

Case Report

Multiple Myeloma Associated with Extramedullary Plasmacytoma Arising from Ileum and Sacrum Presenting with Sacral Nerve Root Compression: A Rare Case Report

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Abstract: Multiple myeloma associated with plasmacytoma of ilium & sacrum with local extension in the form of extramedullary soft tissue plasmacytoma compressing sciatic nerve roots at initial presentation is rare. We describe a 57 year old male patient who presented with progressive pain and weakness at left posterior thigh and gluteal region proceeding gradually towards proximal aspect. The patient was diagnosed as immunoglobulin G, kappa nonsecretory multiple myeloma with plasmacytoma complicated with sacral nerve root compression at the S2 & S3 levels. The patient was treated with Lenalidomide-Dexamethasone.

Keywords: Multiple myeloma, Extramedullary, Neurological.

INTRODUCTION

The association between a solitary plasmacytoma and peripheral neuropathy is rare, and fewer reports are there in literature of similar cases. Here, we report an unusual case of multiple myeloma that initially presented with progressive pain and mild weakness of the left gluteal region along with posterior aspect of the thigh. The patient was diagnosed as nonsecretory multiple myeloma with plasmacytoma at the left gluteal region

CASE REPORT

A 57-year old man was in good health until he experienced progressive weakness and mild pain over the lower part of buttock and back of the left lower limb for last two months. The symptoms first appeared in the lower part of the posterior thigh proceeding gradually towards the buttock. He had no bowel or bladder complaints. Neurological examination was normal except sign of left sacral nerve root compression. Routine laboratory studies revealed haemoglobin 10.8 g/dl, total WBC count 6200/cu mm, total RBC count 3.9 L/cu mm, serum calcium 11.4 mg/dl. Rouleaux formation was seen on peripheral blood smear. CT scan of lumbo-sacral spine demonstrated a large osteolytic lesion with cortical breakdown in the inferior aspect of the left iliac blade and a small area of the adjacent part of sacrum along with involvement of the articular margin of adjacent left sacroiliac joint with widening of

the joint space. This is associated with a well defined extraosseous soft tissue component arising from it measuring approximately 63 x 52 mm in greatest cross sectional dimension. The soft tissue mass has eroded the lateral boarder of the sacrum at S2 and S3 levels and has invaded the adjacent gluteus maximus, minimus and intermedius muscles. Multiple well defined osteolytic lesions of varying sizes in the lumbo-sacral vertebrae and the pelvic bones was seen (Fig. 1).

Magnetic resonance (MR) imaging demonstrated a large extraosseous soft tissue mass at the left gluteal region arising from inferior part of iliac blade and small part of the adjacent sacrum compressing lateral boarder of the sacrum at S2 and S3 levels and invaded the adjacent gluteous maximus, minimus and intermedius muscle (Fig. 2).

CT guided FNAC obtained from the left iliac lesion and soft tissue mass revealed plasma cell neoplasm compatible with plasmacytoma (Fig.3). Serum protein electrophoresis did not demonstrated M band (Fig. 4).

Bone marrow examination revealed 30% plasma cell (Fig. 5). Serum protein electrophoresis reflex to immunofixation revealed no myeloma band; serum light chains (kappa & lamda) – kappa light chain 230.00 mg/dl (3.30-19.40 mg/dl), lamda light chain –

17.40 mg/dl (5.71-26.30 mg/dl) and kappa lamda ratio-13.218 (0.26-1.65). Electrophoresis of urine protein fractions – albumin/alpha-1/alpha-2/beta/gama/bence jones protein – not detected; total protein 35.6 mg/dl (< 110.9 mg/dl); 24 hour urine volume 3330 ml/24 hrs; Beta 2 microglobulin 3953 ng/ml (1556 – 2286 ng/ml). Based on International Multiple Working Group (IMWG) criteria for diagnosis of monoclonal gammopathies the final diagnosis was immunoglobulin G, kappa nonsecretory multiple myeloma with plasmacytoma complicated with sacral nerve root compression at the S2 & S3 levels.



Fig. 1: CT scan of lumbo-sacral spine shows a well defined extraosseous soft tissue mass arising primarily from lytic area of the inferior part of the left iliac and minimally from adjacent sacral lytic lesion eroding lateral boarder of sacrum at S2 and S3 levels

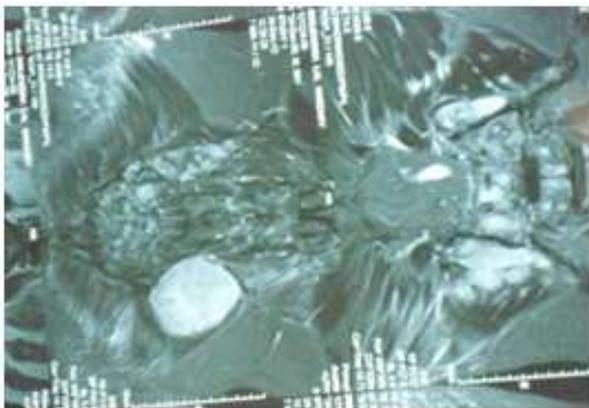


Fig. 2: CT scan of lumbo-sacral spine shows a well defined extraosseous soft tissue mass in the left gluteal region invading the adjacent gluteous muscle group

