CASE REPORT

PLASMACYTOMA PRESENTING AS A STERNAL MASS

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ABSTRACT: OBJECTIVE: To review the tumors of the sternum seen in radiological practice and to bring into focus a case of a sternal mass proved on FNAC examination to be a plasmacytoma.

KEYWORDS: Sternum, Solitary Plasmacytoma, Multiple Myeloma, Sternal Tumours.

INTRODUCTION:

Content Proper: History: A 56 year's old female patient presented to the department of orthopedics SVS medical college hospital on 14th March, 2013, with complaints of a mass in front of the chest which was present since 3 years but had started to increase in size since 6 months.

The mass was not associated with pain.

No other constitutional symptoms.

No similar swellings in other sites.

Figure 1

Figure 2

CLINICAL EXAMINATION:

- Globular mass of 10X10cm over the upper 3rd of sternum towards left with Lobulated surface along with Engorged veins present.
- Surroundings are normal.
- No local raise of temperature. No tenderness. Margins well defined and with Variable consistency.
- Skin is pinchable.
- No reducibility/compressibility.
- No regional lymphadenopathy.
- No significantly positive general examination findings.

Subsequently the patient was referred to the radiology department for further evaluation.

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CHEST RADIOGRAPH (AP) LATERAL CHEST RADIOGRAPH:

Figure 3

Figure 4

Finding on Chest Radiographs AP And Lateral: Soft tissue swelling around the upper part of the body of sternum and manubrium sterni. And in the lateral view it is anterior and posterior to the sternum.

CT TOPOGRAM:

Figure 5

Figure 6A & B

Multiple small lytic areas involving predominantly upper part of sternum with associated soft tissue external and internal components. (Fig. 6a & 6b) moth eaten pattern of bone destruction of the sternum.

A large soft tissue mass encasing the sternal body and manubrium.(Fig. 6C)

Figure 6C

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A large soft tissue mass encasing the sternal body and manubrium.

Figure 7

BONE WINDOW:

No periosteal reaction

No matrix calcification

RECONSTRUCTED IMAGES:

Pathological fracture of lower body of sternum

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Ultrasound Screening:

- Revealed a large relatively homogeneous soft tissue echogenic mass around the sternum.
- Superficial bone irregularity of the underlying sternum.
- The mass showed increased vascularity with decreased resistive index.
- The above findings suggest the nature of the lesion to be a malignant lesion.

The differentials for the above Features Include:

- Plasmacytoma.
- Metastasis.
- Lymphoma.
- Chondrosarcoma.

Limitations While Imaging:

- Patient not co-operative for further investigations.
- Hence directly proceeded to fnac exams.
- Complete workup would include a high quality x-ray, ultrasound, cect, (or ccmr), bone scintigraphy, pet scan (To rule out mets).

FNAC REPORTS:

SITE OF FNAC	: FNAC of swelling in sternal region
CLINICAL HISTORY	: Complains of swelling in sternal region since 3 years.
CYTOLOGY REPORT	
Smears examined are hemorrhagic and moderately cellular. Majority of tumor cells are round to oval with good amount of cytoplasm and eccentrically placed nuclei with coarse chromatin.	
DIAG : FEATURES ARE SUGGESTIVE OF PLASMA CELL TUMOR (PLASMACYTOMA).	
Biopsy advised for further histopathological evaluation & confirm the diagnosis.	
REPORTED BY PATHOLOGIST	M I/C SIGN

There by the predominantly soft tissue mass around the sternum turned out to be plasmacytoma of the sternum.

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SUMMARY:

- Most neoplasms of the sternum are metastases.
- Primary tumors are relatively uncommon in this site; however, primary tumors of the sternum are much more frequently malignant than benign.
- Whether it is believed to be primary or secondary, a new mass in the sternum should be considered malignant until proved otherwise.

PRESENT CASE:

Plasmacytoma: Both plasmacytoma and multiple myeloma may occur in the sternum and are most frequently seen during the 6th and 7th decades of life.

The classic radiologic appearance of osseous plasmacytoma, an area of bone destruction with discrete margins-the so-called “punched-out” lesion-may be absent in sternal plasmacytomas.

