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Case Report

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Paraskeletal Plasmacytoma Manifesting as a Chest Wall Mass - An Uncommon Presentation of Plasma Cell Disorder

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Abstract

Plasma cell tumors are characterised by the uncontrolled growth of differentiated B-lymphocytes, which produce monoclonal immunoglobulins. Involvement of the bone marrow or systemic symptoms can be observed. Plasma cell neoplasms that manifest as mass lesions can include extramedullary plasmacytoma or paraskeletal plasmacytoma. Solitary extramedullary plasmacytomas are rare tumors consisting of plasma cells located in soft tissues, appearing as isolated masses that do not involve bone structures. The term, paraskeletal plasmacytoma is used if they impact the skeletal system. We report a case of a middle-aged Indian male who presented with a chest wall swelling and underwent an incisional biopsy before the diagnosis of paraskeletal plasmacytoma, which was confirmed through pathology but exhibited only minor systemic symptoms. This case report adds another rare example of multiple myeloma manifestation as a chest wall tumour to the medical community's literature and ascertain the need of proper diagnosis distinguishing paraskeletal plasmacytomas from extramedullary disease to avoid unnecessary surgeries/tests/treatments.

Keywords: ***Paraskeletal plasmacytoma, multiple myeloma, extramedullary disease.***

Background

Plasma cell tumors are characterised by the uncontrolled growth of differentiated B-lymphocytes, producing monoclonal immunoglobulins. The manifestations can be without involvement of the bone marrow or systemic symptoms. They are classified as paraskeletal plasmacytoma if they impact the skeletal system or solitary extramedullary plasmacytoma if they do not [1]. Paraskeletal plasmacytomas are more prevalent than extramedullary diseases, with incidence rates for paraskeletal plasmacytomas ranging from 7% to 34.4% at diagnosis, which remain relatively stable at relapse. In contrast, the incidence rate of extramedullary diseases is between 1.7% and 4.5%, with this rate doubling as the disease progresses [2].

We report a case involving a middle-aged Indian male who presented with a chest wall swelling underwent an incisional biopsy before the diagnosis of paraskeletal plasmacytoma was confirmed through pathology and exhibited only minor systemic symptoms. Our review revealed limited literature documenting solitary extramedullary plasmacytomas presenting as an anterior chest wall mass. This case report adds another rare example of multiple myeloma manifestation as a chest wall tumour to the medical community's literature [3].

Case Report

An Indian male of 45 years presented with swelling over his left chest area that gradually developed over the past year, increasing significantly in size over the last six months. He experienced pain and stiffness for 20 days and reported a low-grade fever for a month, with no history of cough, vomiting, trauma, bleeding from orifices, or abnormal movements. Examination showed he was afebrile (98.6°F), with blood pressure at 118/80 mmHg, and other vitals were normal.

On palpation, a mass measuring 4x4 cm was found in the left infraclavicular region which was neither tender nor erythematous. It had smooth surfaces and a firm consistency. He had no palpable lymph nodes or organomegaly on physical examination. Routine blood tests showed normal results (**Table 1**). A Trucut biopsy of the mass revealed fragmented linear cores with diffuse plasma cell sheets and a few binucleated forms, suggesting plasmacytoma (**Figure 1**).

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Subsequent tests included serum plasma electrophoresis (SPEP) and serum free light chains (SFLC). The SPEP showed a gamma globulin peak of 2.61 (**Figure 2**), while the SFLC revealed elevated kappa chains at 168.98 gm/dL. (**Table 1**). Urine electrophoresis results were normal. A whole-body PET scan revealed an enhancing mass lesion in the left anterior chest wall that also involved the left third rib. (**Figure 3**).

