



Case Report

Extramedullary plasmacytoma of larynx: A report of an interesting case

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ABSTRACT

Extramedullary plasmacytoma (EMP) is neoplastic proliferation of monoclonal plasma cells outside the bone marrow. Only 3% of plasma cell neoplasms occur at extramedullary site. Majority of cases occur in head and neck region and very few cases are reported in larynx. We herein report a case of extramedullary plasmacytoma involving the left vocal cord and diagnosed after histopathology and immunohistochemistry (IHC) study.

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1. Introduction

Extramedullary plasmacytomas (EMP) are localized neoplastic proliferation of monoclonal plasma cells arising in tissues other than bone marrow.¹ They are very rare, contributing about 3% of plasma cell neoplasms and majority of cases (about 80%) occur in head and neck region. They account for about 1% of head and neck tumours.²⁻⁴ Most head and neck EMPs occur in sinonasal region and rarely seen in larynx. They account for 0.04 to 0.19% of malignant laryngeal tumours.²⁻⁴ As per our knowledge, total 153 cases of EMP of larynx are reported in the English and Chinese literature and this is 154th case of EMP of larynx. In Indian literature, 8 cases have been reported and this is 9th case of EMP of larynx.^{2,5-11}

2. Case Report

41year old male, presented with history of change in voice and difficulty in swallowing since 8 months. He was a chronic tobacco chewer and smoker. There was no history of other major medical illness. On examination during

laryngoscopy, a polypoidal mass was noticed on left vocal cord which was diagnosed clinically as laryngeal polyp. Biopsy was performed and submitted to histopathology department for a definitive diagnosis. Grossly, we received a polypoidal mass measuring 0.9 x 0.7 x 0.6 cm.

Microscopy revealed a polypoidal mass lined by stratified squamous epithelium which was focally ulcerated with subepithelial tissue showing tumour composed of uniform small round cells having small round nuclei with irregularly condensed chromatin and eosinophilic cytoplasm arranged in clusters and sheets. (Figure 1). On histopathology, it was reported as a small round cell tumour with a differential diagnosis of paraganglioma, vascular tumour and glomus tumour. Immunohistochemistry (IHC) was advised for a definitive diagnosis.

Accordingly, the primary IHC panel included broad spectrum cytokeratin (CK), Synaptophysin, Chromogranin A, CD34, CD31, Fli 1, Calponin and Smooth muscle actin (SMA). The tumour cells expressed CD31 and were negative for Synaptophysin, Chromogranin, CD34, Fli1, Calponin and SMA. As CD31 was positive and other vascular markers like CD34 and Fli1 were negative, we reviewed the morphology again. Considering the small size and condensed nuclear chromatin with occasional plas-

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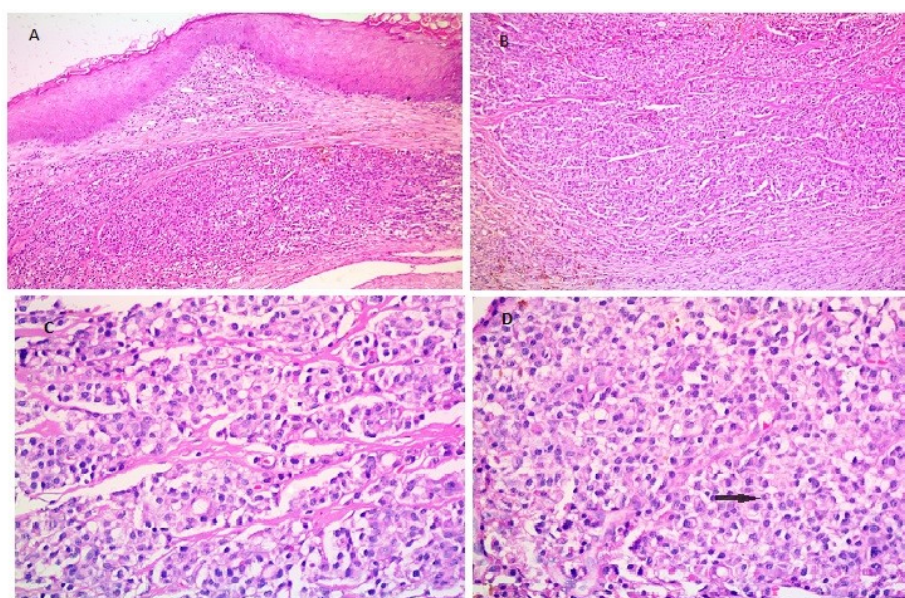


Fig. 1: **A):** Laryngeal polyp biopsy showing a submucosal mass with small round cells arranged in nesting pattern (H&E, 4x); **B, C):** Submucosal small round cells with clumped nuclear chromatin arranged in nests (10x and 40x respectively); **D):** Arrow pointing towards cells having plasmacytoid morphology exhibiting eccentrically placed nuclei with clumped nuclear chromatin and eosinophilic cytoplasm, (40x).

macytoid morphology; we included Epithelial membrane antigen (EMA), CD38, CD138, MUM1, Kappa and Lambda in the 2nd panel of IHC. The tumour cells expressed CD38, CD138, EMA and showed Lambda light chain restriction. Rest of the markers were negative. Mib-1 proliferation index was about 5%. (Figures 2 & 3). Thus, finally we labelled the case as plasmacytoma of larynx. On further workup, the bone marrow examination, whole body CT scan and serum electrophoresis were normal. Hence, we finally concluded it as extramedullary plasmacytoma (EMP) of larynx. After surgical excision, patient was treated with radiotherapy and is disease free after 6 months of follow up.

