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Paraskeletal plasmacytoma presenting as a chest wall mass

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Authors' Contribution:

Sonali Mallik - conception and design, drafting the article, final approval of the version to be published, agreement to be accountable for all aspects of the work

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Abstract

Extramedullary involvement in multiple myeloma is uncommon. It can present as a plasma cell mass in the soft tissue surrounding the bony structures through direct extension or in various other organs via hematogenous spread. Here, we report a case of paraskeletal plasmacytoma that manifested as a chest wall mass in a 60-year-old man.

Key words: Multiple myeloma, plasmacytoma, paraskeletal plasmacytoma, chest wall mass.

Introduction

Multiple myeloma (MM) is a hematological malignancy characterized by clonal proliferation of plasma cells. It constitutes about 1% of all malignant tumors and 10% of all hematological malignancies. The plasma cells primarily proliferate in the bone marrow and produce monoclonal immunoglobulin [1,2]. Infrequently, the clonal plasma cells escape the marrow and continue to grow within soft tissues and various organ systems. The resultant plasma cell tumors that thrive outside the bone marrow microenvironment are known as extramedullary plasmacytomas [3]). They may be present in seven percent of cases with MM at initial diagnosis and may subsequently develop during the disease course in additional 6% cases [4]. We report a case of multiple myeloma with extramedullary plasmacytoma that presented as a chest wall mass.

Case Report

A 60-year-old non-smoker man presented with complaints of right-sided chest pain for two months. The pain was dull aching in nature and localized to the right anterior chest wall without any radiation or referral. Also, he developed a localized swelling over the same region in the right chest wall for one month. The swelling was insidious in onset and gradually progressive in size. He also complained of generalized weakness, body ache, anorexia, and loss of weight. There was no history of fever, breathlessness, and hemoptysis. He was a farmer by occupation. On physical examination, his pulse was

119 beats/min and respiratory rate was 18/min with oxygen saturation of 97% at room air. Pallor was present. There was no peripheral lymphadenopathy. A localized swelling was present over the right mammary region measuring approximately 12cm x 10cm. It was smooth, hard, tender, and fixed to the underlying structure. The margin of the swelling was ill-defined and the overlying skin was normal. Chest auscultation revealed normal vesicular breath sound in both lung fields. Laboratory investigations were as follows: hemoglobin 8.7 gm/dl, total leucocyte counts 9.7 x 103/μL, platelet count 144 x 103/μL, erythrocyte sedimentation rate 140 mm/hour, urea 57 mg/dL, creatinine 2.89 mg/dL and serum lactate dehydrogenase 678 IU/L. Serum total protein level was 10.2 gm/dL with serum albumin and globulin level 2.5 gm/dL and 7.7 gm/dL, respectively. So, the albumin-to-globulin ratio was inverted. Hepatitis B surface antigen, anti-hepatitis C virus antibody, and antibody against human immunodeficiency virus were non-reactive. Routine and microscopic examinations of urine were normal. The chest radiograph showed homogenous opacity in the right mid and lower zones (Figure 1). Contrast-enhanced computed tomography (CECT) of the thorax showed a homogenously enhancing soft tissue density lesion measuring 8.7 cm x 6.1 cm in the right anterior chest wall extending from third to fifth intercostal spaces along with pathological fracture in the anterior aspect of the right fifth rib. Also, there were multiple lytic areas in the sternum, clavicles, scapulae, ribs, and proximal humeri (Figure 2). Ultrasound-guided core needle biopsy was performed from the mass lesion. Histopathology revealed the presence of neoplastic plasma cells containing immature chromatin and atypical mitosis. Few plasma cells were binucleated. The cells were arranged in nests and sheets intervened by blood vessels. These findings were suggestive of plasmacytoma (Figure 3). X-ray of the skull revealed the presence of multiple punched-out lesions. Fluoro-deoxy-glucose positron emission tomography (FDG PET) scan showed diffuse hypermetabolism in the entire visualized bone marrow with extensive lytic lesions in the skull bones, bilateral humeri, clavicles, sternum, multiple ribs, few cervical vertebrae, pelvic bones, sacrum, and bilateral femora. A hypermetabolic enhancing irregular soft tissue mass was seen in the right anterior chest wall involving the third to fifth intercostal spaces and lytic cortical involvement of the fourth and fifth ribs suggesting extramedullary metastatic disease. Serum protein electrophoresis showed an M spike in the gamma region, while immunofixation showed distinct bands in immunoglobulin G (IgG) and kappa light chain

suggesting monoclonal gammopathy. Serum beta 2 microglobulin level was 10800 ng/mL (normal range: 700-1800 ng/mL). Bone marrow biopsy showed the presence of more than 60% plasma cells. The cytogenetic study by fluorescent *in situ* hybridization revealed monosomy 13q(RB1). A diagnosis of multiple myeloma with extramedullary plasmacytoma was made. The patient was in stage 3 according to the international staging system. He was initiated on chemotherapy with a triplet regimen consisting of bortezomib, lenalidomide, and dexamethasone (VRd). Unfortunately, after receiving the first cycle of chemotherapy, the patient succumbed to the disease.