Table 1: Routine Blood Investigations, SPEP, and SFLC suggestive of paraproteinemia

Lab Parameter	Reference Range	Patient's value
Hb	12.0-16.0 gm/dL	15.3
TLC	4.0-11.0 x 10^3/uL	6.8
Serum creatinine	0.74-1.35 mg/dL	0.8
Serum total calcium	8.4-10.5 mg/dL	9.4
Serum total proteins	6.4-8.3 g/dL	7.97
Alb levels	3.5-5.2 g/dL	3.77
Glob levels	2.5 -3.5 g/dL	4.2
α1 chains	0.21-0.35 g/dl	0.26
α2 chains	0.51-0.85 g/dl	0.64
β1 globulin chains	0.34-0.52 g/dl	0.42
β2 globulin chains	0.23-0.47 g/dl	0.26
γ chains	0.80-1.35 g/dl	**2.61**
Free κ light chains	3.3-19.4 mg/L	**168.98**
Free λ light chains	5.71-26.30mg/L	22.43
Free κ/λ ratio	0.26-1.65	**7.53**

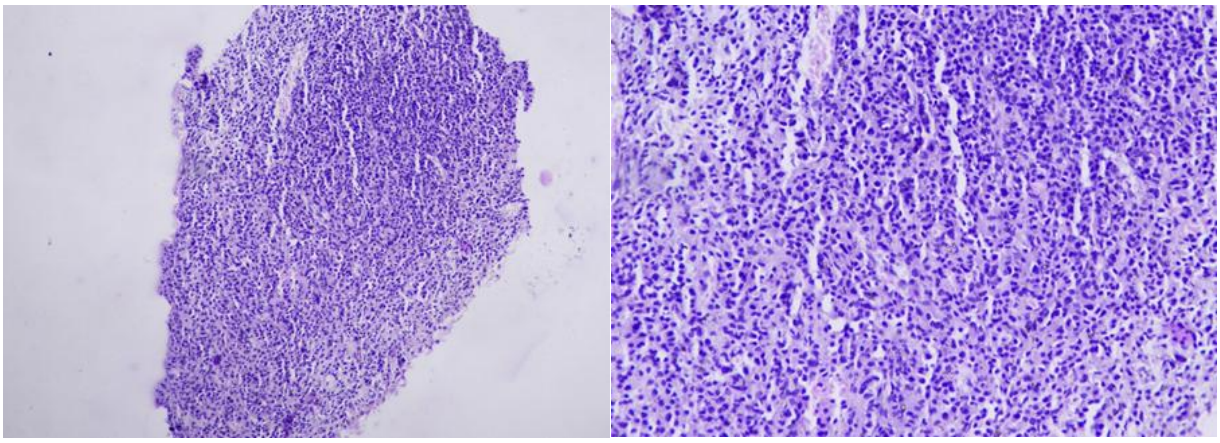

Figure 1: A biopsy specimen stained with hematoxylin and eosin from a chest wall mass reveals diffuse sheets of plasma cells with eccentric nuclei and some binucleated forms, suggesting a possible diagnosis of plasmacytoma.

