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An Uncommon Case of Sacral Plasmacytoma in a Young Male

ABSTRACT

Solitary bone plasmacytoma is an uncommon entity which is characterized by localized proliferation of monoclonal plasma cells. Primarily, it affects the axial skeleton without the evidence of systemic involvement. Involvement of the extremity is unusual. In cases of atypical presentation at a younger age, this entity is mostly suspected radiologically and subsequently confirmed by pathological and biochemical evaluation.

Key words: Peripheral neuropathy, Plasmacytoma, Sacral tumor

INTRODUCTION

The monoclonal gammopathies are a group of disorders associated with monoclonal proliferation of plasma cells.[1] It is classified as multiple myeloma and plasmacytoma. When the plasmacytoma occurs only in bone, it is known as solitary bone plasmacytoma and when involving the soft tissue, it is named as extramedullary plasmacytoma; however, there should be absence of systemic involvement attributing to myeloma in both the lesions. Plasmacytoma arises from plasma cells of bone marrow, whereas extramedullary variety arises from those in the mucosal surfaces.[2] Solitary plasmacytoma is characterized by a localized accumulation of neoplastic monoclonal plasma cells without evidence of a systemic plasma cell disorder.[3] Here, we will be presenting a case of plasmacytoma with unusual clinical presentation.

CASE DETAILS

A 27-year-old male, had initially complained of pain in bilateral calf (left > right) with associated low back ache, since 3 months. The pain was insidious in onset and gradually worsening in nature with no obvious exacerbating or relieving factors. No history of drug allergy or smoking or alcohol consumption. Later, the patient developed sensory symptoms in the form of giving way of the slippers while walking and also he could not get up from squatting position without support. No significant history of any medical illness in the family. Except for reduced power in both ankle joints, detailed neurological examination was grossly normal. Color Doppler flow imaging of bilateral lower limb showing chronic DVT changes in bilateral ATV and PTV, for which the patient was started on anticoagulant. The patient was hemodynamically stable. Blood picture revealed polycythemia with other two cell lines normal. The rest of the routine blood investigation was normal.

X-ray of the pelvis AP view [Figure 1] showed ill-defined lytic lesion with thick surrounding irregular sclerotic rim with narrow zone of transition in the left ala of sacrum. The left sacroiliac (SI) joint appears uninvolved. No evidence of periosteal reaction or overlying soft-tissue component is seen.

The patient was followed up with contrast-enhanced MRI in 1T MRI scanner (Siemens) of the whole spine and pelvis which showed a lobulated heterogeneously enhancing [Figure 2a] altered signal intensity bony lesion with narrow zone of transition involving the left ala of sacrum and left side of the S2 and S3. It was appearing heterogeneously iso- to hypointense on T1WI [Figure 2b] and heterogeneously hyperintense on T2WI/STIR images [Figure 2c] without any blooming foci. The lesion is completely encasing the left S1 nerve root without any change in signal intensity. No intraspinal extension is seen. Bilateral SI joints are uninvolved and normal is seen. No periosteal reaction or soft-tissue component is seen.

Follow-up non-contrast CT scan (NCCT) was done to better document the bony extent and also for guided Trucut biopsy. NCCT of the LS spine and pelvis [Figure 3a] revealed similar finding as that of MRI with erosion of the left S1 neural foramina with the encasement of the left SI nerve root. Few other subcentimetric lytic lesion is noted in the body of LV3 [Figure 3b]. A disc-shaped sclerotic focus is also seen in the body of DV12.

