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Primary Lymph Node Plasmacytoma: A Rare Entity

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ABSTRACT

Primary plasmacytomas are localized growths of clonal plasma cells that occur without evidence of systemic plasma cell disorders like multiple myeloma. They most frequently appear as solitary lesions in bone or the upper aerodigestive tract. Involvement of lymph nodes is extremely rare, accounting for approximately 2% of extramedullary plasmacytomas, and can easily be misdiagnosed as lymphoma. We present the case of a 55-year-old man presented with right side cervical swelling, which was diagnosed as plasmacytoma on lymph node biopsy. Bone marrow biopsy was unremarkable, and no monoclonal protein was detected. Additionally, there was no evidence of end-organ damage, leading to a diagnosis of primary extramedullary plasmacytoma of lymph node.

Keywords: Primary plasmacytoma; Plasma cell dyscrasia; Myeloma; Lymph nodes

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INTRODUCTION

Plasma cell neoplasms are defined by the unchecked expansion of malignant plasma cell clones. The most prevalent type is multiple myeloma (MM), which is marked by a substantial infiltration of clonal plasma cells (typically exceeding 10%) within the bone marrow, presence of monoclonal proteins in the blood and/or urine, and associated systemic features such as hypercalcemia, renal dysfunction, anemia, and bone lesions [1, 2].

Plasmacytomas, on the other hand, are localized proliferations of plasma cells that are morphologically and immunophenotypically similar to those found in MM, but without systemic or bone marrow involvement [3]. These tumors can present as solitary or multiple lesions, and may occur in various organs. Based on their location, plasmacytomas are divided into osseous plasmacytomas (arising from bone) and extramedullary plasmacytomas (EMP) (originating in soft tissues) [4].

Primary plasmacytomas are relatively rare and account for approximately 1.6–4% of all plasma cell disorders. Diagnosing solitary EMP requires histologic confirmation of monoclonal plasma cells in tissue, with less than 10% plasma cells found in bone marrow, and no signs of CRAB features (hypercalcemia, renal impairment, anemia, or bone lesions). According to the updated International Myeloma Working Group (IMWG) criteria, diagnosis is further refined by the requirement that involved free light chain levels remain below 100, and no focal lesions are seen on advanced imaging such as MRI or PET scans. The osseous variant is approximately 40% more common than EMP and often affects the axial skeleton particularly the vertebrae. In contrast, 80% of EMP cases involve the upper aerodigestive tract. Primary lymph node involvement is extremely uncommon, comprising only about 2% of EMP cases, most of which are limited to a single lymph node. Reports of diffuse lymph node involvement by plasmacytoma are exceptionally rare, with only eight cases described in the literature to date [5].

Overall, EMP generally has a more favorable prognosis compared to osseous plasmacytomas, with lower risk of transformation to MM and better progression free survival (PFS) and overall survival (OS) outcomes. Because solitary plasmacytomas—whether osseous or extramedullary—tend to be localized, local treatments such as radiotherapy (and less frequently, surgical excision) are commonly employed. This approach may also be extended to cases with limited multifocal disease, provided the lesions are accessible for targeted radiation [5].

CASE REPORT

A 55-year-old male with no significant medical history presented with a gradually enlarging, painful swelling on the right side of his neck that had persisted for 6–7 months. Clinical examination revealed a tender, subcutaneous mass located on the right lateral aspect of the neck, with overlying skin appearing unremarkable. Patient had no constitutional symptoms.

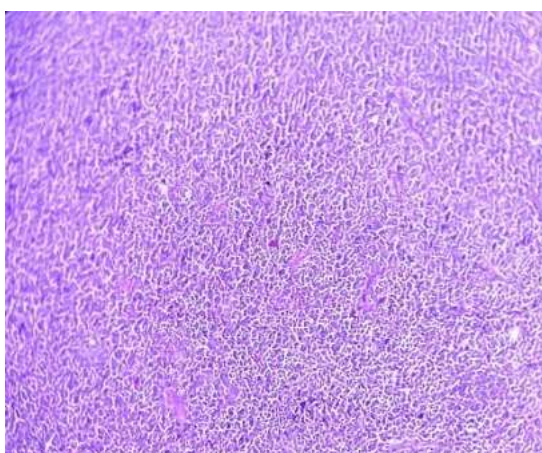
A PET-CT scan demonstrated multiple lymph nodes on the right side neck including levels I to V cervical, supraclavicular, infraclavicular, occipital, and parascapular regions—with sizes ranging from subcentimetric to enlarged. The largest node at level III measured 3.8×2.9 cm. Additionally, several lytic lesions with high FDG uptake were observed in the distal two-thirds of the right clavicle (SUVmax 16.7), left 5th rib (anterior), bilateral humeral heads, right acromion, left 6th rib, L5 vertebra, right sacral ala, right iliac blade (SUVmax 11.9), and the greater trochanter of the left femur.

