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A Case Report on Plasma Cell Leukemia Presenting as a Chest Wall Mass

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Plasma cell leukemia (PCL) is an uncommon neoplasm of plasma cells with an aggressive clinical course and a poor outcome, even with the current standard of care. It can occur either de novo (primary PCL) or as a progression of multiple myeloma (MM). This disease has unique diagnostic criteria, but certain genetic markers and clinical features may overlap with multiple myeloma (MM). Due to the low prevalence of PCL, guidelines on its management are extrapolated from the management of MM and are based on small retrospective studies and case reports/series. We report the case of a sixty-nine-year-old man referred to the hematology department for the diagnosis of pPCL, revealed by thoracic plasmacytomas mimicking a thoracic neoplasm. The diagnostic approach, management, and outcomes of PCL are discussed.

Keywords: Plasma cells leukaemia; multiple myeloma; thoracic mass.

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1. INTRODUCTION

"Multiple myeloma (MM) is a neoplasm of plasma cells, accounting for 10–15% of hematopoietic neoplasms. It is more prevalent in individuals of African descent, occurring twice as frequently compared to Caucasians" [1]. The term plasma cell leukemia (PCL) is typically used when there is a significant number of circulating plasma cells. PCL represents the most aggressive form of plasma cell dyscrasias, defined by the presence of > 2 X 10^9/L peripheral blood plasma cells or accounting for >20% of the differential white cell count, not arising from pre-existing multiple myeloma (MM). Secondary PCL is a leukemic transformation of end-stage MM. Its prognosis is very poor, with a median overall survival of only 7 months with standard chemotherapy.

"The clinical presentation usually involves symptoms attributed to end-organ damage seen in MM (hypercalcemia, renal failure, anemia, and lytic bone lesions) or to leukemia (leukocytosis, thrombocytopenia, and organomegaly) " [2]. We report a case of PCL with an atypical presentation as a chest wall mass and discuss the diagnostic approach as well as treatment options.

2. CASE PRESENTATION

We present the case of a 69-year-old patient, a smoker with a history of 25 pack-years and no significant past medical issues. He was referred to our hospital due to persistent right-sided chest pain lasting over three months, accompanied by apyrexia and a decline in the general state. Upon physical examination, mucocutaneous pallor was noted, along with a hard and painful mass in front of the 3rd right intercostal space. The mass, measuring 8 cm x 5 cm, showed no inflammatory signs and was fixed to the deep plane.

The chest X-ray revealed the presence of a low-density pulmonary opacity in the right inferior hemithorax (Fig. 1), while the chest computed tomography identified a large osteolytic mass centered on the 4th costal arch, with an intrathoracic component measuring 75 x 51 x 39 mm (Figs. 2 and 3).

A ultrasound guided biopsy of the chest mass revealed a diffuse monotonous population of small to medium-sized lymphocytes with plasmacytoid features. The tumor cells tested positive for CD138 and MUM1, displaying monoclonal Kappa restriction by in situ hybridization. Additionally, a high proliferation index was observed with Ki67 (60%). The cells, however, tested negative for CD20 and CD79a. A diagnosis of high-grade plasma cell neoplasm involving soft tissue was established.

Laboratory evaluation revealed a white blood cell count of 22 × 10^9/L, microcytic anemia with a hemoglobin level of 9.9 g/dl, and thrombocytopenia at 82 × 10^9/L. The peripheral blood smear exhibited 79% circulating atypical lymphocytes with plasmacytoid features, confirmed as neoplastic plasma cells through flow cytometry, consistent with PCL.

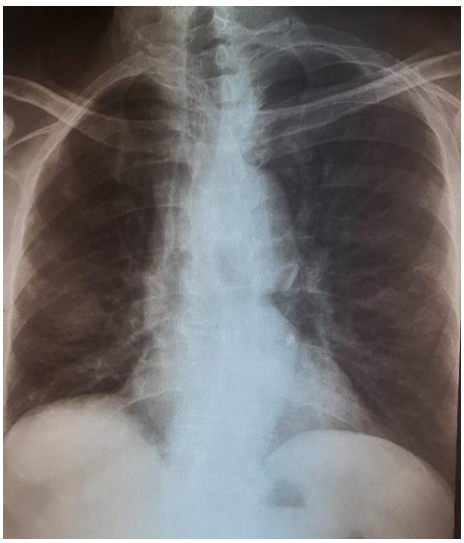

Fig. 1. Frontal chest radiograph depicting a low-density pulmonary opacity in the right inferior hemithorax

