

Extramedullary plasmacytoma in the posterior triangle of neck: A rare entity

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ABSTRACT

Extramedullary plasmacytoma (EMP) is a rare plasma cell neoplasm of soft tissue without involvement of bone marrow or other systemic characteristics of multiple myeloma. It accounts for only 4% of all plasma cell tumors. We have reported herein a rare case of EMP involving soft tissues of the neck.

Key words: Multiple myeloma, neck, plasmacytoma

INTRODUCTION

Plasmacytoma may be primary or secondary to disseminated multiple myeloma. They may arise from osseous (medullary) or nonosseous (extramedullary) sites. Primary extramedullary plasmacytoma (EMP) is rare, accounting for only 4% of all plasma cell tumors, the majority (80%) of which occur in the upper aero-digestive tract. The skin and gastrointestinal tract are less common sites. It is very rare for a tumor to involve soft tissues and musculature of the neck in the posterior triangle, and only few cases has been reported in the literature. We have presented a case of an adult male who presented with an ulcerated mass in the posterior triangle of neck, which turned out to be EMP.

CASE REPORT

A 40-year-old male presented to our hospital with complaints of painless and nontender progressive swelling on the right side of the neck since past 6 months. The lesion had undergone ulceration since past 15 days. Hematological and biochemical investigations were within normal limits.

X-ray of the soft tissue of neck showed an area of opacity in the right lateral aspect of the neck, causing a bulge without adjacent bony destruction. Contrast enhanced computed tomography scan (CECT) was done to evaluate the extent and origin of the soft tissue mass. CECT scan showed a heterogeneously enhancing mass with central necrosis in the soft tissue of posterior triangle of neck, involving the skin and infiltrating into the adjacent neck and paravertebral muscles. Magnetic resonance imaging was done for further characterization. The lesion did not show intraspinal extension [Figure 1a-c]. Based on the imaging features, differential diagnoses of soft tissue sarcoma, neurofibromas, and lymphoma was suggested. The patient underwent wedge biopsy. Histopathological examination showed dense collection of plasma cells arranged in sheets in dermis and infiltrating the subcutaneous tissues, which was suggestive of soft tissue plasmacytoma [Figure 2]. Immunohistochemical study revealed that the cells were positive for Lamda, further confirming the diagnosis of plasmacytoma. Skeletal survey was also done, which did not reveal any lytic or sclerotic lesion. No M band was found on serum electrophoresis. On the basis of radiological and histopathological findings, a diagnosis of EMP was made, and the patient was treated with radiotherapy. Marked resolution of the swelling was noted with the treatment.

DISCUSSION

Plasma cell neoplasms include multiple myeloma, solitary bone plasmacytoma (SBP), soft tissue or nonosseous EMP, and plasmablastic sarcoma. The International Myeloma

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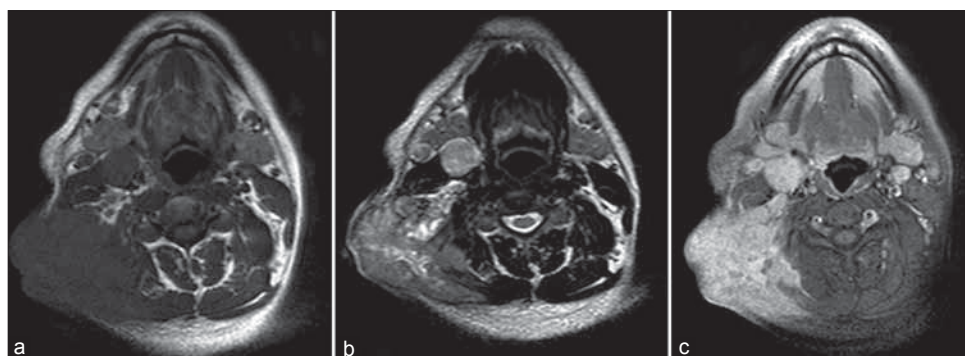


Figure 1: (a) Axial MR T1-weighted image showing the mass lesion isointense to the muscle in right posterior triangle of neck. (b) T2-weighted axial MR images showing lesion to be heterogeneously hyperintense signal. (c) Post-contrast axial MR images showing moderate enhancement with few non-enhancing necrotic areas

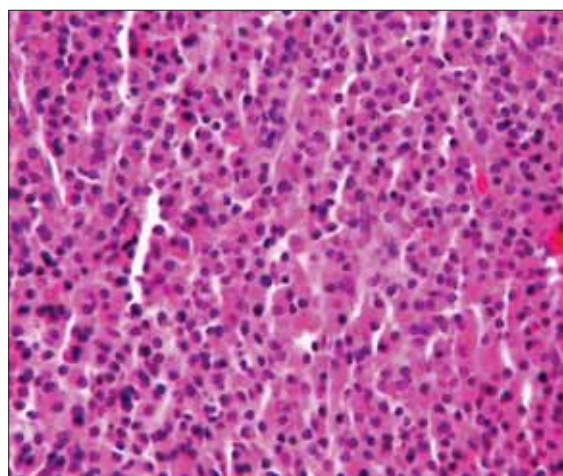


Figure 2: Histopathological examination (H and E, ×40) showing dense collection of plasma cells arranged in sheets in dermis and infiltrating the subcutaneous tissues suggestive of plasmacytoma

