Primary lymph node Plasmacytoma: Presenting as a solitary neck mass
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Abstract

Introduction: Primary plasmacytoma of lymph node is a rare neoplasm. Primary lymph node plasmacytoma represents 2% of all extramedullary plasmacytomas, and only 0.08% of all plasma cell malignant neoplasms. There have been fewer than 20 reported cases worldwide.

Case Report: We hereby report a case of primary lymph node plasmacytoma (PLNP) in a 41 year old man, who presented with a solitary subcutaneous nodule on lateral aspect of his neck, which turned out to be a primary plasmacytoma of the lymph node on histopathological examination. No evidence of the systemic involvement of plasma cell dyscrasia was discovered and thus, the diagnosis of primary lymph node plasmacytoma (PLNP) was made.

Discussion: Plasmacytoma involving lymph node is a rare entity and can occur as a manifestation of regional lymph node involvement in extramedullary plasmacytoma or even more rarely as a primary lymph node plasmacytomas (PLNPs), a complete clinical, radiological and laboratory work up needs to be done to rule out metastatic multiple myeloma and metastatic upper respiratory tract plasmacytomas.

Conclusion: Primary lymph node plasmacytomas (PLNPs) are rare malignant neoplasms representing 0.08% of all plasma cell malignant neoplasms. A diagnosis of plasmacytoma should prompt further clinical, biochemical and radiologic evaluation to determine whether the lesion is purely solitary or a localized presentation of multiple myeloma.

Key words: Lymph node, Multiple myeloma, Plasmacytoma

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Introduction

Plasmacytoma is a local tumourous collection of monoclonal plasma cells [¹]. Extramedullary plasmacytoma is a rare form of plasma cell dyscrasia and represents 1.6~4% of all plasma cell tumours [²,³]. Lymph node involvement in plasmacytomas is rare. The available literature had not more than twenty nine documented cases involving lymph nodes.
both as a primary or secondary phenomenon [4].

**Case report**

A 41 year old male patient presented with a solitary mass on one side of his neck, which was present since 3 years and gradually increasing in size. On examination, a nodular subcutaneous mass measuring around 4x3cm was situated on right lateral aspect of neck. The mass was non tender and skin over the mass was normal. Patient had no constitutional symptoms. Thorough physical examination had failed to reveal any other similar mass at other site of the body. Other groups of lymph nodes in the body were not palpable. USG neck confirmed the mass as a single enlarged lymph node. Excision biopsy of mass was done, and sent for histopathological evaluation. On gross examination, the mass was capsulated, nodular, grey brown measuring 3.5x3x2.5cm. External surface was smooth, grey brown in colour, with few dilated blood vessels. Cut section of the mass was grey brown, homogeneous with focal grey white and hemorrhagic areas [Figure1,2].

![Image](image-url)

**Figure 1:** Gross photograph of the capsulated, nodular, grey brown mass measuring 3.5x3x2.5cm with few dilated blood vessels over the external surface.
Figure 2: Cut section of the mass with homogeneous grey brown areas, focal grey white and hemorrhagic areas

Microscopic examination revealed effacement of normal architecture of lymph node by diffuse infiltrate of atypical plasma cells and plasmacytoid cells [Figure3] leaving behind only few residual atrophic lymphoid follicles. The infiltrate was seen extending into the perinodal fat. Immunohistochemistry with CD 38 and CD 138 for plasma cells showed monoclonal plasma cells in the lymph nodes. Based on the morphology, positive IHC for plasma cell markers and negative surface B-cell antigen (CD20), a histological diagnosis of plasmacytoma was rendered.

Figure 3: Microscopic photograph of lymph node with diffuse infiltrate of atypical plasma cells and plasmacytoid cells, H and E stain, high power.
Further evaluation was done with set of investigations to rule out multiple myeloma. Complete blood count, including peripheral smear examination was asked for and parameters were within normal limit. Bone marrow study and X-ray of axial skeleton revealed no evidence of plasma cell proliferation or any osteolytic lesion. Urine for Bence Jones protein was negative, serum electrophoresis did not show any monoclonal immunoglobulin. Chest X-ray, USG abdomen were unremarkable. Based on the clinical, histopathological, biochemical and radiological findings, a final diagnosis of primary lymph node plasmacytoma (PLNP) was made and sent for radiotherapy.

**Discussion**

Primary plasmacytoma of the lymph nodes is quite rare. In order to diagnose primary plasmacytoma of the lymph nodes, there must be no evidence of plasma cell proliferation elsewhere [5]. There have been fewer than 20 reported cases worldwide [6].

Extramedullary manifestations of plasma cell dyscrasias most commonly affect the upper respiratory tract, including the nasal cavity, sinuses, oropharynx, salivary glands, and larynx, followed by the gastrointestinal and urogenital tracts, skin and lung [7]. Most lymph node plasmacytomas are attributable to metastasis from multiple myeloma, or other extramedullary plasmacytomas. Extramedullary plasmacytoma represents 1.6~4% of all plasma cell tumors. Primary lymph node plasmacytoma represents 2% of all extramedullary plasmacytomas, and only 0.08% of all plasma cell malignant neoplasms [8].

PLNPs may manifest with disseminated lymphadenopathy that usually does not progress to multiple myeloma, in contrast with other types of extramedullary plasmacytomas. PLNPs can be diagnosed only after exclusion of metastatic multiple myeloma (which metastasizes to lymph nodes in up to 40% of cases of advanced-stage disease) and metastatic upper respiratory tract plasmacytomas (which represent 76% of extramedullary plasmacytomas and infiltrate cervical lymph nodes in approximately 15% of cases).

In plasma cell tumor of the lymph nodes various differentials like Castleman’s disease of the plasma cell type, lymphoplasmacytic lymphoma,
diffuse large B cell lymphoma with plasma cell differentiation, have to be considered and ruled out \[4\].

Primary lymph node plasmacytomas (PLNPs) are cured with surgical excision or radiation therapy \[9\]. Solitary extramedullary plasmacytomas are highly radiosensitive tumours. Due to the small number of patient cases and low local failure rates associated with this condition, the dose-response relationship remains unclear. Tumours of less than 5 cm in diameter have an excellent chance of being locally controlled with doses of radiation in the region of 40 Gy in 20 fractions \[7\]. The survival time associated PLNP is significantly longer than that of patients with multiple myeloma \[9\].

**Conclusion**

Primary lymph node plasmacytomas (PLNPs) are rare malignant neoplasms representing 0.08% of all plasma cell malignant neoplasms, which can manifests as an enlargement of the cervical lymph nodes. A diagnosis of plasmacytoma should prompt further clinical, biochemical and radiologic evaluation to determine whether the lesion is purely solitary or a localized presentation of multiple myeloma.

**References**