The neck mass was surgically excised and submitted for histopathological examination. Grossly, it appeared encapsulated, grey-brown in color, and measured $3.5 \times 2.5 \times 1$ cm. The cut surface was grey-white to grey-brown. Microscopic examination revealed a diffuse infiltration of atypical cells that replaced the normal lymph node architecture. These cells exhibited round to oval, centrally or eccentrically located nuclei, some with prominent nucleoli, and moderate to abundant eosinophilic cytoplasm (Photomicrograph 1 and 2). Scattered residual lymphoid follicles were also seen.

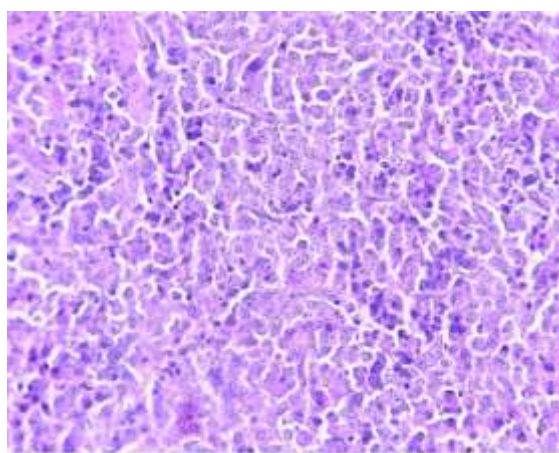
Immunohistochemical (IHC) staining showed that the neoplastic cells were positive for CD138 and CD38, confirming plasma cell lineage. The cells were negative for CD3, CD5, CD19 and CD20, ruling out T-cell and B-cell lymphomas. Light chain restriction was demonstrated by positive staining for kappa and negative for lambda. Based on the morphological features and immunophenotypic profile, a diagnosis of plasmacytoma was established.

Further workup was conducted to exclude multiple myeloma. A complete blood count showed normal parameters and bone marrow examination revealed no evidence of plasma cell infiltration or other abnormalities. Serum protein electrophoresis did not demonstrate an M-protein spike and no CRAB features were present.

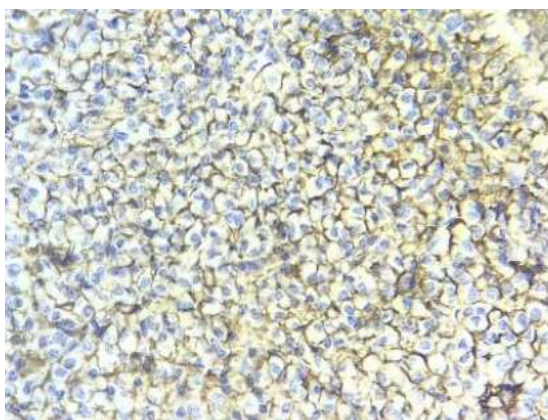
Taking into account the clinical, pathological, biochemical, and radiological findings, the patient was diagnosed with primary lymph node plasmacytoma, a rare form of extramedullary plasmacytoma.



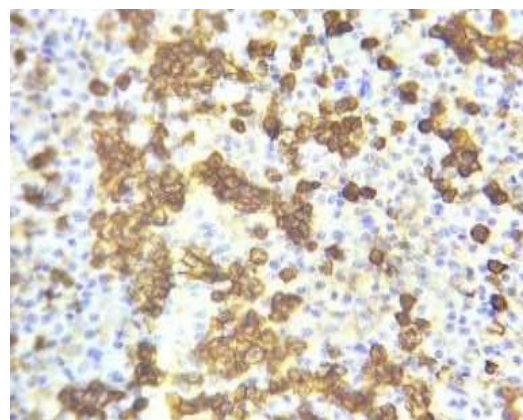
Photomicrograph 1 (H&E Stain; 40x) showing diffuse infiltration of atypical cells that replaced the normal lymph node architecture.