Tru-cut biopsy of the lesion showed sheets of plasma cells which is suggestive of plasmacytoma. FDG PET scan of whole

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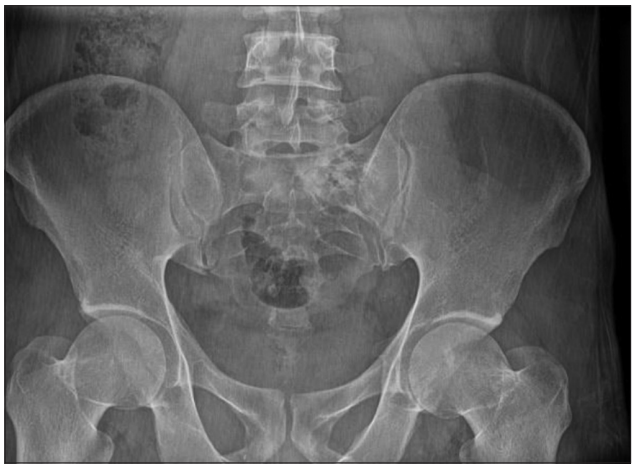

Figure 1: AP radiograph pelvis showing ill-defined lytic lesion with thick surrounding irregular sclerotic rim with narrow zone of transition in the left ala of sacrum

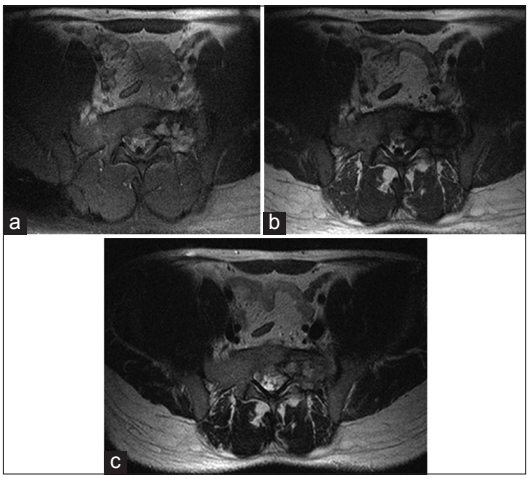

Figure 2: (a-c) CEMRI of the whole spine and pelvis showed a lobulated heterogeneously enhancing bony lesion with narrow zone of transition involving the left ala of sacrum and left side of the S2 and S3. Heterogeneously iso to hypointense on T1WI and heterogeneously hyperintense on T2WI/STIR images

body revealed increased SUV value in all the lesions described in NCCT [Figure 4a and b]. Another lesion is seen in the posterior right 6th rib showing increased SUV value [Figure 4c].

Bone marrow aspirate and biopsy shows plasma cells <9% [Figure 5]. Serum electrophoresis showed monoclonal gammopathy (M spike) in gamma globulin region. EMG/NCV of bilateral lower limb showed generalized (DMN) variant with increased F-wave.

With the clinical background and associated imaging and blood examination biochemical parameters, the patient was diagnosed as a case of solitary sacral plasmacytoma and accordingly treatment instituted. The patient responded to the treatment and presently kept on follow-up as per standard guideline.

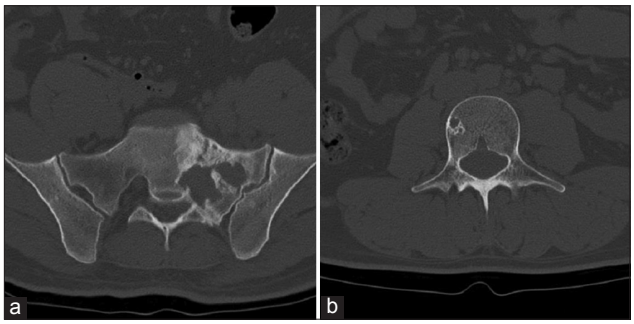

Figure 3: (a and b) NCCT of the LS spine and pelvis revealed lytic lesion and erosion of the left S1 neural foramina with the encasement of the left S1 nerve root. Subcentimetric lytic lesion is also noted in the body of LV3

DISCUSSION

Plasma cells represent the end product of the B-cell maturation. Pathological proliferation of the plasma cell can cause either local tumor (plasmacytoma) or disseminated disease (multiple myeloma).[4] Plasmacytoma is relatively uncommon entity. It is usually unifocal, causing localized destructive lesion of the skeleton in a red marrow area. Most common sites are the vertebral bodies of the thoracolumbar and lumbar regions. The next most common sites are the pelvis, especially the ilium, femur, and humerus. Patients mostly belong to 30–60-year age group.[4] Common symptoms are bone pain and backache.