Working Group in 2003 has recognized a separate classification of plasmacytomas that occur as multiple sites of disease in soft tissue, bone, or both soft tissue and bone as multiple solitary plasmacytoma. Plasmacytoma (osseous or nonosseous) is distinguished from multiple myeloma by absence of hypercalcemia, renal insufficiency, and anemia, normal skeletal survey, absence of bone marrow plasmacytosis, and serum or urinary paraprotein <2 g/dl. Primary EMP can be solitary or multiple.^[1]

Primary EMP is rare, accounting for only 4% of all plasma cell tumors. It classically arises in the upper aero-digestive tract, with a predilection for the head and neck.^[1,2] Alexiou *et al.*,^[1] reviewed more than 400 published articles and found that 82.2% of EMPs occurred in the upper aerodigestive tract, with 17.8% arising in the gastrointestinal tract, urogenital tract, skin, lung, and breast, respectively. Ooi *et al.*,^[3] analyzed 21 cases of EMPs and found almost equal involvement of the head and neck, thorax, and abdomen. It is very rare for the tumor to involve soft tissues and musculature of the neck in the posterior triangle, with only few cases reported in literature.^[3,4]

Primary EMPs manifests as soft-tissue tumors with variable mass effect. Large lesions show aggressive features such as infiltration and destruction of adjacent bone, muscle, fat, and vascular encasement. On cross-sectional imaging, these tumors appear as well-demarcated, mildly heterogeneous, soft-tissue-attenuating masses, and shows moderate to marked enhancement. CT examination would also be useful in showing the underlying bony abnormality. MR offer advantages over CT on account of its high soft-tissue contrast and multiplanar depictions. These lesions are isointense on T1-weighted images, intermediate signal intensity on T2-weighted images, and moderate-to-marked contrast enhancement. However, the imaging features are not specific.^[5,6]

The distinction between different plasmacytomas is of great significance as the prognosis of multiple myeloma is poor with a mean survival of 2-3 years. The prognosis is significantly better for EMP patients than for SBP patients, with two-thirds surviving more than 10 years with currently available radiotherapy techniques.^[7,8] The risk of distant metastasis or conversion to multiple myeloma is <30%. Distant metastases tend to appear within 2-3 years of the initial diagnosis, and a close follow-up is warranted during this period.^[9] EMP carries the best prognosis of all plasma cell neoplasms with a 70% 10-year survival.^[10]

To conclude, EMP, although very rare, can arise from the soft tissues of the posterior triangle of neck and should be kept as differential diagnosis. However, due to the non-specific imaging characteristics, the role of imaging is important to determine the tumor extension, to plan the biopsy, and for follow-up of the patients.

REFERENCES

- Alexiou C, Kau RJ, Dietzfelbinger H, Kremer M, Spiess JC, Schratzenstaller B, *et al.* Extramedullary plasmacytoma: Tumor occurrence and therapeutic concepts. *Cancer* 1999;85:2305-14.
- Soesan M, Paccagnella A, Chiarion-Sileni V, Salvagno L, Fornasiero A, Sotti G, *et al.* Extramedullary plasmacytoma: Clinical behavior and response to treatment. *Ann Oncol* 1992;3:51-7.

3. Ooi GC, Chim JC, Au WY, Khong PL. Radiologic manifestations of primary solitary extramedullary and multiple solitary plasmacytomas. *AJR Am J Roentgenol* 2006;186:821-7.
4. Kaler AK, Shankar A, Jena M. Extramedullary Plasmacytoma of Soft Tissues and Gingiva. *J Allied Health* 2012;11:9. Available from: <http://www.ojhas.org/issue42/2012-2-9.htm> [Last accessed on 2012 Jul 25].
5. Chalazonitis AN, Tzovara J, Ghiatas A. Extramedullary plasmacytoma of the sphenoid sinus. *JBR-BTR* 2005;88:343.
6. Ching AS, Khoo JB, Chong VF. CT and MR imaging of solitary extramedullary plasmacytoma of the nasal tract. *AJNR Am J Neuroradiol* 2002;23:1632-6.
7. Galièni P, Cavo M, Pulsoni A, Avvisati G, Bigazzi C, Neri S, *et al.* Clinical outcome of extramedullary plasmacytoma. *Haematologica* 2000;85:47-51.
8. Miller FR, Lavertu P, Wanamaker JR, Bonafede J, Wood BG. Plasmacytomas of the head and neck. *Otolaryngol Head Neck Surg* 1998;119:614-8.
9. Mayr NA, Wen BC, Hussey DH, Burns CP, Staples JJ, Doornbos JF, *et al.* The role of radiation therapy in the treatment of solitary plasmacytomas. *Radiother Oncol* 1990;17:293-303.
10. Iseri M, Ozturk M, Ulubil SA. Synchronous presentation of extramedullary plasmacytoma in the nasopharynx and the larynx. *Ear Nose Throat J* 2009;88:E9-12.

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