Discussion

The development of extramedullary disease (EMD) in MM can occur either due to direct growth from bony lesions or hematogenous dissemination of clonal plasma cells. Paraskeletal plasmacytomas are formed in the surrounding soft tissues of the skeletal lesions due to disruption of cortical bone. In the case of hematogenous dissemination of tumor cells, the plasmacytomas grow in organs or soft tissue without any contact with bones. (3) However, some authors suggested that paraskeletal plasmacytomas should be considered a distinct form of MM rather than a subtype of EMD. The rationale for this distinction is that plasma cells in paraskeletal plasmacytomas may belong to the same "biological compartment" as the bone and have similar morphological phenotype; whereas plasma cells that metastasize to a distant site carry different biological characteristics with immature plasmablastic morphology. On the other hand, solitary plasmacytoma is a distinct entity that presents as a soft tissue mass with no or minimal bone marrow plasmacytosis. So, it should not be considered as EMD [5,6].

Extramedullary involvement can be present at initial diagnosis as well as can happen during relapse of MM. Paraskeletal plasmacytomas are more common than EMD. The incidence of PS plasmacytomas ranges from 7% to 34.4% at diagnosis and it remains almost similar at relapse. But the rate of EMD is reported to be 1.7% to 4.5% at diagnosis and it doubles at relapse of the disease. The paraskeletal plasmacytomas are commonly seen in relation to the skull, vertebrae, ribs, sternum, and pelvis; whereas EMD is seen involving subcutaneous tissue, skin, liver, breast, kidney, pleura, lymph nodes, and central nervous system [7].

Plasmacytomas may be present as palpable masses. Imaging techniques are required for the detection and characterization of lesions. Magnetic resonance imaging can assess the extent of soft-tissue involvement. It can be useful in differentiating paraskeletal plasmacytomas from EMD [8]. FDG-PET is the most useful imaging technique in MM with soft tissue involvement having sensitivity and specificity of 96% and 78% respectively. Thus, for the detection of PS or EMD, whole-body PET/CT remains the imaging technique of choice [9]. A histopathological diagnosis of plasma cell neoplasm is established when the plasma cells show cytologic atypia. However, if the cells exhibit mature cellular features, the possibility of an extranodal marginal B-cell lymphoma with extensive plasmacytic differentiation should be considered. In such instances, further work-up including flow cytometry, protein electrophoresis with immunofixation, and bone marrow evaluation is required [10].

According to the available literature on extramedullary involvement of chest wall in MM, the most common clinical features include chest pain and localized swellings in the chest wall. The CT scan of the thorax usually reveals soft tissue density lesions that may involve the ribs. The confirmatory diagnosis of plasmacytoma is made based on fine needle aspiration, image-guided biopsy or incisional biopsy of the mass [11-14].

The treatment approach for newly diagnosed multiple myeloma is based on eligibility for autologous stem cell transplantation (ASCT) and risk stratification of the patient. Although several chemotherapeutic regimens have shown favorable outcomes, the current standard options are bortezomib-lenalidomide-dexamethasone (VRd) and daratumumab-lenalidomide-dexamethasone (Drd) [15]. In patients with PS involvement not immediately proceeding to ASCT, the treatment of choice may be the VRd regimen or daratumumab-bortezomib-melphalan-prednisone (Dara-VMP) regimen. In the case of EMD, transplant-ineligible patients are managed with VMP or VRd regimen. The addition of daratumumab has been shown to improve the efficacy of the regimens. Local radiation therapy should be administered in bulky plasmacytomas, in the lesions causing compressive myelopathy and persistent local disease after systemic therapy [7].

Soft-tissue involvement in MM is associated with significantly shorter progression-free survival (PFS) and overall survival [4]. In the relapse setting, the development of

plasmacytoma indicates a poorer prognosis [16]. However, the median survival is better in PS plasmacytomas compared to EMD (3.5 *vs* 1.8 years) [17].