When a vertebral body is affected, on imaging, there is bone expansion with considerable thinning of the overlying cortex. Sharply defined margin without any surrounding sclerosis reaction is characteristic. The lesion may cause collapse of the vertebral body. Soap bubble appearance may be seen in large lesion in flat bones. Kosaka *et al*.[5] reported that three cases of sacral solitary plasmacytoma showed relatively low signal intensity on T2-weighted images. Our study provided a somewhat similar but interesting finding regarding the disease entity. Our case showed characteristic lytic lesion involving the sacrum with few other smaller lytic lesions could be seen in the lumbar vertebral bodies and in the rib. These lesions showed contrast enhancement in post-Gadolinium T1 fat-saturated images. It is also seen encasing the nerve roots and thereby causing symptoms.

Resemblance to giant cell tumor would be close; however, eccentric location with the involvement of the SI joint would have more characteristic of sacral giant cell tumor.

Other most important differential would be osteolytic metastasis. However, metastasis is more common in older age group and metastasis commonly involves the pedicles.

Chordoma is another relatively uncommon malignant bony lesion, common site of affection being sacrum. It causes lytic bony lesion; however, it is usually midline tumor and mostly seen in the sixth and seventh decade.

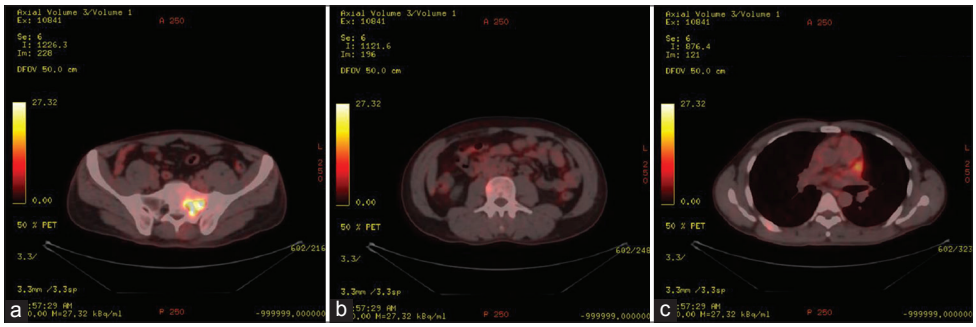

Figure 4: (a-c) FDG PET scan of whole body metabolically active lesion in left sacral ala and multiple lumbar vertebrae. Another lesion is also seen in the posterior right 6th rib showing increased SUV value

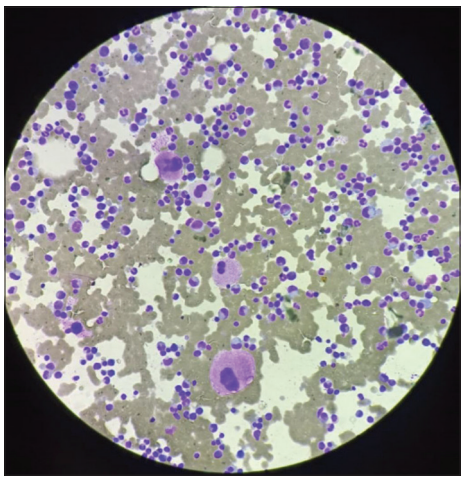

Figure 5: HPE of biopsy from sacral lesion showing plasma cells

CONCLUSION

In conclusion, sacral solitary lytic lesions are commonly encountered in daily practice. Though a wide range of differentials exists, imaging plays a crucial role in narrowing the differential. Thorough knowledge about the imaging findings backed up with clinical data can clinch the final diagnosis.

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