- CT better depicts subtle lytic lesions, small soft-tissue masses with a multilobulated appearance and lack of intrinsic calcifications, and expansile lesions, which may be similar in appearance to osseous metastases.

Metastases:

- Usually subsequent to direct infiltration from adjacent organs or from hematogenous spread.
- Cancers of the breast, lung, thyroid, kidney, and colon, and hematologic malignancies such as lymphomas, are among the most common sources of sternal metastases.

Lymphoma:

- Primary non-Hodgkin lymphoma of the sternum is much more commonly described than is sternal Hodgkin disease, which is only reported anecdotally.
- Subclassification of sternal lymphomas is difficult, as is that of osseous lymphomas that occur elsewhere.
- At CT, lymphoma appears as a soft-tissue mass with bone destruction and diffuse mild enhancement after intravenous injection of contrast material.

Primary Malignancies:

- Among primary malignant tumors of the sternum, which are rare overall, the most common is chondrosarcoma.
- Chondroid matrix mineralization, which is sometimes recognizable on lateral chest radiographs, is better depicted on CT images, on which it appears as multiple stippled calcifications.

Osteosarcoma:

- Sternal osteosarcoma, in comparison with osteosarcoma of the extremities, is more commonly reported in older patients (Median age, 42 years).
- A significant percentage of sternal osteosarcomas are secondary lesions found in patients who previously underwent radiation therapy.
- Radiographically, sternal osteosarcoma appears as a mass like lesion that consists of bone and soft tissue and contains areas of osteolytic change, calcification, and osteoid matrix.

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Benign Lesions:

- In the largest series of neoplasms of the sternum, only six benign lesions were seen at the Memorial Sloan-Kettering Cancer Center in 64 years and only three benign lesions were seen at the Mayo Clinic in 87 years.
- The spectrum of these lesions includes enchondroma, osteochondroma, hemangioma, hemangiopericytoma, enostosis, osteoid osteoma, fibrous dysplasia, Paget disease, Langerhans cell histiocytosis, aneurysmal bone cyst, eosinophilic granuloma, giant cell tumor, brown tumor, nonossifying fibroma, and chondromyxoid fibroma.

DISCUSSION:

- Solitary bone plasmacytoma (SBP) is defined as the clonal proliferation of plasma cells identical to those of plasma cell myeloma, which manifests itself as a localized osseous growth. Plasma cell neoplasms account for approximately 1% to 2% of all human malignancies and occur at a rate of about 3.5/100,000 per year. SBP is composed of monoclonal plasma cells, which are cytologically, immunophenotypically, and genetically identical to those seen in MM.^[1]
- MM is a multi-focal plasma cell proliferation in the bone marrow, produces excess immunoglobulin, and infiltrates bone. Free light chains are also produced along with intact proteins; these light chains are detected by urine protein electrophoresis and are designated as Bence-Jones proteins. Excess cytokines activate osteoclasts, leading to bone destruction and subsequently to discrete lytic lesions or diffuse osteopenia. Increased bone resorption also leads to hypercalcemia associated with neurologic manifestations. Patients are susceptible to recurrent bacterial infections due to the suppression of normal humoral immunity, which is frequently the cause of death. Bence-Jones proteins are toxic to renal tubular cells and thus may contribute to renal insufficiency or failure.^[2]
- Chondrosarcoma is the most common tumor among primary malignant tumors of the sternum, although its incidence is extremely low. Of the 11,087 bone tumors in the Dahlin tumor series at the Mayo Clinic, only 66(0.6%) were primary malignant tumors of the sternum. Of these 66 tumors, 22(33%) were chondrosarcomas; 20(30%) were myelomas, including plasmacytomas; 14(21%) were lymphomas; 8(12%) were osteosarcomas; 1(1.5%) was a fibrosarcoma; and 1 (1.5%) was a Ewing tumor.^[3] Zehani et al.^[4] in a study of primary malignant tumors of the sternum observed for 16 years between 1993 and 2009, reported that primary tumors of the sternum were very rare and accounted for only 0.5% of all of the primary bone tumors they encountered and that these tumors were often malignant, osteolytic, and aggressive. They also described 6 cases of primary malignant tumors of the sternum, of which 3 were plasmacytomas, 1 was a chondrosarcoma, 1 was an osteosarcoma, and 1 was a large B-cell lymphoma. Plasmacytoma arises in bone or soft tissue. SBP is defined as a tumor confined to a bone with no multiple osteolytic lesions, while extramedullary plasmacytoma (EMP) is defined as a tumor that occurs only in a soft tissue with no multiple osteolytic lesions. EMP mainly involves the head, neck, and lungs.^[2] SBP is relatively rare and accounts for 3% to 5% of all plasma cell neoplasms.^[1] The median age at the diagnosis of SBP is 55 years, and SBP occurs 10 years earlier than MM. Males are more frequently affected than females, and one study reported that two-thirds of all patients were male.^[2] SBP more commonly involves the axial skeleton, and distal appendicular diseases are extremely rare. As in MM, marrow areas with active hematopoiesis are targeted, including the vertebrae, ribs, skull, pelvis, femur, clavicle, and