Conclusions

Extramedullary plasmacytoma is an uncommon presentation of MM and also a rare cause of chest wall mass. The presence of extramedullary plasmacytoma bears poor outcome. Further PS should be differentiated from EMD because of further prognostic implications.

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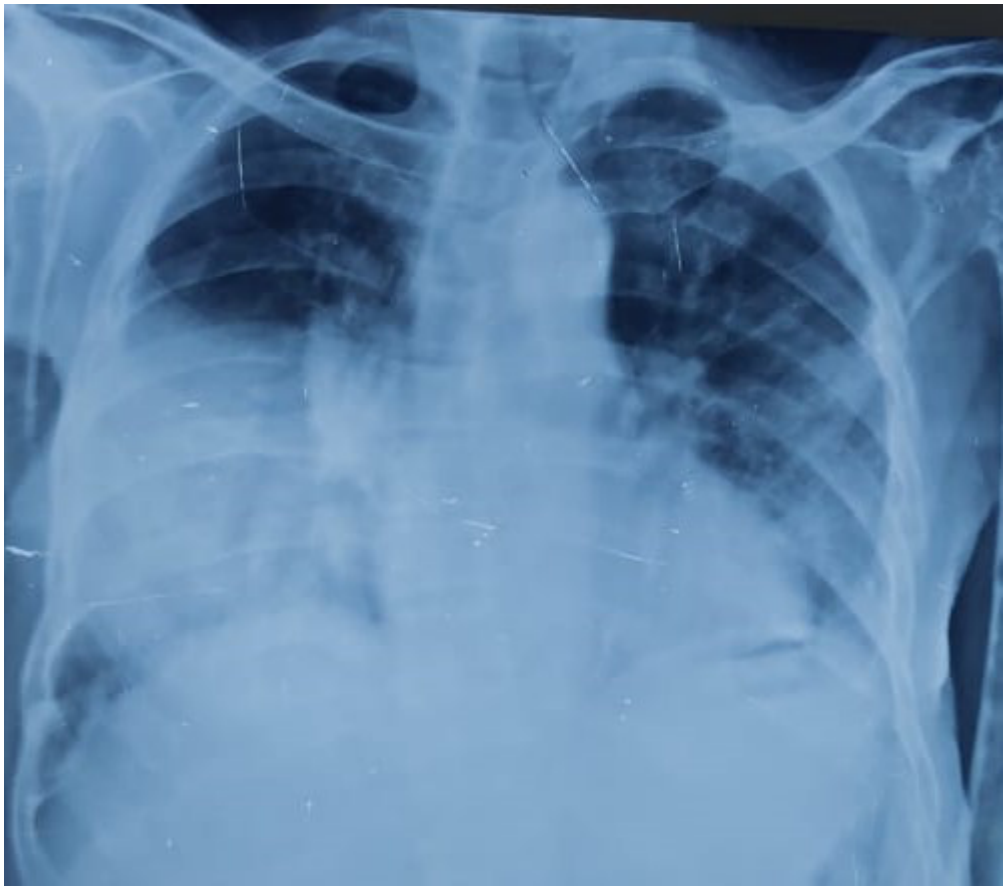

Figure 1. Chest radiograph showing homogenous opacity involving right mid and lower zones.