3. Discussion

Solitary plasmacytoma is an extremely rare form of haematological malignancy, which is classified as solitary bone plasmacytoma or solitary extramedullary plasmacytoma. EMP is defined as solitary soft tissue lesion with monoclonal plasma cells, absence of bone marrow involvement, and absence of end organ damages secondary to plasma cell proliferative disorder.¹² It is an immunoproliferative, monoclonal disease of the B-cells. It originates as an imitation of malignant transformed plasma cells. They tend to migrate and return in the bone marrow. Rarely, they settle in soft tissue or extracellular connective tissue area. This is the origin of monoclonal plasma cell foci located outside the bone marrow.⁵ The common age group for presentation is 50- 70 yrs. It occurs more commonly in males than females with a ratio of 3:1.^{5,13} EMPs are reported at

various sites such as aero digestive tract, soft tissue, lymph node, breast, bladder, thyroid, testes, parotid gland, skin and central nervous system.^{1,6} About 80% occur in head and neck region with most cases involving nasal cavity, paranasal sinuses and nasopharynx and very rarely seen in larynx.⁶ In larynx they are commonly seen at epiglottis, vocal cord, ventricular bands, arytenoids and the subglottic space. Clinical symptoms are closely related to location, size of tumour and laryngeal impairment. The common symptoms are difficulty in swallowing, change in voice and sometimes acute airway obstruction etc.^{2,12}

The diagnosis of EMP is made after excluding multiple myeloma with negative X-ray and/ or magnetic resonance imaging of bones and with normal bone marrow biopsy. There should be no signs of serum or urine monoclonal proteins, anaemia, hypercalcemia or renal impairment.⁴ EMP is diagnosed on the basis of characteristic histological features of diffuse or nodular proliferation of small round cells with eccentric nuclei having coarse chromatin and abundant slightly basophilic cytoplasm.^{1,3} The differential diagnosis includes tumours with small round cell morphology like lymphomas, neuroendocrine tumours, rhabdomyosarcoma, neuroblastoma along with melanoma, undifferentiated carcinomas and benign lesions like plasma cell granuloma and inflammatory lesions.^{3,7} Immunohistochemistry is required for a definitive diagnosis by ruling out the differentials, if there is any diagnostic dilemma on haematoxylin and eosin morphology. The EMP express CD138, CD 38, CD56, EMA and also exhibits either Kappa or lambda light chain restriction.^{1,7,8,11}