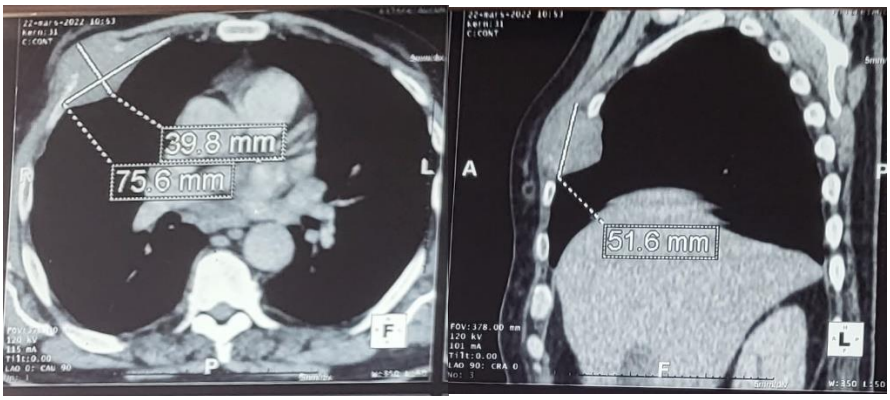

Fig. 2. Axial and sagittal sections of a thoracic CT scan revealing a large osteolytic mass centered on the 4th costal arch

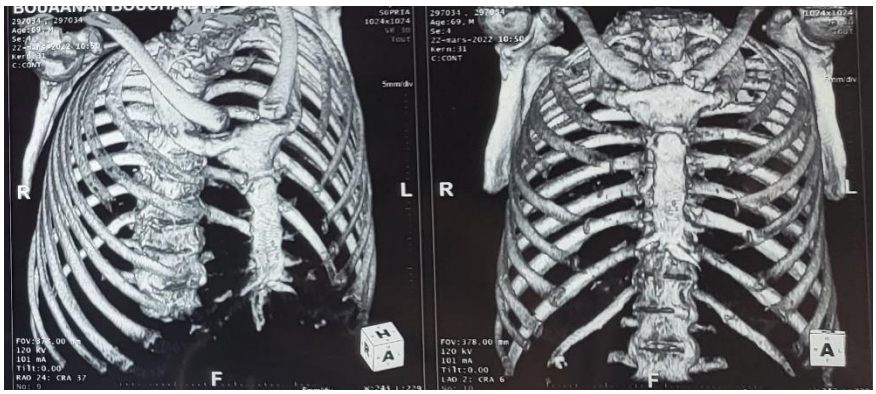

Fig. 3. MIP 3d reconstruction CT scan displaying lysis of the anterior arch of the 4th rib

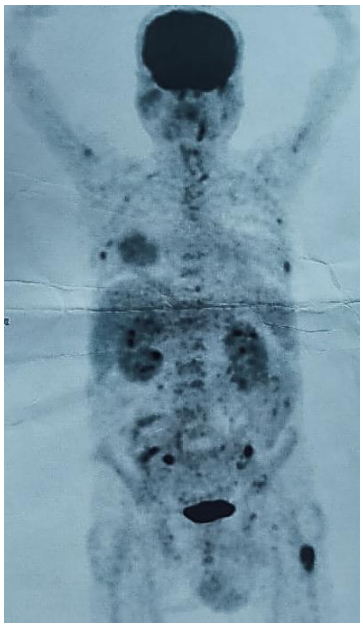

Fig. 4. Maximum intensity projection image of PET/CT displaying a hypermetabolic process centered on the anterior arch of the 4th right rib, exhibiting a lytic appearance. Multiple bone locations with increased metabolic activity are evident, including the thoracic spine (T10), humeral, costal (2nd, 6th, 5th, and 7th ribs), pelvic, and left femoral regions

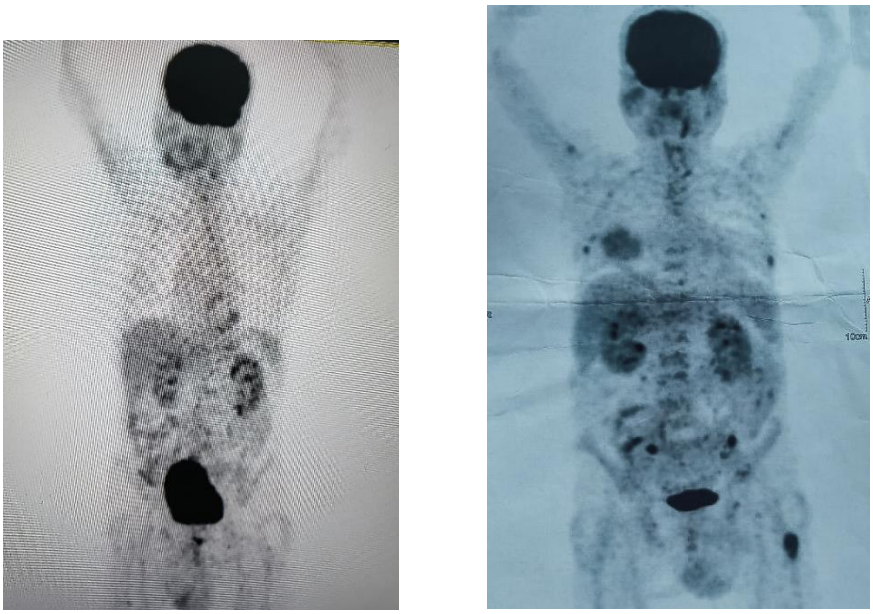

Fig. 5. The maximum intensity projection image of PET/CT performed after the 3rd cycle of chemotherapy reveals a favorable response to treatment, demonstrating complete metabolic regression in the costal, humeral, pelvic, and femoral bone locations