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scapula, in order of decreasing frequency.^[1] The thoracic vertebrae are more frequently involved than the lumbar, sacral, and cervical spines. SBP originates in the medullary cavity and erodes through cancellous and cortical bone.

- Thus, punched-out defects are seen on radiographs and usually measure 1 to 4 cm. Patients may present with pathologic fractures or skeletal pain. Further, cord compression may be the presenting feature of solitary plasmacytoma involving the vertebrae. Soft tissue extension of the tumors may result in palpable masses, particularly when the rib is involved. Therefore, SBP should be distinguished from MM.
- The International Myeloma Working Group has established the following criteria for the diagnosis of SBP.^[5] The diagnosis of SBP requires a solitary bone lesion; a biopsy which shows histological evidence of bony involvement by plasma cells; negative results of complete skeletal radiographs, including the humerus and femur, that show no lytic lesions; absence of clonal plasma cells in a random sample of bone marrow; no evidence of anemia, hypercalcemia, or renal involvement suggestive of systemic myeloma; and immunofixation of serum and concentrated urine that shows no monoclonal proteins.^[6]
- Like MM, SBP has a lytic appearance on plain radiographs. CT and particularly, magnetic resonance imaging (MRI) depict the extent of SBP more clearly. MRI is useful for identifying additional unsuspected plasma cell lesions that do not appear in a skeletal survey.
- Some recent studies have emphasized the necessity of CT and MRI along with 99m Tc-MIBI scintigraphy, while others have mentioned the increased diagnostic sensitivity of fluorine-18 fluorodeoxyglucose PET. Electrophoresis of serum and urine samples reveals monoclonal proteins in 24% to 72% of SBP patients, although protein levels are considerably lower in SBP patients than in MM patients. All SBP patients should undergo serum and urine immunofixation even when electrophoresis results are normal because monoclonal proteins may not be detected in approximately one-third of all patients.^[1,6]
- SBP can be definitively diagnosed by a biopsy. The gross and microscopic features of solitary plasmacytoma are identical to those of MM. Histologically, plasmacytoma appears as sheets of plasma cells. These are small round blue cells with 'clock-face' nuclei and abundant cytoplasm with a perinuclear clearing or 'halo.' Plasmacytoma exhibits monoclonal kappa or lambda light chains, whereas plasma cells of reactive chronic osteomyelitis are polyclonal.^[3,4]
- Radiotherapy is the treatment of choice for SBP; however, there exists controversy regarding extensive wide resection. Treatment fields should be designed to encompass all diseases shown by MRI or CT scanning and should include a margin of normal tissue. Localized radiotherapy should be administered even if the tumor is completely removed for diagnostic purposes. The local response rate has been shown to be 80% to 90%, and there is no clinical evidence that adjuvant or prophylactic chemotherapy prevents the ultimate development of MM.
- Approximately 55% of patients with SPB develop MM within 10 years of successful treatment; 10% develop local recurrences or solitary plasmacytomas at different locations.^[7] Although solitary plasmacytoma occurs less commonly in the sternum than MM, it must be considered in the differential diagnosis of bone and soft tissue tumors, particularly in the absence of lytic lesions in a skeletal survey and in the presence of clinical evidence of end organ damage.
- Unfortunately, more than half of the patients with solitary plasmacytoma develop MM during their lifetime.

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