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Multiple Myeloma with Extraosseous Involvement: Imaging Findings

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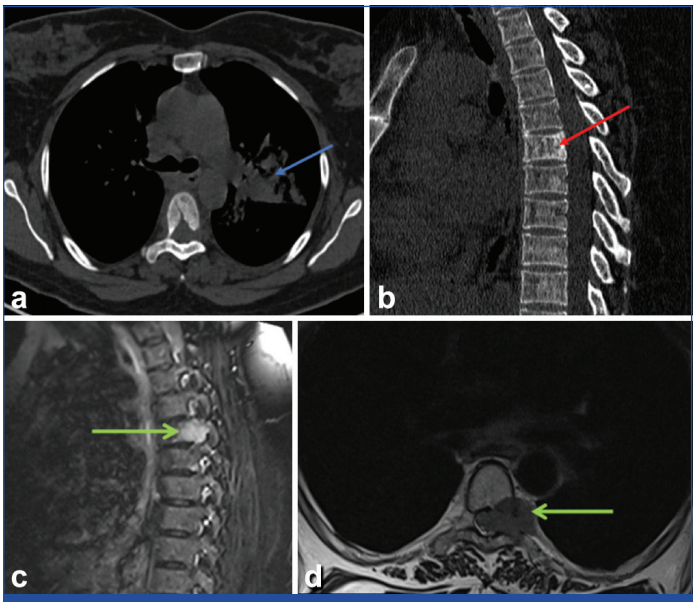
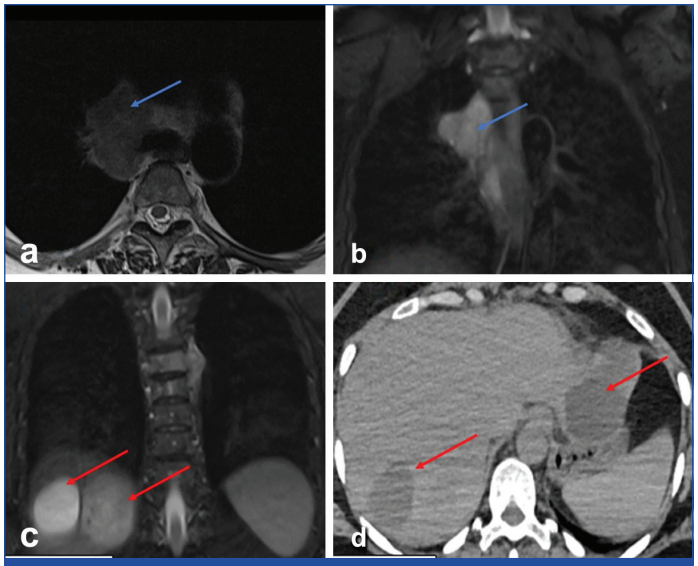
A 50-year-old female patient presented with complaints of backache, occasional on-and-off fever, and wheezing for six months. There was no history of weight loss or other significant complaints. Clinical examination showed tenderness at the mid-dorsal vertebral level. Blood investigations revealed anaemia (Hb: 10 g%), a reversal of the A/G ratio with low albumin levels (3.4 g/dL), and high globulin levels (6.7 g/dL), along with a rise in Lactate dehydrogenase (368 U/L). Alkaline phosphatase was within normal limits at 51 IU/L, and the rest of the investigations were normal. To evaluate the cause of wheezing and pyrexia, a Computed Tomography (CT) chest was performed, which demonstrated infective changes in the left lung field [Table/Fig-1a] and an ill-defined left paravertebral and epidural soft tissue mass at the D5-D6 level with a lytic lesion of the D5 vertebra [Table/Fig-1b] causing mass effect on the cord. Magnetic Resonance Imaging (MRI) of the dorsal spine was conducted for the complaints of backache, which revealed a relatively well-defined lobulated T2 hypointense and STIR (Short Tau Inversion Recovery) hyperintense lesion in the left paravertebral region with involvement of the left pedicle of the D5 vertebral body [Table/Fig-1c,d]. Based on the blood and radiological work-up, multiple myeloma and metastasis were considered as differential diagnosis. The patient then underwent a bone marrow biopsy, which revealed more than 80% plasmablasts. A dedicated comprehensive myeloma panel was conducted, showing positive Ig G and kappa bands, thus confirming the diagnosis. Urine analysis for Bence-Jones proteins also turned out to be positive. All the investigations were consistent with multiple myeloma. The patient was started on injections of bortezomib, thalidomide, dexamethasone, allopurinol, and analgesics in appropriate doses.



[Table/Fig-1]: a) Axial CT shows patchy and confluent consolidation in left upper lobe (blue arrow); b) Sagittal CT dorsal spine shows lytic lesion of D5 vertebra (red arrow); c,d) MRI dorsal spine: sagittal STIR sequences show relatively well defined lobulated STIR hyperintense, T2 hypointense lesion (green arrows) in paravertebral region involving left pedicle of D5 vertebra.

