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Case Report

Ear, Nose and Throat Section

Solitary Extramedullary Plasmacytoma of the Thyroid Cartilage: A Case Report

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ABSTRACT

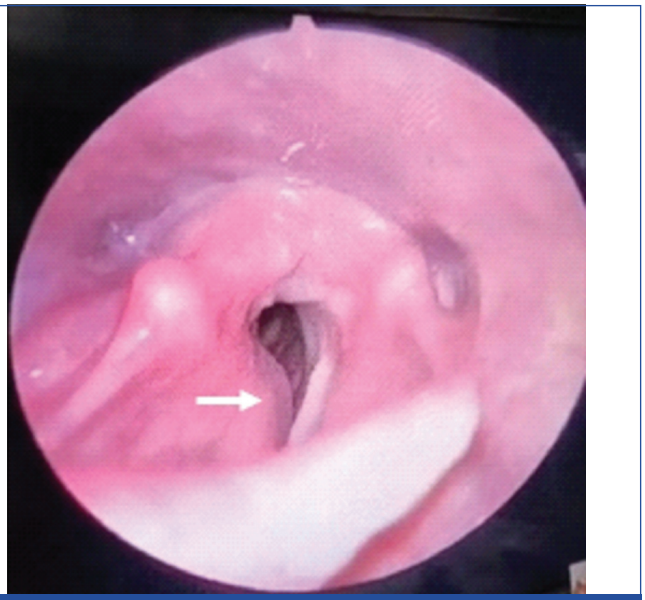
Extramedullary Plasmacytoma (EMP) belongs to the category of haematolymphoid neoplasms of the larynx. This is a case report of a 51-year-old female patient who presented to the Ear, Nose and Throat (ENT) department with a change in voice and a swelling in the neck. On video laryngoscopy, a smooth bulge was observed arising from the right ventricle towards the midline. Magnetic Resonance Imaging (MRI) of the neck showed a large, relatively well-defined T1 hypointense, T2 and T2 FS hyperintense, mildly enhancing mass centred in the right lamina of the thyroid cartilage. Histopathology reported plasmacytoma with CD138 positivity and lambda restriction. Although EMP in the supraglottic region has been reported, plasma cell neoplasm involving the thyroid cartilage is extremely rare. Reaching such a rare diagnosis requires a collaborative effort, where each clinical and diagnostic finding plays a crucial role in achieving accuracy.

Keywords: Multiple myeloma, Plasma cell neoplasm, Tumours of larynx

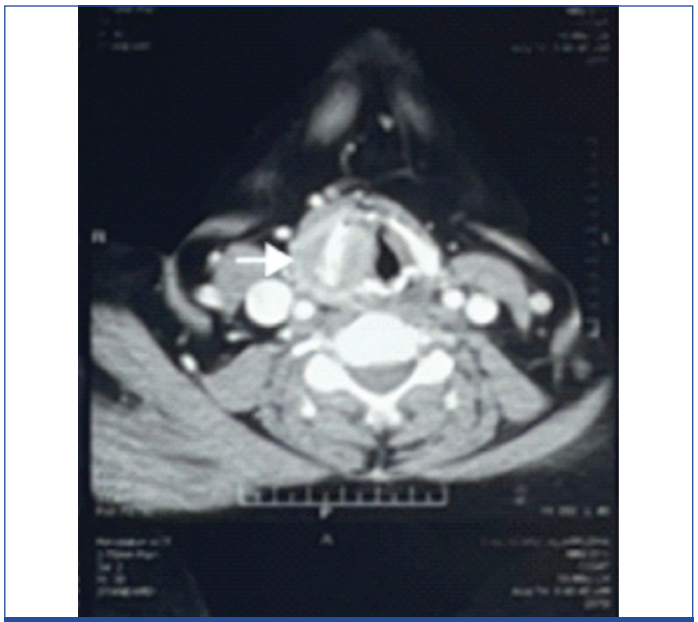
CASE REPORT

A 51-year-old female homemaker presented to the ENT department with a persistent change in voice for the past one and a half months, which was present throughout the day. She also complained of a swelling in the neck, which she had noticed one month earlier. She had no co-morbidities, no previous surgeries, and no history of radiation exposure. There was no family history of malignancy or thyroid disease. On examination of the neck, there was a swelling measuring 3×2 cm at the level of the right lamina of the thyroid cartilage. It was firm, immobile, and had regular borders. The swelling moved with deglutition. Routine blood investigations were within normal limits, and thyroid function tests (T3, T4, TSH) were also normal.

Video laryngoscopy showed a smooth bulge arising from the right ventricle towards the midline [Table/Fig-1]. Ultrasonography (USG) of the neck showed a well-defined hypoechoic lesion measuring 3 cm × 2.4 cm × 3.3 cm involving the right lamina of the thyroid cartilage, with extension on either side of the lamina and anteriorly abutting the strap muscles. Contrast-enhanced Computed Tomography (CECT) of the neck and thorax confirmed the USG findings. CECT also showed destruction of the right lamina of the thyroid cartilage [Table/Fig-2]. MRI revealed a T1 hypointense and T2/T2 FS hyperintense lesion [Table/Fig-3].