Photomicrograph 2 (H&E Stain; 400x) showing cells which are round to oval, centrally or eccentrically located nuclei, some with prominent nucleoli, and moderate to abundant eosinophilic cytoplasm



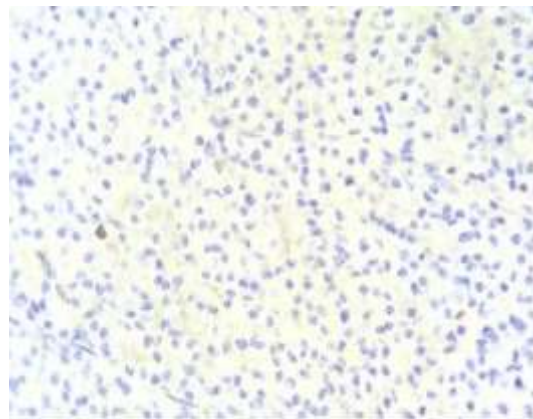
Photomicrograph 3 (IHC Stain; 400x) showing CD138 Positivity



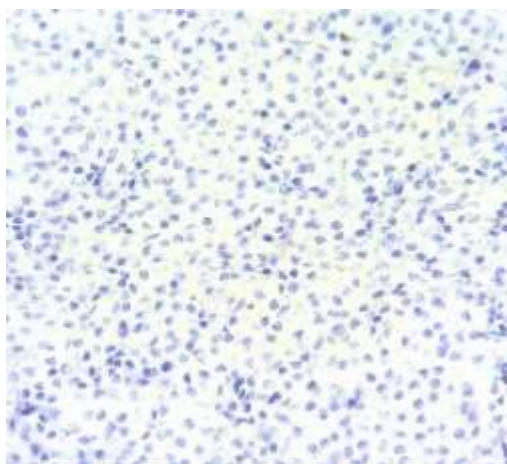
Photomicrograph 4 (IHC Stain; 400x) showing CD38 Positivity



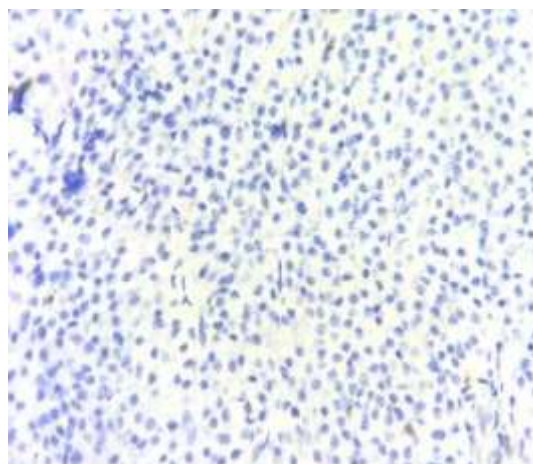
Photomicrograph 5 (IHC Stain; 400x) showing Kappa Positivity



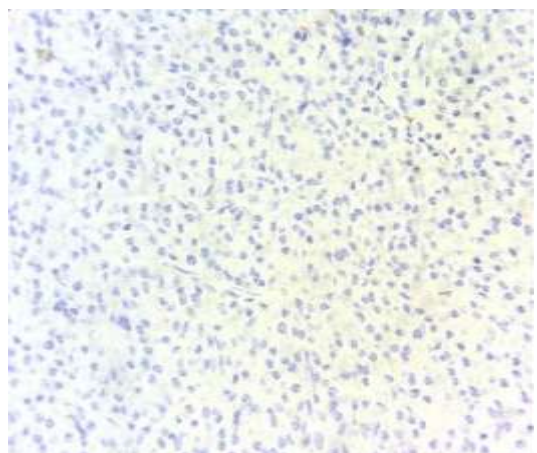
Photomicrograph 6 (IHC Stain; 400x) showing lambda negativity