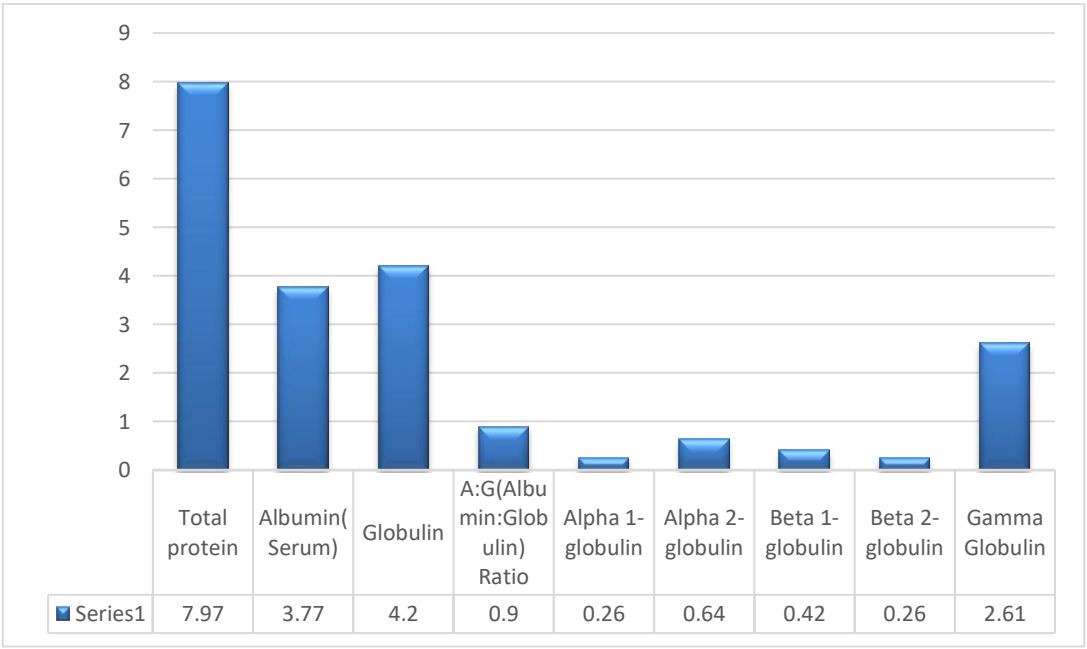

Figure 2: The patient underwent serum protein electrophoresis, which revealed a peak of 2.61 g/dL, suggesting possible paraproteinemia.

