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EXTRAMEDULLARY PLASMACYTOMA: RARE PRESENTATION OF THE ANTERIOR MEDIASTINAL MASS

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RESUME
Plasmacytomas are tumors originating from plasma cells, categorized as solitary bone plasmacytomas (SPB) or extramedullary plasmacytomas (EMP), the latter being rare and accounting for 3–5% of cases. EMPs typically occur in the upper aerodigestive tract but can arise elsewhere, including the mediastinum. Diagnosis involves confirming monoclonal plasma cells without systemic disease. EMPs respond well to radiotherapy, with a generally favorable prognosis and a lower likelihood of progression to multiple myeloma compared to SPB. Adjuvant chemotherapy is debated and reserved for persistent cases.

INTRODUCTION

Plasmacytomas are localized tumors originating from plasmacytic cells, typically found in the bone. The extra medullary form is uncommon, accounting for less than 5% of plasmacytomas. More than 90% of extra medullary plasmacytomas occur in the head and neck, especially in the upper airways, while thoracic locations are very rare. This report presents a case of mediastinal plasmacytoma to examine the epidemiological and anatomoclinical aspects of this rare entity and to discuss its prognosis.

CASE REPORT

The patient was a 63-year-old woman being monitored for hypertension, which was well-controlled with amlodipine. She had never smoked and had no history of respiratory exposure.

The clinical presentation evolved over twenty days, characterized by the onset of fluctuating, atypical thoracic pain without distinct radiation. This was subsequently accompanied by the emergence of a mass anterior to the sternum, resulting in anterior chest wall deformity. The patient also experienced a nonproductive cough that was irritative in nature, along with progressively worsening dyspnea, which led to a medical consultation and a chest radiograph. The physical examination revealed a deformed chest wall with a slight right parasternal curvature. The peripheral lymphatic areas were free.

The chest X-ray (Figure 1) revealed a mediastinal opacity extending to both sides, with irregular external borders. Bilateral pleurisy was also present. Haemogram showed no abnormalities in any of the three blood lines. The thoracic CT scan (Figure 2 & 3) revealed a tissue mass in the anterior mediastinum, with heterogeneous enhancement on contrast injection. This process invaded the anterior chest wall. Laterally, it made intimate contact with the pericardium and mediastinal pleura. Posteriorly, it pushed back all the mediastinal structures, with no obvious separation line. A transthoracic biopsy under CT scan control was performed, the histological study was in favour of plasmacytoma.

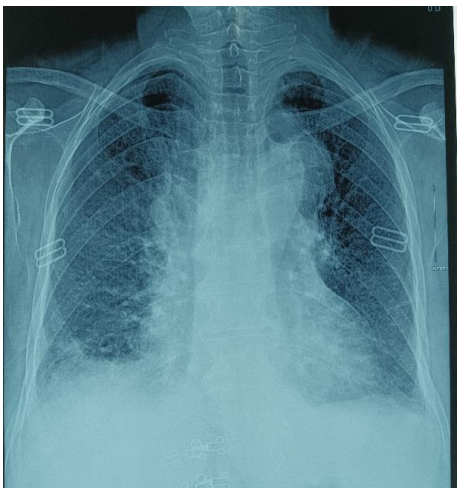

Figure 1: Chest X-ray showing a mediastinal opacity.

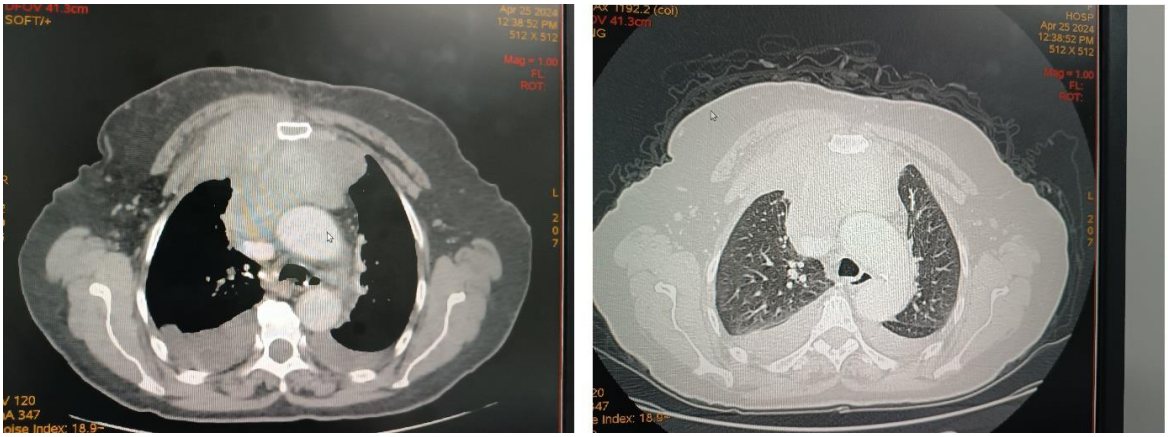

Figure 2: Chest CT revealing the tissue mass in the anterior mediastinum.