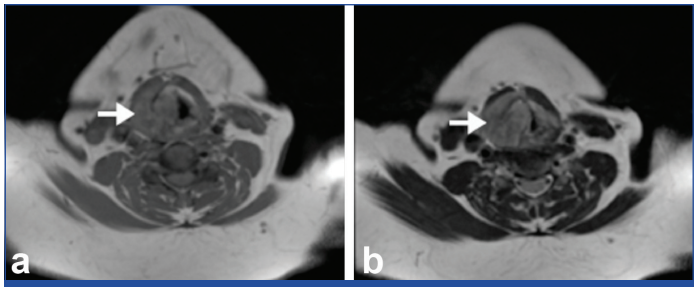
[Table/Fig-1]: Video laryngoscopy showed a smooth bulge arising from the right ventricle crossing midline (white arrow). Right vocal cord could not be visualised. Left vocal cord normal and mobile.



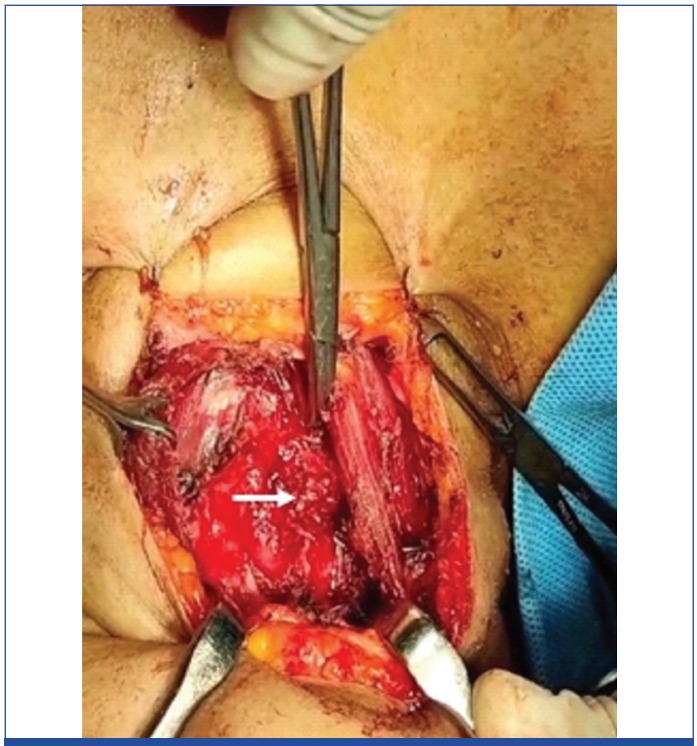
[Table/Fig-2]: Contrast Enhanced CT (CECT) of infrahyoid neck at the level of thyroid cartilage shows a near-homogeneously enhancing, soft-tissue density lesion (white arrow) centred within the lamina of the right thyroid cartilage.

A biopsy of the thyroid cartilage lesion was taken through a neck exploration procedure [Table/Fig-4]. Histopathology showed fragments of fibrocollagenous tissue with diffuse infiltration by plasmacytoid cells having abundant eosinophilic cytoplasm and eccentrically placed round nuclei [Table/Fig-5a]. Immunohistochemistry (IHC) showed CD138 positivity and lambda light-chain restriction [Table/Fig-5b].

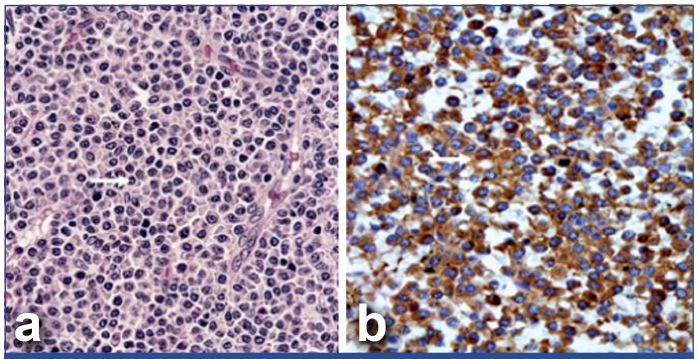
Further investigations were conducted to rule out Multiple Myeloma (MM). Complete blood count and peripheral smear were within normal limits. Beta-2 microglobulin was elevated at 6.8 mg/L. Serum protein electrophoresis was normal, and Bence Jones protein was not detected. A skeletal survey revealed no lytic lesions. Bone



[Table/Fig-3]: T1-weighted (a) and T2-weighted (b) MR images of the neck at the infrahyoid level show a lesion centered in the lamina of the right thyroid cartilage, appearing hypointense on T1WI (white arrow) and mildly hyperintense on T2WI (white arrow). The lesion demonstrated non homogeneous post-contrast enhancement (not shown).



[Table/Fig-4]: Friable and vascular lesion involving the right lamina of thyroid cartilage (white arrow).



[Table/Fig-5]: Microscopic picture of plasmacytoma; a) Fibrocollagenous tissue with diffuse infiltration by plasmacytoid with abundant eosinophilic cytoplasm and eccentrically placed nucleus (H&E; 400x); b) Immunostaining for CD138 shows positive staining in plasmacytoma (CD138; 400x).

marrow aspiration was also negative. In the absence of systemic involvement, the patient was diagnosed with EMP.

