

A Rare Case of Mediastinal Plasmacytoma Mimicking Lung Cancer



Medical Science

KEYWORDS : Extra-medullary plasmacytoma , Bronchogenic carcinoma, para-esophageal mass, Immunohistochemistry

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ABSTRACT

Plasmacytomas are a localized proliferation of plasma cells in the bone marrow, and are less frequently seen in extra-osseous organs or tissues. Extra-medullary plasmacytoma is a rare malignant neoplasm, and is especially uncommon when it arises from the mediastinum¹. Here, we report a case of mediastinal extra-medullary plasmacytoma in a 70-year-old man. He was admitted with right sided pleural effusion and para-esophageal mass which was provisionally diagnosed as bronchogenic carcinoma. However subsequent investigations revealed it as a case of plasmacytoma by tissue biopsy and immunohistochemistry (IHC) later confirmed by bone marrow biopsy. Patient completed first cycle chemotherapy comprising of cyclophosphamide and thalidomide.

INTRODUCTION

Plasmacytoma, a neoplastic proliferation of plasma cells, is a form of plasma cell dyscrasia that may manifest as multiple myeloma, primary amyloidosis or monoclonal gammopathy of unknown significance. Plasmacytoma may be primary or secondary to disseminated multiple myeloma and may arise from osseous (medullary) or non-osseous (extra-medullary) sites. Primary extra-medullary plasmacytoma can be solitary or multiple. Mediastinum is rarely involved by extramedullary plasmacytoma (EMP).

EMP constitutes about 4% of all plasma cell tumors. The most common site for extra-medullary involvement is the upper aero-digestive tract . The mediastinum is rarely involved in extramedullary plasmacytoma . Only 5% of patients with EMP's have coexistent multiple myeloma .

CASE REPORT

70 yr old male with past history of myocardial infarction admitted with complaints of breathlessness, cough and dull aching right sided chest pain since 15 days. Cough was non-productive with no hemoptysis. He had no fever, weight loss or wheezing within this time period. He had no known chemical or occupational exposure.

Clinical examination revealed presence of pallor and clubbing. Respiratory system examination revealed decreased air entry on right hemithorax. A routine evaluation was initiated, which included basic blood investigations, chest radiograph and CT thorax.

Chest radiograph (Figure 1) showed right sided moderate pleural effusion

Computerized tomogram of thorax (Figure 2) is showing well defined lobulated lesion in right para-esophageal region with pleural effusion and pleural based lesion likely metastatic.



Figure 1- Chest radiograph showing right sided pleural effusion.



Figure 2 – computed tomography of thorax showing fairly defined lobulated lesion in paraesophageal region

Pleural fluid investigations showed lymphocytic predominant transudative effusion. Pleural fluid ADA was 42 IU/L. Pleural fluid cytology was negative for malignant cells. CT guided biopsy from para-esophageal mass was positive for malignancy and differential diagnosis given were adenocarcinoma and mesothelioma. On IHC cells were positive for CD 138 (Figure 3). Renal function and calcium levels were normal. Bone marrow biopsy (Figure 4) showed marked increase in plasma cells (65%). Serum protein electrophoresis showed decrease in albumin with increase in alpha 1 and alpha 2. M band was not detected.

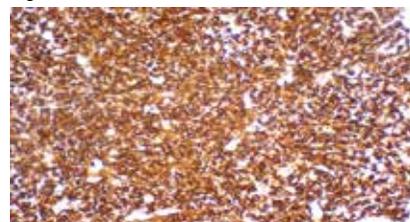


Figure 3- IHC showing CD 138 positive cells

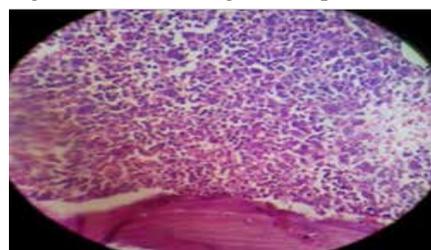


Figure 4- Marked increase in plasma cells in bone marrow biopsy

Patient received first cycle of chemotherapy as advised by Hemato-oncologist, consisting of cyclophosphamide and thalidomide.

DISCUSSION

Multiple myeloma is a neoplastic disorder caused by the proliferation of transformed B lymphoid progenitor cells that give rise to a clone of malignant immunoglobulin-secreting plasma cells. Multiple myeloma usually manifests as a diffuse bony disease (myelomatosis), but it can sometimes present as a solitary plasmacytoma of a bone or as extra-medullary (extra-osseous) plasmacytomas¹. The EMP arising from the mediastinum is extremely rare. Although rare, various patterns of thoracic involvement of multiple myeloma have been reported, including a lung mass, multiple pulmonary nodules, diffuse reticulonodular infiltration, lymph node enlargement, a mediastinal mass, nodular pleural thickening and pleural effusion, and tracheobronchial infiltration².

EMP must be distinguished from reactive plasma cell lesions and lymphoma. It should be demonstrated that the infiltrate consists entirely of plasma cells and that there is no B cell component. In this regard CD138, MUM1/IRF4, CD20 and PAX5 are the most useful markers although it should be recognized that CD20 and PAX5 are sometimes expressed in plasma cell malignancies. Monoclonality and/or an aberrant plasma cell phenotype should be demonstrated with useful markers being CD19, CD56, CD27, CD117 and cyclin D1^{3,4}. All diagnoses should be made or reviewed by specialist haematopathologists in accordance with NICE guidelines for improving outcomes in haematological cancers⁵. There have been no significant publications since the previous guideline on the role of MR scanning in the diagnosis or prognosis of patients with EMP. Additionally, because of the fewer numbers of patients, experience in the role of PET scanning for patients with EP is also scanty. Recommended investigations for all patients diagnosed with EP therefore remain unchanged from the previous guideline

The sequences of proceedings suggest that the mediastinal plasmacytoma provided an early hint to the diagnosis of multiple myeloma, and therefore we conclude that the multiple myeloma was coexisting with the mediastinal lesion. Another aspect of our case is that it presented like a diagnostic dilemma; we initially thought that the diagnosis of occult lung cancer with mediastinal involvement. However, diagnosis of plasmacytoma with multiple myeloma was reached after extensive investigations.

Extramedullary plasmacytoma should be treated by radical radiotherapy encompassing the primary tumour with a margin of at least 2 cm. Newer agents including thalidomide and bortezomib have also been used successfully in small numbers of patients with relapsed plasmacytoma^{6,7}. Our patient received thalidomide along with cyclophosphamide due to bone marrow involvement.

CONCLUSION

Extramedullary plasmacytoma is a rare presentation of multiple myeloma. In this case the presentation of the patient mimicked lung malignancy. This should be considered as a rare differential diagnosis of carcinoma lung.

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