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Solitary Plasmacytoma of Manubrium Sterni - A Case Report

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Case Report

Solitary Plasmacytoma of Manubrium Sterni – A Case Report

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Abstract

Solitary plasmacytoma is a tumour consisting of neoplastic monoclonal plasma cells without systemic involvement. It constitutes about 5% of all plasma cell dyscrasias and is mainly detected in the spine, pelvis, ribs, and rarely in the sternum. Here we report a case of a 45-year-old man presenting with pain and swelling in the midline of the chest over manubrium sterni. Plain radiograph and CT scan revealed an expansile, osteolytic lesion in the manubrium sterni. There was no M Band in protein electrophoresis, and the iliac bone marrow aspirate did not show plasmacytoma. However, histopathology of the lesion was suggestive of plasmacytoma.

Key words: Manubrium sterni, Plasmacytoma, Solitary.

Introduction

Solitary plasmacytoma of bone is a primary bone neoplasm occurring mainly in the axial skeleton. It represents about 5% of all plasma cell dyscrasias and is mainly detected in the spine, pelvis, ribs, and rarely in the sternum.[1] The natural history of the neoplasm is progression to multiple myeloma; some patients may develop local recurrences in bone. The disease-free interval after treatment is about 40% at five years and 15 to 30% at 10 years as per literature.[2,3]

There is no consensus about prognosis and management though radiotherapy is preferred. However, there is some debate about the dose of radiation and its effect on the response. The role of surgery and chemotherapy is also not clear.

Case Report

A 45-year-old gentleman presented to the pulmonary medicine department with chest pain and swelling in the anterior chest wall in the midline. The pain was dull aching, and continuous with occasional radiation to the neck. He had a fine needle aspiration cytology report showing non-specific inflammation. A 4x4 cm swelling was seen over the upper third of the sternum on examination. The pulmonologist suspected skeletal secondary, myeloma and tuberculosis. He was referred to Orthopaedic surgery to confirm the diagnosis and further management. Routine blood tests were normal except for a high erythrocyte sedimentation rate (ESR=62 mm/hr). The chest radiograph PA view was normal; the lateral view showed an expansile, lytic lesion of manubrium sterni with breaching of both anterior and posterior cortices (Figure 1). Computerized Tomography (CT) scan confirmed the presence of an expansile, osteolytic lesion involving the upper third of the sternum infiltrating into the anterior mediastinum (Figure 2). Laterally the lesion extended to the costochondral junction. These features were consistent with a malignant tumour. Though the clinical features and radiological findings indicated a primary neoplasm, a whole-body $Tc99^{m}$ bone scan was done to look for skeletal lesions, which was grossly normal. Work-up for possible primary sites was did not demonstrate any pathology. There was no M Band in protein electrophoresis. Bone marrow study from the posterior superior iliac spine was negative for myeloma.

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We did an open biopsy of the lesion through a midline incision. Histopathological examination showed a cellular tumour composed of diffuse infiltration by sheets of plasmacytoid cells having eccentrically placed nuclei. This appearance was suggestive of a plasmacytoma (Figure 3). The skull radiograph was free of punched-out lesions.

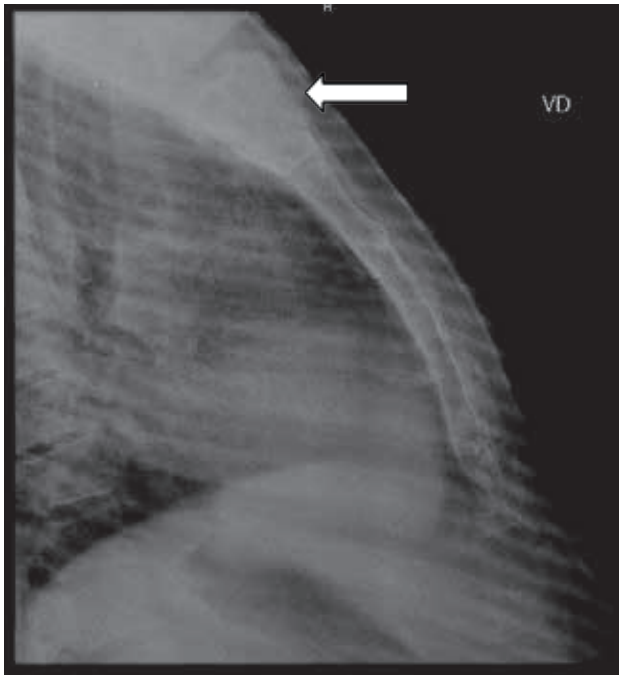

Figure 1: Lateral view of chest showing expansile lesion of Manubrium Sterni (Arrow)

