleate forms (Fig. 7). Serum protein electrophoresis revealed the presence of M band (Fig. 8).

Case-2

A 55-year-old male, smoker presented with complaints of left-sided chest pain and breathlessness and loss of appetite for 7 months. The pain was moderate in intensity, constant, and localized primarily to the lower part of the left sided chest wall. The pain increased to some extent on movement.
Physical examination revealed pallor. Investigations showed Hb-8.6 g/dl, ESR-120 mm/h. Chest X-ray revealed an expansile growth arising from left chest wall (Fig. 9). Scanogram confirmed the same. (Fig. 10) CT scan of the mediastinal window shows an expansile swelling of the left rib with destruction of the bony parenchyma (Fig. 11). FNAC from the bony swelling showed plasmacytoid cells both mononucleate and multinucleate (Figs. 12 and 13). Quantitative serum protein estimation showed gamma globulin 2.63 g/dl (30.60%) (Fig. 14). Serum electrophoresis showed M band in the gamma globulin region (Fig. 15). Bone marrow aspiration showed more than 75% of the marrow cells comprising of plasmacytoid cells. Erythropoisis, myelopoisis and megakaryocytes were grossly depressed in keeping with plasma cell dyscrasia. (Fig. 16).
Discussion

Diagnostic criteria of symptomatic multiple myeloma are [9]:

1. Presence of monoclonal plasma cells in the bone marrow more than 10% and or the presence of biopsy proven plasmacytoma.

2. Presence of monoclonal protein in the serum and/or urine.


R–renal insufficiency (serum creatinine > 2 mg%).

A–anaemia (Hb < 10 g% or 2 g below normal level).

B–lytic bone lesion/osteoporosis.
In our study, case-1 shows - Hb-8 g/dl, creatinine-2.3 mg/dl, serum calcium 14.6 g/dl, lytic bone lesion involving skull, humerus with the presence of monoclonal protein in serum. FNAC from pulmonary lesion showed the presence of plasma-cytoid cell. Bone marrow aspiration revealed >10% of myeloma cells involving the marrow (see Fig. 17).

Case-2 presented with anaemia (Hb-8.6 g/dl), expansile swelling arising from the left thoracic wall with destruction of the bony parenchyma, serum electrophoresis revealed the presence of M band while bone marrow aspiration showed >75% of plasma cells.

Findings of both these cases are consistent with the diagnosis of symptomatic multiple myeloma with the involvement of the thorax which is a rare entity.

Association of multiple myeloma with lung plasmacytoma is found to be extremely rare [8,10]. The most typical thoracic manifestations of multiple myeloma are bony involvement of the thoracic cage and the other manifestations are pneumonia, intra-parenchymal mass lesions, lymphadenopathy of the mediastinum, reticulonodular shadows, interstitial pattern and intrapulmonary calcification [11].

Comparative analysis of pulmonary manifestations in multiple myeloma by various authors showed different types of presentation. In a prospective study of Oymak et al. [12], the patients presented with pneumonia in 6 cases, mass lesions in 2 cases. Out of 38 cases, 13 showed lung manifestations. In our case-1, patient presented with pneumonia associated with mass lesion arising from the apex of the left lung involving the left side rib whereas case-2 presented with chest pain and mass lesion involving chest wall. In another study by Shin et al., 2 cases presented with mass and multiple nodule on chest X-ray [2]. In a study by Duggal and co-workers a 60 year old male patient presented with erosion of the right 6th rib [10]. In our case-1, the patient presented with erosion of the left 1st rib.

Multiple myeloma involving the thorax is a rare entity, but it should always be kept in mind whenever an elderly patient presents with thoracic lesions.

Conflict of interest
None declared.

References