Case Report

Monoclonal Gammopathy of Unknown Significance (MGUS) – Solitary Plasmacytoma of sternum

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Abstract

Plasmacytoma is a rare B- lymphocyte neoplastic disorder that usually presents as the generalized disease multiple myeloma. Less than 5% of cases present as a solitary mass of monoclonal plasma cells in the bone or soft tissue. Although solitary osseous Plasmacytoma may arise in any bone, while Sternal involvement is uncommon. We report here a case of solitary plasmacytoma of sternum in a 55 years old male patient without signs and symptoms suggestive of multiple myeloma. Patient was treated with radiation therapy with a dosage of 40 Gy in 20 fractions.

Key words: Monoclonal Gammopathy, Sternal tumors, Multiple Myeloma.

INTRODUCTION

Solitary plasmacytoma of bone is an uncommon form of plasmacytoma and is localized to involved bone [1]. The new World Health Organization (WHO) criteria defines solitary plasmacytoma of bone (SPB) as a localized bone tumor consisting of plasma cells identical to those seen in plasma cell myeloma, which appears as a solitary looking lesion on radiological examination [2]. Plasma cell neoplasms are categorized classically in four groups: multiple myeloma, plasma cell leukemias, solitary plasmacytoma of bone and extramedullary plasmacytoma [3].

A plasma cell neoplasm is a monoclonal proliferation of B - cell that has undergone potentially malignant transformation to become a plasmacytoid cell. These neoplasms can manifest as several different clinical entities, all of which are associated with production of homogeneous immunoglobulins composed of single class of heavy and light chains or fragments of these components [4]. We report here a case of solitary plasmacytoma of bone in a 55 years old pt, involving upper part of sternum without signs and symptoms of multiple myeloma. To best of our knowledge this is the first case to be reported from Pakistan.

CASE REPORT

A 55 years old male patient presented with an enlarging lump over his upper sternum during the previous 05 months. He believed that the mass had enlarged progressively over the last month without any associated complaints like fever, chills, headache, sweating, bone pain, cough or dysphagia. His past history included exploratory laparatomy and gastric repair plus omentoplasty operated for gastric perforation. He had no relevant medical and family history. The patient was afebrile with normal vital signs. Neither liver nor spleen was palpable, no lymphadenopathy were noted. Just inferior to his sternal notch, there was a large rounded lump of 6 x 4.5 cm which was non tender, hard in consistency and fixed to underlying structures.

Initially laboratory values were as follows: Hemoglobin 13.6 g/dl, TLC count 6900 with normal differential count, serum calcium 9.7 mg/dl, serum albumin 3.7 g/dl, urine analysis showed 1+ protein and calcium oxalate crystals, but no glucose, RBCs or WBCs. Electrolytes, renal and liver function tests were unremarkable. Chest x-ray was unremarkable. A metastatic bone survey was negative.

Whole body ^{18}F-FDG PET CT Scan was performed which revealed mildly FDG avid (SUV max: 2.8),

expansile lytic lesion with soft tissue component was noted in the manubrium, measuring 7x4.3x5 cm, abutting pectoral muscles and displacing mediastinal structures posteriorly (however there was no encasement of major vessels) with extension into mediastinal fat. Fat planes with adjacent structures were maintained. No other FDG avid lesion noted in the body surveyed.