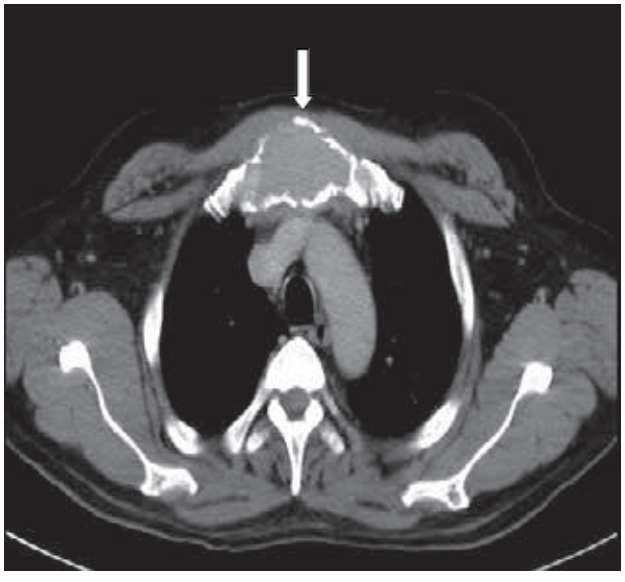

Figure 2: Axial CT scan showing expansile lytic lesion of manubrium sterni, infiltrating into the mediastinum.

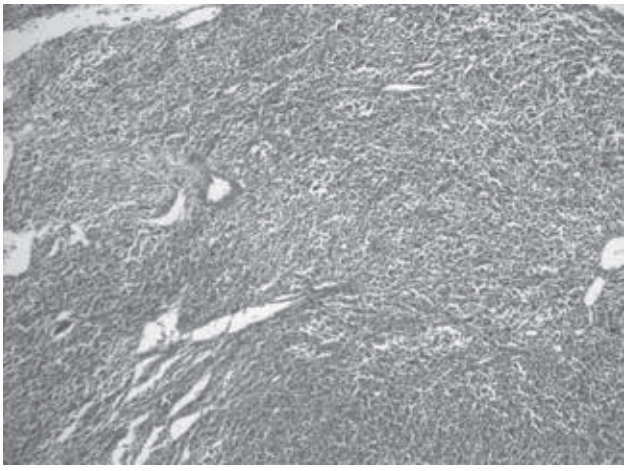

Figure 3: Histopathology (10X) of the lesion showing malignant plasma cells.

Further evaluation revealed a monoclonal gammopathy, confirming the diagnosis of plasmacytoma. He was later referred to an oncologist. Six cycles of the category 'A' chemotherapy with thalidomide, dexamethasone, and zoledronic acid were given. The patient had persistent pain, tenderness, and mild swelling at the local site even after six cycles of chemotherapy. He was treated with a course of palliative radiotherapy, after which symptoms subsided. Thalidomide treatment was continued for three more months. The patient remained well for two years under regular follow-up without recurrence or progression to multiple myeloma.

Discussion

Solitary plasmacytoma is defined as a monoclonal proliferation of plasma cells indistinguishable from multiple myeloma, which manifests as a localised bony growth. The most common sites described are in areas of active haemopoiesis, such as the axial skeleton. About 10 to 15% of cases occur in the ribs and sternum.[1,4] Solitary plasmacytoma is more common in men (M:F = 2:1), with a median age of 55 years at presentation. That is about 10 years earlier than the presentation of multiple myeloma. The patient presents with pain at the site of the lesion.[2]

Similar cases of solitary lesion in the sternum have been reported in the literature.[1,2,4,5] It is challenging to do the initial staging of solitary plasmacytoma. Conventional radiographic skeletal survey and

bone scan understage the disease in some patients. Magnetic Resonance Imaging (MRI) of the entire spine is needed to exclude additional lesions.[6] Alper et al.,[7] compared these investigations with a $Tc99^{m}$-MIBI scan to increase the sensitivity of diagnosis. More promising preliminary results are available in the literature regarding the usage of Positron Emission Tomography (PET).[8,9]

The surgical method of obtaining a biopsy is usually decided based on the size of the tumour. Incisional biopsy is the preferred approach to a lesion, and the biopsy scar is excised during definitive surgery. However, complete excision is not always possible.

Patients diagnosed with solitary plasmacytoma on incisional biopsy should be further evaluated to rule out the possibility of multiple myeloma. There must not be any signs of systemic disease. A complete skeletal survey must not show any other lesions, and the bone marrow away from the solitary lesion should not have more than 5% plasmacytosis. Immunofixation, immunoelectrophoresis, and immunoglobulin quantitations are carried out along with routine haematology and biochemistry investigations. These patients require definitive chemotherapy and local radiotherapy.[6]

In those patients whose tumour dimensions are less than 4 cm, routine haematology and biochemistry tests and a whole-body CT scan are performed. If no contraindication, complete surgical excision and primary reconstruction are undertaken. Complete resection is curative,[5] but adjuvant chemotherapy and local irradiation are considered depending on histopathology and further staging.[12]

The most crucial factor that is detrimental to the long-term survival of these patients is the development of multiple myeloma.[13]

Conclusion

Excision of the lesion can be performed if the solitary myeloma is less than 4 cm. Incisional biopsy is the preferred approach to a tumour greater than 4 cm. Complete excision is thought to be curative but not always possible. Adjuvant chemotherapy and local radiotherapy are to be considered depending on the histopathology and staging of the neoplasm. Optimum patient selection is crucial in both groups as certain patients are highly likely to get cured. Routine clinical examination and laboratory screening is mandatory to detect local or systemic recurrences.

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