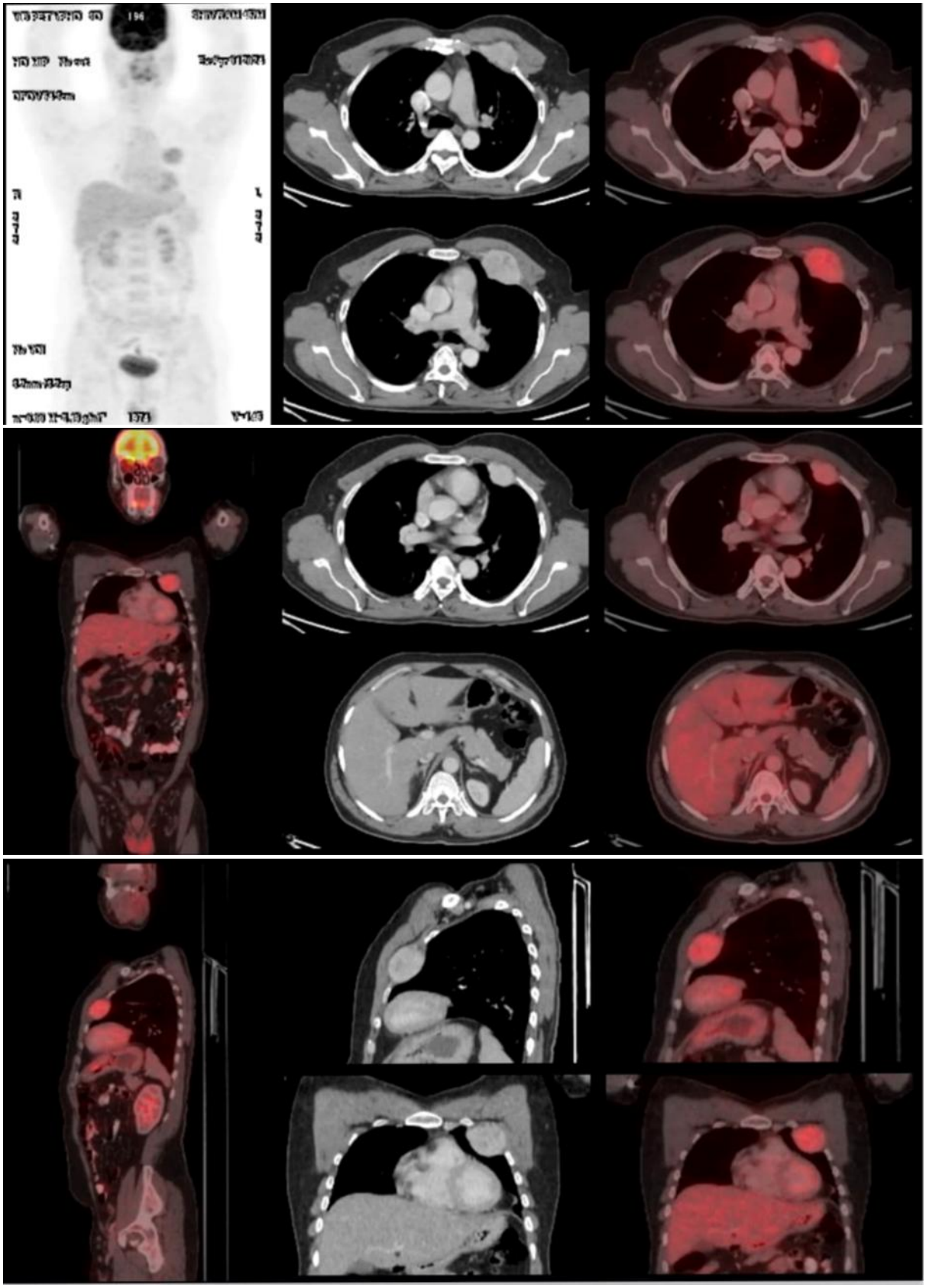

Figure 3: A positron emission tomography (PET) scan reveals a variably enhancing soft tissue mass that causes erosive changes in the left third rib and the adjacent third costochondral junction (SUVmax - 4.2; dimensions 4.9 x 3.3 x 4.3 cm). This mass is situated close to the medial ends of the left pectoralis minor and major muscles at the front, as well as the costal pleura of the left upper lung at the back, as shown in the axial and coronal view sections.

A trephine bone marrow biopsy was done based on the suggestive findings of paraproteinemia on serum protein electrophoresis (SPEP) and the presence of an enhancing lesion on the PET scan. This revealed cortical bone, with evaluable marrow spaces appearing normocellular and exhibiting a mild interstitial increase in plasma cells, which accounted for 17%. Flow cytometric immunophenotyping identified 3.5% of total plasma cells, of which 93% were clonal plasma cells. The immunophenotype details are provided in Table 2.

Table 2: Results of flowcytometric immunophenotyping showing markers positivity of plasma cells

Markers	Positive	Negative
B/Plasma cell	CD38, CD138, CD19(dim to neg), CD56, CD81 (heterogenous), CD117, cyKappa	CD27, CD200, cyLambda
T/NK cell	-	-
Myeloid	-	-
Other	CD45(dim)	-

Fluorescence in situ hybridisation (FISH) analysis identified several abnormalities. It detected three copies of 4p16 (FGFR3) and a deletion of one copy of 14q32 (IGH) in 30% of the cells. Additionally, there were three copies of chromosomes 5p and 15q and four copies of chromosome 9q. However, no abnormalities were found regarding the deletion of 1p32 (CDKN2C), the gain of 1q21, t(4;14) (p16; q32), 8q24 (MYC), 11q13 (CCND1), t(14;16) (q32; q23), or 17p13 deletion.

Based on these findings, the patient has been diagnosed with paraskeletal plasmacytoma with monoclonal gammopathy of undetermined significance (MGUS). Treatment was initiated with weekly subcutaneous bortezomib (2 mg), dexamethasone, oral cyclophosphamide, and external beam radiotherapy (EBRT), with a total planned dose of 30 Gy administered over 10 session fractions.

Discussion

Plasma cell neoplasms that manifest as mass lesions can include extramedullary plasmacytoma or paraskeletal plasmacytoma. Solitary extramedullary plasmacytomas are uncommon tumors consisting of plasma cells located in soft tissues, appearing as isolated masses that do not involve bone structures. The annual incidence of solitary extramedullary plasmacytoma is 0.1 cases per 100,000 people, primarily impacting individuals aged 50 to 80, with a median age ranging from 55 to 60 years [3].