Figure-1
(A & B): Showing lytic lesion in sternum

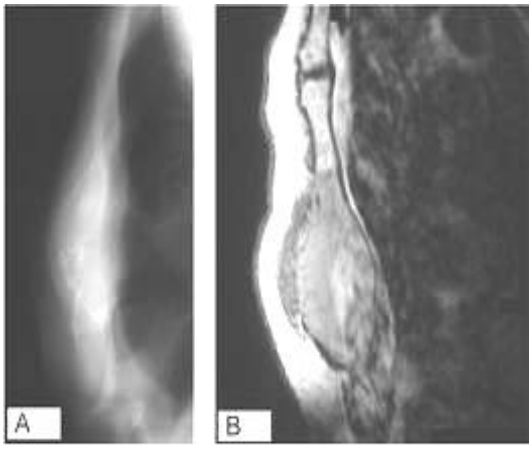


Figure-2
FDG-PET showing lytic lesion in upper part of sternum

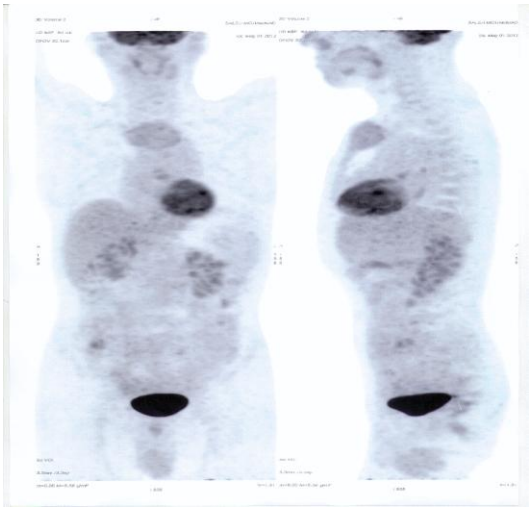


Figure-3
Showing no any metastatic FDG avid lesion

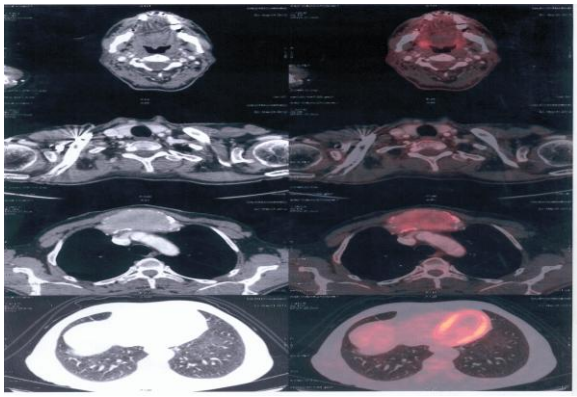
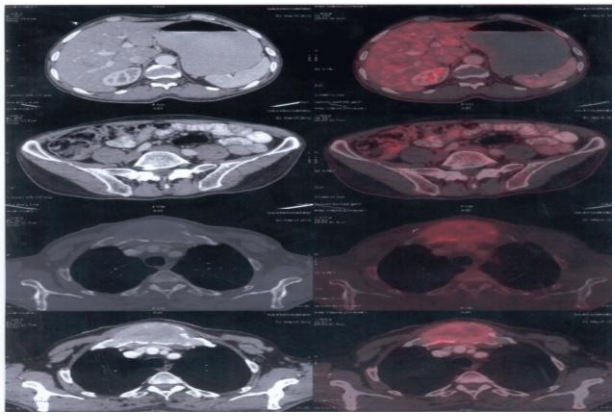


Figure-4
Showing no any metastatic FDG avid lesion



A trucut biopsy from sternal growth was performed, which revealed dense plasma cell infiltration high lightened by CD138 with predominant expression of Lambda light chains, no staining for kappa chains. CK and CD56 were not expressed and the diagnosis was consisting with plasmacytoma.

Bone marrow biopsy showed: Normal marrow cellularity with M: E ratio was 3:1, Erythroid precursors and myeloid series showed normal maturation and morphology, megakaryocytes were adequate; plasma cells were 4% in average.

The serum protein electrophoresis showed an IgG lambda monoclonal protein spike with an initial gamma fraction of 2.86 g/dl. B2- microglobulin was 2.96 mg/l within normal range. Urine test for Bence Jones Protein was negative.

The patient was diagnosed with solitary plasmacytoma of sternum. He had received a course of radiation therapy with a dose of 40 Gy in 20 fractions. He tolerated the radiotherapy without significant toxicity.
The patient has been followed every 2 months over the last 04 months and is stable.

DISCUSSION

Plasmacytoma is a rare neoplastic disorder arising from B- lymphocytes. It usually present as the generalized disease multiple myeloma; however, a few pts (fewer than 5%) with plasma cell malignancies present with either a single bone lesion that is solitary plasmacytoma of bone(SBP), or less commonly, a soft tissue mass of monoclonal plasma cells also known as solitary extramedullary plasmacytoma(SEP) [5]. Solitary bone plasmacytomas often progress to multiple myeloma, where as solitary extramedullary plasmacytoma tend to have high cure rate with localized treatment. Solitary bone plasmacytomas have a male-female ratio of 2:1, with a median age of 55 years and often occurs in the axial skeleton, especially the vertebrae, ribs and pelvis [6, 7]. In our case after histopathologically confirmed the diagnosis of plasmacytoma of bone, we performed FDG-PET combined with whole body CT-scan which confirmed the lesion as solitary. Hughes et al [8], suggested criteria for the diagnosis of solitary plasmacytoma according to the guidelines of working group of the United Kingdom Myeloma Forum (UKMF) which are:

- No M protein in serum/ or urine.
- Single area of bone destruction due to clonal plasma cells, as in our case.
- Bone marrow not consistent with multiple myeloma, plasma cells <5%.
- Normal skeletal survey.
- No anemia, hypercalcemia or renal impairment, as it is the case in our pt.
- No related organ or tissue damage.

Restrepo et al, described that sternal abnormalities can arise from trauma, degenerative and inflammatory conditions and neoplasms. Most neoplasms of sternum are metastases, while primary sternal neoplasms are relatively uncommon: however primary tumors are much more frequently malignant and most are chondrosarcomas [9].
Soutar et al, suggest that radiotherapy is the treatment of choice for solitary plasmacytoma [6]. In patients with solitary plasmacytoma of bone, the most common pattern of progression consists of new bone lesions, rising myeloma protein level and development of marrow plasmacytosis. The median survival of pts with solitary bone plasmacytoma is 7.5 to 11 years [7]. Unfortunately, the majority of pts (>75%) with solitary bone plasmacytomas develop multiple myeloma with a median time to progression of 2 to 4 years [6, 7].
When combined with surgical excision either pre or post radiotherapy the curative rates are higher [10].
After completion of radiotherapy, pts should be monitored for local recurrence and/or new bone lesions as well as progression to multiple myeloma. Serial serum and urine electrophoresis, complete blood count and renal function should be monitored every 4 to 6 months. A skeletal survey should be performed every year or sooner if the M-protein increases. At present, there is a considerable interest in thalidomide in plasma cell dyscrasias but there is no data for its use for solitary plasmacytomas.

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