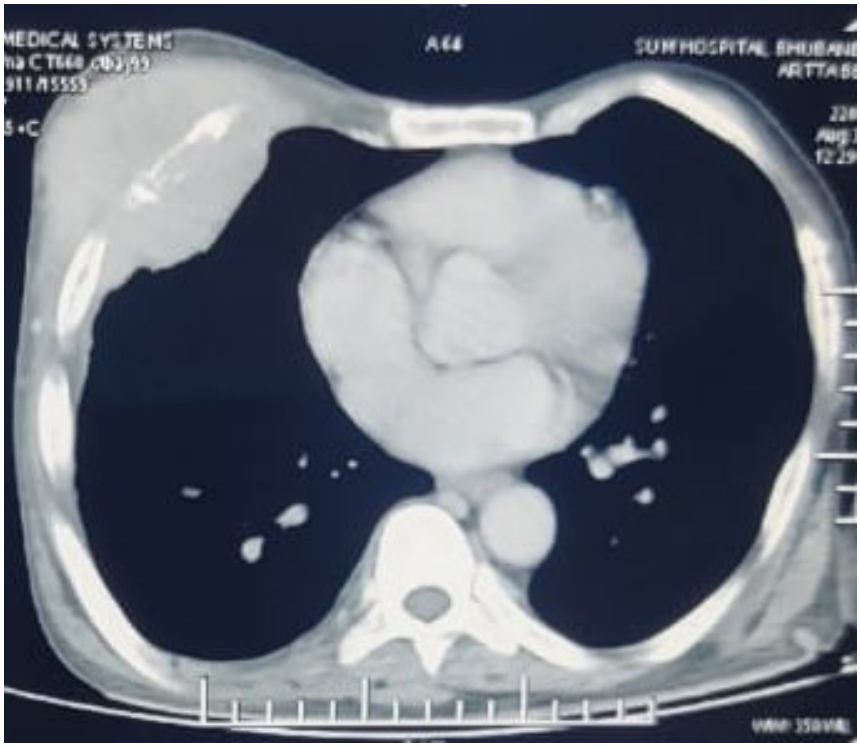

Figure 2. CECT of the thorax (axial image) showing a homogenously enhancing soft tissue density lesion measuring 8.7 cm x 6.1 cm in the right anterior chest wall along with pathological fracture in the anterior aspect of the fifth rib.

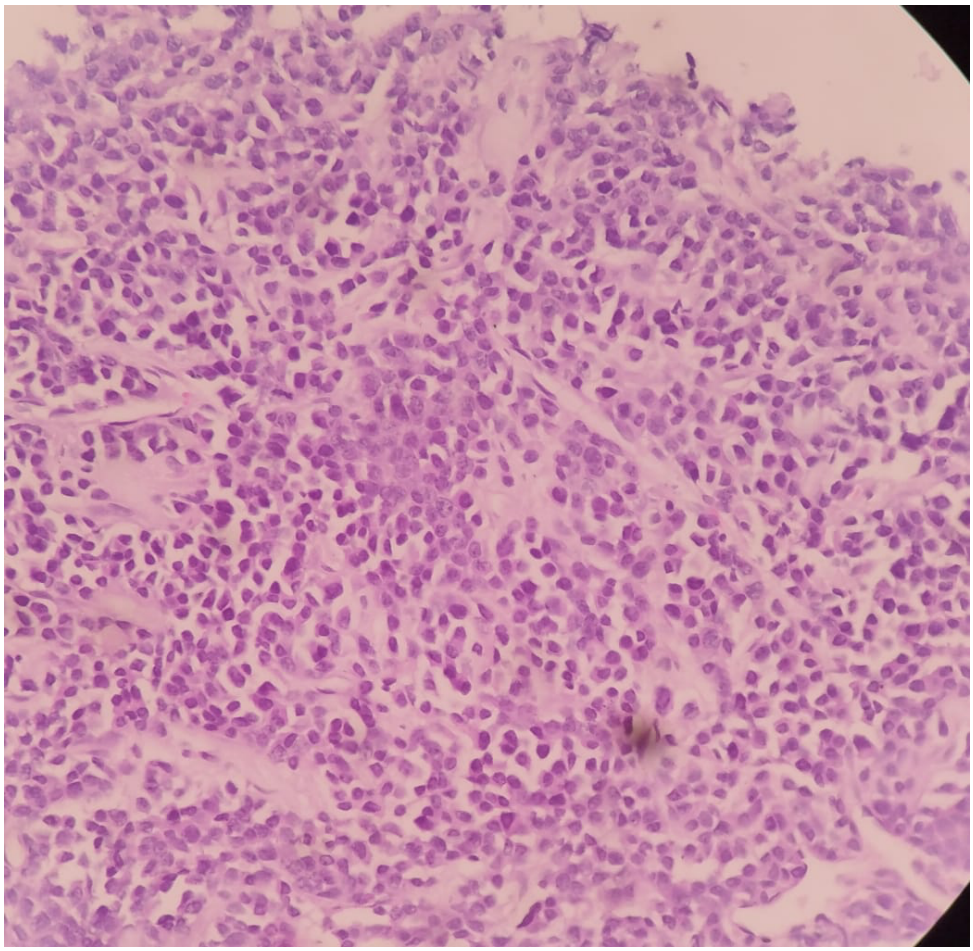

Figure 3. Haematoxylin & eosin stained biopsy specimen from chest wall mass revealing the presence of neoplastic plasma cells having immature chromatin and atypical mitosis. Few plasma cells are binucleated. The cells arranged in nests and sheets intervened by blood vessels (original magnification × 400).