Holler et al. conducted a literature review from 1998 to 2021, documenting 1,134 instances of extramedullary plasmacytoma (EMP) with information on tumour locations. In these cases, the majority (62.4%) were in the head and neck region, whereas the rest were in other areas of the body. Importantly, 76.9% of patients did not see any recurrence or transition to multiple myeloma (MM). On the other hand, 12.8% experienced local recurrences, and 10.2% advanced to MM. A trend toward a higher incidence was observed with radiotherapy alone MM [4].

Many extramedullary plasmacytomas evolve into multiple myeloma, with population-based studies suggesting a transition rate exceeding 70%. The median duration before progression is approximately 19 months, although this period can vary significantly, ranging from 7 to 293 months. Factors that may influence this progression include the expression profiles of certain chemokine receptors on malignant plasma cells and irregularities in cell adhesion molecules [6].

In our PubMed literature search on para skeletal plasmacytoma, we found a case by Mallik et al., who was similar to our case, a middle-aged man with chest wall mass [7]. Typically, paraskeletal plasmacytomas are located in the skull, sternum, ribs, vertebrae, and pelvis. On the other hand, extramedullary disease (EMD) may impact the skin, subcutaneous tissue, breast, liver, kidney, pleura, lymph nodes, and central nervous system [2].

Recent research into the chest wall's involvement in multiple myeloma indicates that significant clinical symptoms are chest pain and localized swelling. Thoracic CT scans frequently show soft tissue density lesions that can impact the ribcage, with a definitive diagnosis achieved through tissue biopsy, similar to what was performed in our case [8,9].

Fluorodeoxyglucose positron emission tomography (FDG-PET) is 96% sensitive and 78% specific for soft tissue involvement and can differentiate paraskeletal plasmacytomas or extramedullary disease in multiple myeloma. However, further evaluations, such as flow cytometry, serum protein electrophoresis, and bone marrow aspiration and biopsy, are necessary [10].

For patients diagnosed with paraskeletal plasmacytomas who are not candidates for immediate autologous stem cell transplantation (ASCT), the management options primarily include the Velcade (bortezomib), Revlimid (lenalidomide), and dexamethasone (VRd) regimen or the daratumumab-bortezomib-melphalan-prednisone (VMP) regimen. For those with extramedullary disease (EMD) who cannot undergo transplantation, standard management typically also involves either the VRP or VMP regimen. Furthermore, local radiation therapy should be considered for larger plasmacytomas that are causing compressive myelopathy or for local disease that persists after systemic treatment.

Conclusion

Extramedullary plasmacytoma and paraskeletal plasmacytoma are rare types of multiple myeloma that often present as unusual chest wall masses. The presence of extramedullary plasmacytoma is associated with poorer outcomes. Distinguishing paraskeletal plasmacytomas from extramedullary disease (EMD) is essential, as they have different prognostic implications and can help avoid unnecessary tests and surgeries. Healthcare professionals should remain vigilant and well-informed about both conditions to evaluate patients' prognosis effectively.

List of abbreviations

SPEP: Serum Protein Electrophoresis
SFLC: Serum Free Light Chains
Hb: Hemoglobin
TLC: Total Leukocyte Count
PET: Positron Emission Tomography
SUVmax: Maximum Standardized Uptake Value
CD: Cluster of differentiation
NK Cell: Natural killer cell
FISH: Fluorescence in Situ Hybridisation
MGUS: Monoclonal Gammopathy of Undetermined Significance
EBRT: External Beam Radiotherapy
EMP: Extramedullary Plasmacytoma
MM: Multiple Myeloma

Declarations

Ethics approval and consent to participate

Written informed consent was obtained from the patient. Ethics committee approval was not required.

Conflicts of Interest

The authors have no conflicts of interest to declare.

Funding Statement

None.

Authors' contributions

Rohit Raina, Preeti Singh Dhoat and Barath G R were involved in data collection and drafting the manuscript. Amandeep Kaur, Niket Verma, Maninder Kansal and Deepak Chaudhary were involved in reviewing, editing and finalising the manuscript.

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Supplementary Materials

None

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