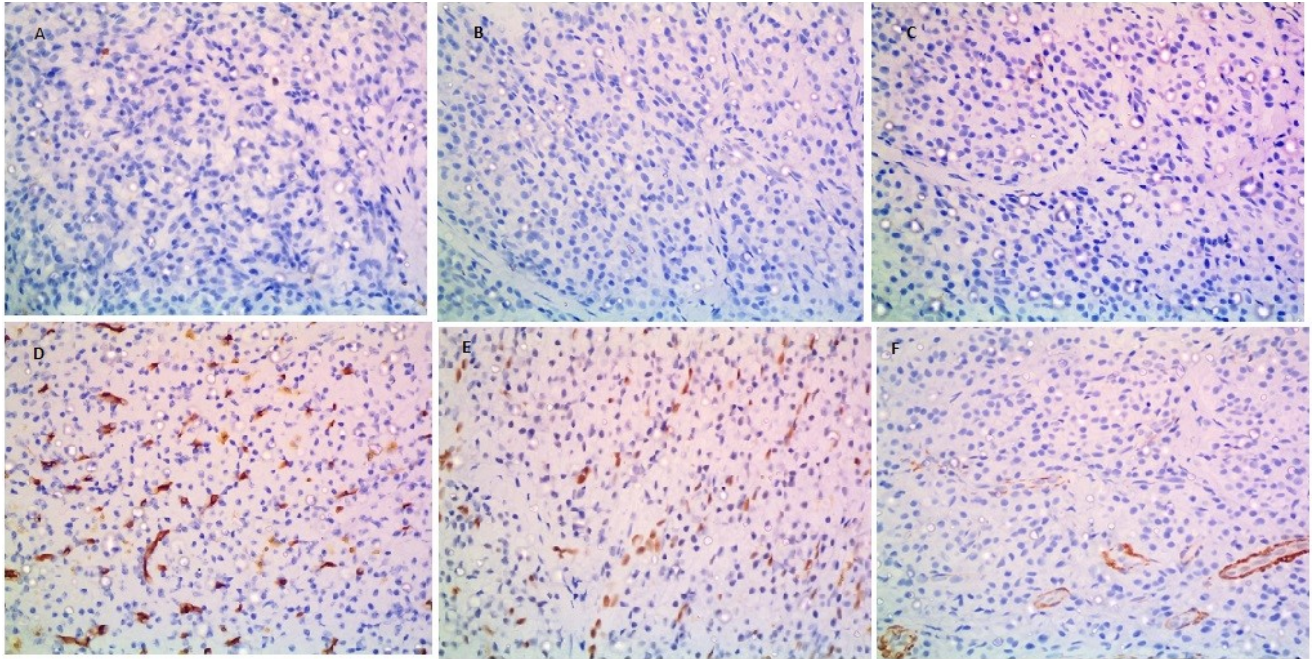


Fig. 2: Immunohistochemistry (IHC) markers. Tumour cells were negative for **A):** CK, **B):** Synaptophysin; **C):** Chromogranin; **D):** CD34, **E)** FLI-1, **F)** Calponin (40x)

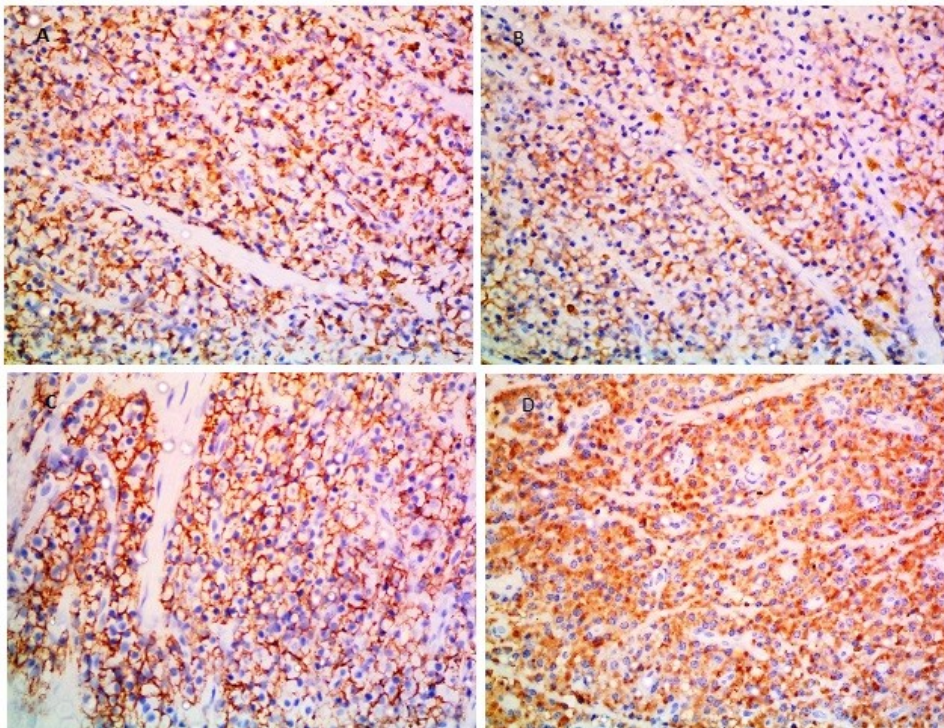


Fig. 3: – Immunohistochemistry (IHC) markers. Tumour cells expressed **A):** CD31; **B):** CD 38; **C):** CD138 and; **D):** Showed lamda light chain restriction

EMP is radiosensitive. The prognosis is favourable after radiation therapy with doses of 40-50 Gy. The local disease control rates are 80-100%. Other treatment modalities include the surgical resection, laser excision or combined therapy.^{3-5,12} EMP in head and neck region carries better prognosis than the other site, regardless of tumour grade or stage.^{1,12} The prognosis of EMP is relatively better than solitary plasmacytoma of bone or multiple myeloma (MM). The follow up is recommended since some cases of conversion to multiple myeloma after 15 years of initial diagnosis with EMP are found.^{3,12} Large size, presence of M- protein and light chain, amyloid deposit and high nuclear grade are poor prognostic factors.¹¹

Our patient was treated with surgery and radiotherapy and is disease free at 6 months of follow up.

4. Conclusion

EMP in larynx is an extremely rare tumour and can be misdiagnosed as it mimics many other tumours like vascular tumours, neuroendocrine tumours and other small round cell tumours. We must consider EMP in the list of differential diagnosis of tumours with small round cell morphology as treatment modalities and prognosis differs significantly. High degree of suspicion and immunohistochemistry is helpful in diagnosis of this entity.

5. Conflict of Interest

None.

6. Source of Funding

None.

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