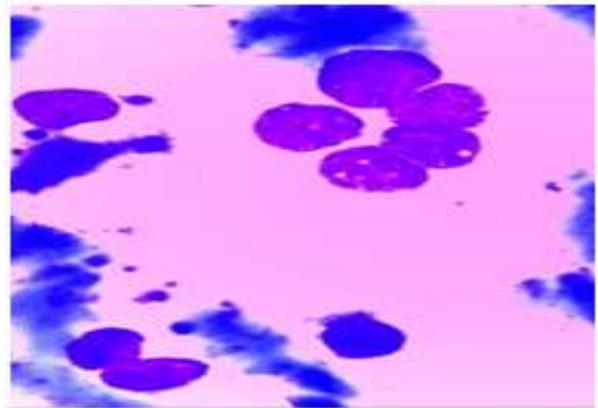


Fig. 3: Histopathology of the FNAC obtained from the mass and iliac lesion shows cluster of plasma cell suggesting plasmacytoma (40 X)

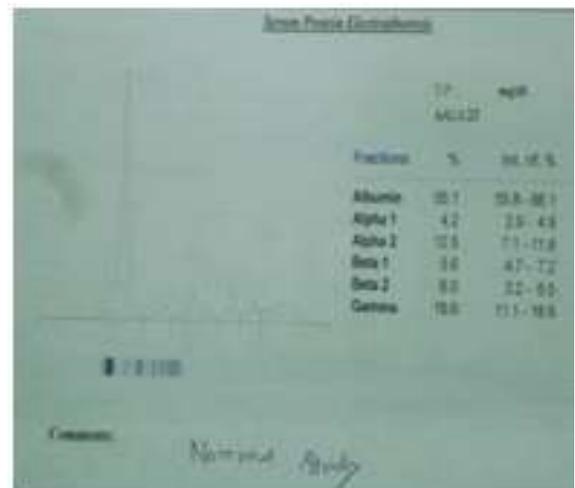


Fig. 4: Serum Protein Electrophoresis shows no M band

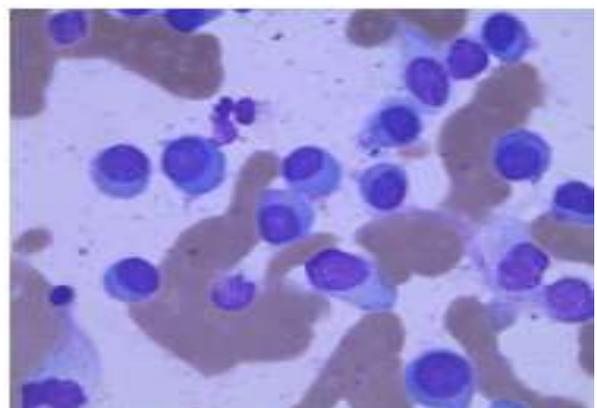


Fig. 5: Histopathology of the bone marrow aspiration shows huge amount of mature and immature plasma cells (40X)

DISCUSSION

Sacral involvement by solitary plasmacytoma of any type is unusual while involvement of ileum is rare. In a large series of 61 cases of solitary plasmacytoma, the involvement of sacrum was observed only in one case [1]. Other recent series

include no examples of solitary sacral lesions [2, 3]. The clinical feature related to our patient is due to compression of sacral nerve roots by the plasmacytoma at the S2 and S3 sacral nerve roots. The association between peripheral neuropathy and multiple myeloma is well documented [4] and occurs in about 3% of patients. However, it is observed in about 50% of patients with solitary plasmacytomas [5]. The peculiarity of development of peripheral neuropathy associated with plasmacytomas is that motor weakness is more prominently manifested than sensory loss [5]. It usually begins distally and spreads proximally in a symmetrical pattern [5]. It is well established that that disease in the sacrum or its related spaces may produce symptoms related to the sacral or sciatic nerves because of mass effect, and that CT scan is essential in evaluation of the sacrum [6] or other pelvic bones. The present case emphasize these points.

To our knowledge plasmacytoma arising from both sacrum and iliac blade compressing sacral nerve roots has not documented in literature. The apparent rarity of the association between polyneuropathy and solitary plasmacytoma arising from sacrum, ileum and corresponding soft tissue is surprising considering the appearance in our case which may be connected with the neurological mode of presentation of the condition. Studies have pointed out that neuropathy of multiple myeloma almost always appears as a complication of the established disease whereas that associated with a solitary lesion presents with the neuropathy [4]. Less frequently it is found in ileum and sacrum.

Our patient presented with a biopsy-proved sacroiliac solitary bone plasmacytoma with extensive involvement of the corresponding soft tissues. However, involvement of adjacent bone and soft tissue sites by direct extension is rare and does not constitute evidence of multiple myeloma. In our case solitary bone plasmacytoma was located in sacrum and ileum that extended into the corresponding gluteal region in the

form of plasmacytoma. CT and MR imaging findings can be very helpful in a patient with plasma cell dyscrasia and should be kept in mind, even though biopsy is usually required. To our knowledge, the imaging findings of such an unusual presentation of plasmacytoma have not been previously described. Usually, a full skeletal survey is not routinely carried out during the investigation of an obscure peripheral neuropathy, and the chances that a small plasmacytoma will be overlooked must be high, if it is not in the area covered by a routine radiological investigation. The diagnosis of this potentially treatable cause of an otherwise progressive neuropathy must, therefore, be considered in all patients with an obscure progressive neuropathy affecting predominantly the lower limbs and in whom all routine investigations are normal. We suggest that all such patients should have a full skeletal survey.

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