DISCUSSION

Plasma cell neoplasms are classically categorized into four groups; multiple myeloma (MM), plasma cell leukemias, solitary plasmacytomas of the bone (SPB), andextramedullary plasmacytomas (EMP). These tumors may be further described as localized or diffuse in presentation.[1]

Plasmocytoma is an immunoproliferative monoclonal disease of the B cell line; it's classified as non-Hodgkin lymphoma.[2] EMP refers to localized plasma cell neoplasms occurring in tissues outside of the bone. This rare subtype accounts for approximately 3-5% of all plasmacytomas, with a global annual incidence of 3 per 100,000. The median age of diagnosis is 55 years, and 34% of affected patients are male.[3,4]

Plasmacytomas that occur extramedullary are predominantly located in the upper aerodigestive tract, accounting for approximately 80% of cases. However, it can arise any part of body in 17.8% (liver, lung, spleen, pancreas, kidney, lymph nodes, digestive tract, thyroid, heart, testis, ovary, and skin). These lesions typically present as solitary masses, although they may also arise as part of an underlying multiple myeloma.[5]

A diagnosis of solitary plasmacytoma necessitates fulfillment of the following criteria.[6]

1) Tissue biopsy and histological and immunohistochemical confirmation of the presence of a homogenous infiltrate of monoclonal plasma cells, which typically express CD138 and/or CD38.
2) Negative bone marrow analysis demonstrating no evidence of clonal plasmacytosis.
3) Absence of extramedullary skeletal involvement; and.
4) No laboratory findings indicative of anemia, hypercalcemia, or renal dysfunction.

A unilateral bone marrow (BM) aspiration and trephine biopsy are recommended for all patients with suspected solitary plasmacytoma (SP). In order to exclude > 10% of monoclonal PCs in the BM, a BM aspiration with immunophenotyping to define the proportion of monoclonal cells by kappa/lambda labeling should be performed.[7] A BM plasmacytosis exceeding 10% confirms a diagnosis of multiple myeloma (MM). The BM aspiration holds both diagnostic and prognostic significance. If monoclonal PC infiltration is detected at baseline, repeat aspiration and biopsy should be conducted upon suspicion of progression to MM.

In our case, the initial blood test was normal, and the serological examination showed a gradual increase in gammaglobulin levels and light chain lambda during the recurrence. The diagnosis of EMP was confirmed through a CT-guided transparietal biopsy of the mediastinal tissue mass, the bone marrow biopsy showed no medullary plasma cell infiltration.

EMP is highly responsive to radiation, with 80 to 100% of patients achieving local control in cases with low-grade histology. Radiotherapy doses of 35 to 45 Gy are commonly used for patients with tumors larger than 5 cm. Therapy is generally given daily at a rate of 1.8–2.0 Gy per fraction and a margin of at least 2 cm should be employed.[8,9]

Surgery may be considered for localized EMP, but it should be reserved for the treatment of pathological fractures and neurological complications.[7]

The role of adjuvant chemotherapy (regimens used for multiple myeloma) after radiation therapy in the treatment of SP remains controversial precluding a definite recommendation. It can be considered for patients with persistent disease, based on PET/ CT, after initial radiotherapy.[9-10]

Among all plasma cell tumors, EMPs have the best prognosis.[11-12] The presence of local involvement of adjacent bone or lymph nodes does not necessarily indicate systemic disease or a worse prognosis. Additionally, the progression to multiple myeloma (MM) is less likely in patients with EMPs compared to those with solitary plasmacytoma of the bone (SPB).

CONCLUSION

This case highlights a rare instance of mediastinal extramedullary plasmacytoma (EMP) in a 63-year-old woman. Diagnosis was confirmed via CT-guided biopsy, revealing no bone marrow involvement. EMPs are rare but highly responsive to radiotherapy, with a favorable prognosis and a low risk of progression to multiple myeloma.

Competing interests

The authors declare that they have no competing interests.

Authors contributions

All the authors have contributed to this manuscript.
All the authors have read and approved the final version of the manuscript.

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