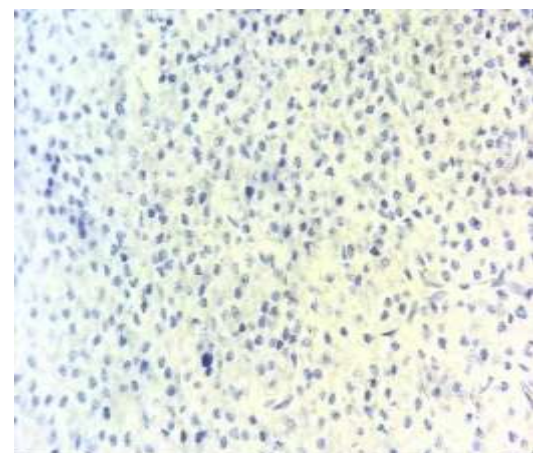
Photomicrograph 7 (IHC Stain; 400x) showing CD3 negativity



Photomicrograph 8 (IHC Stain; 400x) showing CD5 negativity



Photomicrograph 9 (IHC Stain; 400x) showing CD19 negativity



Photomicrograph 10 (IHC Stain; 400x) showing CD20 negativity

DISCUSSION

Plasmacytomas are uncommon plasma cell neoplasms characterized by the localized proliferation of monoclonal plasma cells. They are broadly categorized into primary plasmacytomas, which are confined to a localized site, and MM, which exhibits systemic involvement, including bone marrow infiltration and paraproteinemia. Distinguishing between these conditions is essential due to their markedly different prognosis and treatment approaches. Primary plasmacytomas may arise in bone (osseous plasmacytoma) or soft tissues - EMP. Osseous plasmacytomas are more prevalent and carry a higher risk of progression to MM compared to EMPs. EMPs most commonly affect the aerodigestive tract, with involvement of other locations being exceedingly rare. Among the rarest presentations are primary plasmacytomas involving multiple lymph nodes without systemic disease, which represent a minute subset of plasma cell disorders.[5]

Diagnosing primary plasmacytomas—particularly those involving multiple lymph nodes—requires thorough investigation to exclude MM. This includes bone marrow aspiration and biopsy, serum free light chain assay, PET imaging, and a full laboratory workup (e.g., complete blood count, kidney and liver function tests, urinalysis, and serum/urine protein electrophoresis with immunofixation). Immunohistochemical analysis assessing immunoglobulin heavy and light chain expression can further support the diagnosis [5]

Primary lymph node plasmacytomas may present with widespread lymphadenopathy but typically do not progress to MM, which sets them apart from other forms of EMP. Only after exhaustive testing to demonstrate the absence of bone marrow involvement, monoclonal immunoglobulins, light chains, and clinical sequelae of multiple myeloma can a diagnosis of diffuse primary plasmacytoma of lymph nodes be confirmed [5].

Differential diagnoses for plasma cell tumors of the lymph nodes include conditions such as plasma cell-type Castleman disease, lymphoplasmacytic lymphoma, follicular lymphoma, monocytoid B-cell lymphoma, and diffuse large B-cell lymphoma with plasma cell differentiation. These must be carefully ruled out [6].

Plasmablastic lymphoma (PBL) and plasmablastic myeloma (PBM) are both rare, aggressive plasma cell malignancies that can present with overlapping clinical and pathological features, complicating the diagnostic process. Both entities typically express CD38, CD138, and MUM1, show a high Ki-67 index, and lack CD20 expression. PBM may express CD56, whereas Epstein-Barr virus-encoded RNA (EBER) can be positive in PBL [7].

Morphologically, PBL is marked by a predominance of plasmablasts and/or immunoblasts with only occasional mature plasma cells, whereas PBM tends to demonstrate a greater proportion of mature plasmacytic cells. Nonetheless, these differences are often subtle, and morphology alone may be insufficient to reliably differentiate between the two, necessitating additional diagnostic tools [8].

Treatment strategies for plasmacytomas depend heavily on their location and extent. Solitary primary plasmacytomas, owing to their high radiosensitivity, are often managed successfully with localized radiotherapy and may even be cured. In contrast, treatment protocols for multifocal primary plasmacytomas are less well-defined due to their rarity. When the disease is too disseminated for localized treatment, systemic therapy becomes necessary [6].

CONCLUSION

This case underscores the exceedingly rare and often misclassified entity of primary plasmacytoma involving diffuse lymph nodes. Notably, it demonstrates that such cases can be effectively treated with systemic therapies commonly used in multiple myeloma, achieving an excellent therapeutic response.

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