Case Report

Solitary plasmacytoma with secondary amyloidosis

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Abstract

Solitary plasmacytoma is a localized lesion and extremely rare amongst the plasma cell tumors. Unlike multiple myeloma there is no evidence of disseminated disease. We present the case of a seventy five year old female presenting with chest pain which turned out to be a solitary plasmacytoma of eighth rib on the right side which itself is a rare disease and the histopathology specimen revealed the presence of amyloid material making it all the more interesting as it turned out to be a case of secondary amyloidosis in this rare tumor.

Key words: Solitary plasmacytoma, Secondary amyloidosis

Case report

A seventy five year old female presented to us with right sided chest pain of six months duration and breathlessness of one week duration. The chest pain was more or less localized to the infra axillary area which was a dull aching pain with tenderness on deep palpation. She gave history of MMRC Grade II dyspnea and no wheezing during this episode. She gave history of recurrent episodes of wheezing for the last six years. She also gave history suggestive of rhino bronchial allergy. She was getting regular treatment for moderate persistent asthma for the last two years and was well controlled. She was on treatment for systemic hypertension for the past five years. There was no history of loss of appetite, loss of weight, cough or haemoptysis. She gave history of blunt trauma to chest about five years back. General examination revealed pallor. There was no significant lymph node enlargement. Respiratory system examination showed kyphosis and fullness over the right infra axillary area and tenderness could be elicited. There was impaired resonance in the right infra axillary area with reduced vocal fremitus. Breath sounds and vocal resonance were decreased in the above area. Hence a provisional diagnosis of right lower lobe peripheral mass lesion with possible chest wall involvement in a moderate persistent well controlled asthmatic was made.

Her investigations were as follows; Hb: 10.8gm%, TC- 10400, DC P42 L 39    E19, ESR -40 mm/1st hr, RBS-112mg%, Sputum AFB X 2 samples -negative, Urea - 10mg%, S.creatinine - 0.8.Her liver function tests were normal. Sputum cytology was negative. Mantoux test was non reactive.
Her Chest X-ray PA view revealed a well defined opacity involving the right lower zone close to the costophrenic angle with a pleural base and making an obtuse angle with the chest wall which localises the lesion to the extra pleural region fig (1). This mass was clearly made out (bold arrow) in the right lateral projection also fig (2). Hence the differential diagnoses of a pleural based mass lesion, chest wall mass, encysted effusion or a peripheral lung mass with chest wall extension were considered. In view of her high eosinophil count, a peripheral smear was done which revealed normocytic normochromic blood smear with eosinophilia. Her urine Bence jones protein was negative and serum electrophoresis to rule out multiple myeloma did not reveal the presence of M band fig (3).

Lateral view of her skull did not reveal any punched out lesions. A CT-Chest taken showed an expansile lytic lesion with cortical break and multiple bone fragments associated with intra and extra thoracic soft tissue component involving the anterolateral aspect of 8th rib on the right side and the possibilities considered were metastasis or a lesion confined to rib like tuberculous osteomyelitis or solitary plasmacytoma. fig (5&6)

Fig.5 CT chest mediastinal window showing the peripheral lesion arising from the rib
Hence we proceeded with a Trucut biopsy of the chest wall lesion and a bone marrow study, as we wanted to rule out a plasma cell dyscrasia in this patient fig (3). Bone marrow biopsy showed mild increase in the eosinophilic precursors with 2% plasma cells. Erythroid, myeloid precursors and megakaryocytes were seen normally. Her trucut biopsy specimen yielded sheets of plasma cells infiltrating abundant eosinophilic material which was suggestive of amyloid material. This was hence subjected for immunohistochemical studies and congo red staining which was CD 138 +ve and congo red positive with kappa restriction Fig (7,8,9, 10&11).
Congo red staining shows brisk uptake

Hence a diagnosis of solitary plasmacytoma of the right eighth rib with secondary amyloidosis was made. She was advised local radiotherapy and limited resection. The patient however was not willing for further management and opted for palliative care.

**Discussion**

Solitary plasmacytoma of bone (SPB) is a rare localized lesion that accounts for only 4% of all malignant plasma cell tumors. It is relatively benign plasma cell proliferative disorder with monoclonal plasma cell proliferation. Myeloma on the other hand is a clonal haematopathy. Plasmacytomas can be divided into multiple, solitary osseous, and solitary extraosseous or extramedullary plasmacytomas. The incidence of SPB has been reported to be 3/10 00 000 annually. The average age on presentation was 59.5 years with a range from 39 to 77 years. It may progress to multiple myeloma in 10% cases. To make a diagnosis of solitary plasmacytoma, there should be: 1) Single area of bone destruction due to clonal plasma cells 2) Bone marrow not consistent with multiple myeloma 3) Normal skeletal survey 4) No end organ damage other than solitary bone lesion. According to the current recommendations, the detection of a monoclonal component in the serum or urine does not exclude a diagnosis of solitary plasmacytoma. CD138 or syndecan 1 positivity of the trucut specimen can be considered as an excellent marker for plasmacytic differentiation. Kappa restriction demonstrated in immunohistochemistry signifies the clonality of light chain present in the lesion. Congo red staining with yellow green birefringence under polarized light is the diagnostic technique for demonstrating amyloid material in tissue specimen and our specimen was proved to have amyloid deposits which is hence secondary amyloidosis in a SPB. Treatment is by preoperative radiotherapy with complete en-block resection, followed by adjuvant chemotherapy. Radiation therapy was used as the primary treatment for solitary plasmacytoma. Aviles et al, observed that most patients treated with adequate radiation therapy alone will develop multiple myeloma within the first 3 years after diagnosis and treatment. Low doses of melphalan and prednisolone contributed to an improvement in disease-free survival and overall survival in patients with SPB, compared with patients who were treated with radiotherapy alone. Hence preoperative radiotherapy with complete en-block resection, followed by adjuvant chemotherapy, is the currently accepted best treatment strategy for SPB.

**References**