Blood chemistry indicated an inflammatory syndrome, with an elevated CRP of 170 mg/L and hyperproteinemia at 100 g/L, without evidence of tumor lysis syndrome. Serum protein electrophoresis and immunofixation revealed a monoclonal spike with IgA Kappa. The bone marrow biopsy demonstrated diffuse infiltration with 90% atypical plasma cells, confirming the diagnosis of PCL with chest wall involvement.

A PET/CT scan was performed, revealing fluorodeoxyglucose (FDG) avidity in the parietal mass and other bony localizations, including costal, humeral, pelvic, vertebral, and femoral sites (Fig. 4).

The patient was referred to the hematology department, where he underwent induction chemotherapy with VTD-PACE (Velcade, thalidomide, dexamethasone, cisplatin, doxorubicin, cyclophosphamide, etoposide), administered in 28-day cycles for a total of 6 cycles.

After the 3rd cycle, the patient showed significant improvement marked by complete remission, indicated by the absence of circulating blood plasma cells, a plasma cell rate in the bone marrow of 1%, negative electrophoresis of plasma proteins, and no bony lesions observed in the PET scan (Fig. 5).

3. DISCUSSION

"Plasma cell leukemia (PCL) is a rare and aggressive plasma cell neoplasm, representing only 3% of cases. Primary PCL (pPCL) arises without prior MM in 60-70% of cases, while secondary PCL can occur du to leukemic progression in MM-treated patients" [1].

"Patients often present with nonspecific symptoms such as fatigue, loss of appetite, fever, and abdominal distension, which can mimic multiple myeloma" [3]. Distinguishing features include younger age (median age of 55 compared to 65 for MM patients) and worse performance status at diagnosis [4,5]. PCL typically involves extramedullary and minimal bone involvement, along with symptoms like lymphadenopathy and hepatosplenomegaly [6]. "Thoracic extramedullary plasmacytomas can manifest in six different patterns: lung mass, pulmonary nodules, myelomatous infiltration of lymphatics with amyloid deposition, thoracic lymphadenopathy, pleural effusion with pleural-based nodules, and tracheobronchial infiltrates. Primary thoracic involvement is rare, less than 1% of cases" [7].

"The diagnosis of PCL relies on laboratory parameters, as outlined in the consensus statement by the International Myeloma Working Group" [8]. "PCL is defined by the presence of

>20% circulating plasma cells and/or an absolute plasma cell count >2 × 10^9/L. A peripheral blood smear often reveals an atypical appearance of white blood cells. Flow cytometry in plasma cell leukemia typically expresses CD38 and CD138, similar to MM. However, there is a reduced expression of CD56, CD117, CD71, and HLA-DR antigens compared to MM. PCL is more likely to express CD20, CD45, CD19, CD27, and CD23" [9]. Serum and urine protein electrophoresis with immunofixation identify a monoclonal immunoglobulin. Skeletal surveys aid in establishing bone involvement. A bone marrow biopsy with cytogenetics should be performed in all patients diagnosed with PCL. Any soft tissue mass should be biopsied to assess possible extramedullary involvement.

"PCL is highly aggressive, with a high proliferation index, often leading to tumor lysis syndrome that can occur before or after chemotherapy initiation" [10]. "Treatment decisions for primary PCL rely on extrapolated data from small studies and MM trials, given its rarity. Typically, PCL treatment involves aggressive induction therapy incorporating both proteasome inhibitors and immunomodulatory drugs. Bortezomib-based regimens have demonstrated the highest overall response rate, ranging from 69% to 79%, while lenalidomide-based therapies show response rates of up to 60%" [7]. "Following induction therapy, patients undergo autologous stem cell transplantation. However, PCL is associated with short remissions and early relapse, requiring maintenance therapy with agents like lenalidomide, bortezomib, and thalidomide.The median overall survival for pPCL patients has increased from 3 to 4 months in the early 1980s to 13 months due to improved detection and enhanced chemotherapy regimens" (6). In our case, the patient achieved complete remission after the 3rd cycle of VTD-PACE induction chemotherapy.

4. CONCLUSION

We present a case of PCL with an uncommon presentation. Swift diagnostic assessment of a chest wall mass, coupled with peripheral smear evaluation, facilitated the prompt diagnosis of PCL, enabling early initiation of therapy. Given the rarity of PCL, chemotherapy regimens are primarily adapted from those used for MM. Early referral to a bone marrow transplant center is recommended, as combined chemotherapy and hematopoietic stem cell transplantation offer the best chance of achieving and maintaining complete remission.

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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