To assess the response to treatment, a follow-up MRI of the whole spine was performed six months after the initial diagnosis, showing no significant change in the previously described paravertebral lesion. In addition to the paravertebral lesion, there was a new appearance of two intercommunicating large T2 heterointense conglomerate lesions in the right paratracheal and right paraesophageal regions of the mediastinum [Table/Fig-2a,b] and a few large STIR/T2 heterogeneously hyperintense lesions in both lobes of the liver [Table/Fig-2c,d]. Histopathological examination with samples from the liver and paravertebral lesion showed atypical cellular infiltrate with plasmacytoid features, confirming the diagnosis of extraosseous myelomatous deposits. As of now, the patient has completed 16 cycles of chemotherapy and has not shown any significant improvement.



[Table/Fig-2]: a) Axial T2 MRI and b) Coronal STIR MRI shows intercommunicating large T2 isointense and STIR hyperintense conglomerate lesions (blue arrows) in right paratracheal and right paraoesophageal regions of the mediastinum; c) Coronal STIR shows few large heterogeneously hyperintense lesions (red arrows) in both lobes of the liver; d) Axial plain CT shows few well-defined hypodense lesions (red arrows) in both lobes of liver.

Multiple myeloma is a monoclonal gammopathy characterised by the proliferation of plasma cells, typically in the bone marrow. It is defined by more than 10% of clonal plasma cells in the bone marrow or biopsy-proven extramedullary plasmacytoma and by evidence of end-organ damage, including bone lesions and renal insufficiency. It is a common malignancy in patients over the age of 40 years. Patients most commonly present with bone pain, anaemia, renal failure, pathological fractures, and recurrent infections. Diagnostic features include anaemia, hypercalcaemia, renal insufficiency, one or more osteolytic lesions on skeletal radiography [1]. Other laboratory findings include Bence Jones proteinuria, reversal of the albumin: globulin ratio, and decreased or normal alkaline phosphatase, as seen in present case. Bone marrow biopsy is the confirmatory tool.

Extraosseous myeloma refers to any manifestation of multiple myeloma where there is plasma cell proliferation outside the skeleton. It is more common in younger patients diagnosed with multiple myeloma. The incidence of clinical and radiological extraosseous myeloma is approximately 10-16%. Although multiple autopsy series have noted an incidence of 63% [2]. It has been shown that extraosseous manifestations are more common following autologous or allogeneic stem cell transplantation. The hypothesis is that extraosseous sites serve as reserve sites for individuals who do not respond well to stem cell transplantation, resulting in a higher rate of extraosseous recurrence [1]. Evaluation with sensitive modalities like MRI/Positron Emission Tomography (PET)-CT is recommended in patients diagnosed with multiple myeloma to evaluate for extraosseous deposits. Extraosseous myeloma occurs as soft tissue homogeneous density masses with no evidence of necrosis or calcification on CT. On MRI, they appear as T2 hypointense lesions and show F-18 fluorodeoxyglucose (FDG) uptake on FDG-PET [2]. The most common sites of extraosseous multiple myeloma are reticuloendothelial system - lymph nodes (5-23%), liver (28-30%), spleen (30-45%), pleura (3-6%), kidney (10-30%), pancreas (4-17%), leptomeningeal involvement (less than 1%) [1-3]. Myeloma involvement of lymph nodes is typically seen as enlarged discrete homogeneous nodal masses that mimic lymphoma [4]. Hepatic involvement may be diffuse, unifocal, or multifocal. Diffuse involvement can be due to innumerable lesions and is more common. The multifocal pattern of involvement usually presents with hepatomegaly with innumerable small low-attenuating lesions, which is the appearance most commonly seen. On Ultrasound (USG) imaging, the focal pattern of involvement shows hypoechoic or target/bullseye appearance. On CT, they show low attenuation without calcification or significant contrast enhancement. On MRI, focal lesions may be hyperintense or hypointense on T1-weighted images and hyperintense on T2-weighted images with minimal gadolinium enhancement [1].

In present case, liver involvement was multifocal in nature. Extraosseous involvement of the liver is more common in IgA myeloma than in other kinds of myeloma, as described in a study conducted by Oshima K et al., although the cause is unknown [5]. But in present case, IgM kappa chain was confirmed by immunohistochemistry with liver involvement. Heckmann M et al., study showed a similar case of pericostal and paravertebral tumour masses besides hepatic, mediastinal, and pericardial tumours. Immunohistochemistry from one of these lesions showed myelomatous origin with IgA expression [6]. In present case, although there were hepatic and mediastinal tumours, there was no pericardial involvement, and present case involved IgM kappa chain. Panda S et al., study showed two cases of mediastinal masses that initially presented with mediastinal widening on chest X-ray. Further biopsy of the mediastinal masses turned out to be positive for kappa chains [7]. Present case also had mediastinal masses, similar to the cases described by Panda S et al., [7].

Extraosseous multiple myeloma is a rare condition that signifies poor survival. It is more prevalent among young individuals and patients who have undergone stem cell transplantation. In a patient with a history of multiple myeloma presenting with an unknown mass, it should be considered as a differential diagnosis. A confirmatory diagnosis by histopathological examination should be made as the imaging features may resemble those of metastasis.

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