She was treated with targeted radiotherapy (IMRT) for two months. A total dose of 50 Gy was administered in 2 Gy fractions. She is currently on follow-up with regular blood tests and video laryngoscopy every two months. A follow-up CT scan performed after one year did not show any new lesions.

DISCUSSION

The EMP constitutes less than 5% of plasma cell neoplasms, of which 80% occur in the head and neck region. The most commonly involved site in the head and neck is the pharynx, followed by the nasal cavity. Laryngeal involvement is extremely rare and accounts for approximately 5% of cases [1]. The most common symptom of laryngeal EMP is slowly progressive hoarseness [1]. Plasma cell neoplasms are characterised by neoplastic proliferation of a single clone of plasma cells producing monoclonal immunoglobulins [2]. A neoplasm of plasma cells usually involves the bone marrow diffusely and is termed Multiple Myeloma (MM). A single lesion localised to the bone marrow is referred to as medullary plasmacytoma, and when localised to the bone alone, it is referred to as Solitary Bone Plasmacytoma (SBP). Soft-tissue involvement without bone marrow infiltration is termed Extramedullary Plasmacytoma (EMP) [3]. Solitary EMP is a single soft-tissue lesion composed of clonal plasma cells without associated bone marrow involvement or end-organ damage [3]. Although the aetiology of EMP is unknown, viral pathogenesis—particularly Epstein-Barr Virus (EBV)—has been implicated [4].

In the larynx, the most commonly involved site is the supraglottis. Involvement of the vocal folds and subglottis is rare. The order of frequency of laryngeal involvement is: epiglottis > ventricles > vocal cords > false cords > aryepiglottic fold > arytenoid > subglottic region [5]. The larynx is involved in 5-20% of EMP cases, but involvement of the cartilage is rare. Two theories have been proposed to explain cartilage involvement: direct invasion from an adjacent plasmacytoma, or metaplasia of cartilage into bone with the formation of a marrow cavity in which a plasmacytoma can originate [6].

Diagnosis of EMP is based on IHC and exclusion of systemic plasma cell proliferative diseases [7]. Radiological imaging such as CT and MRI is useful to evaluate the spread of EMP to the spine, pelvis, femur, and humerus. Positron Emission Tomography (PET) scan is also useful prior to the final diagnosis of EMP [7]. Additional systemic workup, including bone marrow biopsy and serum/urine protein electrophoresis, is performed to rule out MM [7].

Differential diagnoses for a submucosal laryngeal lesion include chondrosarcoma, liposarcoma, paraganglioma, and neuroendocrine tumours. Chondrosarcoma and neuroendocrine tumours appear as heterogeneously enhancing lesions on contrast-enhanced CT, whereas EMP typically shows homogeneous enhancement. Liposarcoma can be distinguished by the presence of intratumoral fat-attenuating areas on CT [1].

EMP is radiosensitive, with favourable outcomes following treatment with radiation doses of 40-50 Gy [8]. Other treatment options include surgical resection, laser excision, or combined modality therapy. Progression to MM is less common in EMP compared to SBP [3]. The optimal radiotherapy dosage regimen remains debated. Mill WB and Griffith R, recommend 5,500-6,000 cGy over 6-7 weeks [9].

Prognosis is significantly worse for patients presenting with cervical lymphadenopathy or multifocal disease, with more than 40% experiencing relapse or metastasis during follow-up [10]. There is a risk of progression to MM, with approximately 6% of patients in large series developing MM—most commonly within the first two years, though progression may occur much later [11]. Rarely, secondary therapy-related myeloid neoplasms, such as acute myeloid leukaemia, have been reported following treatment.

Current evidence is limited due to the rarity of the disease, reliance on case reports and small case series, and short follow-up durations, which may underestimate late relapse or progression to MM [10,11]. There is no consensus on the optimal treatment modality, as randomised controlled trials are lacking. The choice between radiotherapy and surgery is often individualised based on tumour size, location, and patient factors [10,11]. Lifelong follow-up is essential due to the risk of late recurrence and transformation into systemic disease [11].

MM has a poor prognosis, with a 5-year survival rate of 18% and a mean survival of 2-3 years [12]. MM develops in 20-30% of patients with SBP and in 50-60% of those with EMP after the initial diagnosis [12]. EMP has a better prognosis than MM; approximately 66% of patients with EMP survive for five years, compared with only 18% of patients with MM [13].

Plasmacytoma of the head and neck region has been reported to have a better prognosis than those occurring at other sites. This may be due to early presentation, when the lesions are small and well localised [14]. Prognosis is influenced by tumour location, cartilage and bone destruction, and regional lymph node involvement [15].

CONCLUSION(S)

Although EMP of the larynx is rare, it should be considered in the differential diagnosis of laryngeal tumours. As it is highly radiosensitive, radiotherapy is an effective treatment modality. Despite its comparatively good prognosis relative to SBP or MM, regular follow-up is essential due to the long-term risk of progression to MM.

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