Extramedullary plasmacytomas of the larynx and parapharyngeal space: Imaging and pathologic features

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Abstract
Extramedullary plasmacytoma is a rare plasma cell neoplasm that can occur in the head and neck. In this article we describe a case of multiple synchronous extramedullary plasmacytomas involving the upper airway, pharynx, and larynx. The clinical, imaging, and pathologic features of this neoplasm are discussed, together with potential treatment options.

Introduction
Extramedullary plasmacytomas are rare neoplasms that occur in soft tissues at single and multiple sites. Extramedullary plasmacytomas occur in the upper aerodigestive tract and oral cavity. In this report we describe the clinical, imaging, and pathologic features and report a case in which multiple lesions involved the upper airway and pharynx.

Case report
A 71-year-old man presented with a 6-month history of a hoarse voice. He had no other symptoms, and the initial clinical examination was unremarkable. Microlaryngoscopy revealed a mucosal nodule on the soft palate with several other nodules present in the left and right supraglottic regions (figure 1).

Biopsies were taken from a left supraglottic nodule, which demonstrated the replacement of the submucosal stroma with dense sheets of plasma cells. On immunohistochemical staining, these cells showed kappa immunoglobulin light-chain restriction consistent with plasmacytoma (figure 2).

Blood test results were normal apart from a 4g/L IgG kappa paraprotein in the serum with no immunoparesis. Magnetic resonance imaging (MRI) of the neck (figure 3) confirmed the presence of supraglottic nodules. An additional 3-cm soft-tissue nodule in the right parapharyngeal space was also assumed to be a plasmacytoma. Further investigations—including Bence-Jones protein urinalysis, skeletal survey, bone marrow aspiration, and trephine biopsy, as well as spinal MRI—revealed no evidence of myeloma.

The patient underwent laser excision of the soft palate and supraglottic nodules, with subsequent radiotherapy to the neck to which he responded well. At 2 years post-treatment, he showed no evidence of disease recurrence.

Discussion
Dalrymple and Bence Jones first described plasma cell neoplasms in 1846. These can be classified into 3 main categories:

- multiple myeloma
- extramedullary plasmacytoma of soft tissues
- solitary plasma cell tumors of bone

The overall incidence of all plasma cell tumors is 3:100,000/year; multiple myeloma is the most common form. Plasma cell tumors represent less than 1% of all malignant tumors of the head and neck; of these, extramedullary plasmacytomas comprise less than 4%. Extramedullary plasmacytomas are usually found in the upper aerodigestive tract and oral cavity. Common sites include the nasal cavity, nasopharynx, paranasal sinuses, and larynx. The most common laryngeal sites are the epiglottis, vocal folds, false vocal folds, ventricles, and subglottis, in descending order or frequency. Extramedullary plasmacytomas have also been reported to occur in the lungs, gastrointestinal tract, liver, and brain.
Extramedullary plasmacytomas of the larynx are generally submucosal but may be polypoid and may involve single or multiple contiguous sites. It is believed that extramedullary plasmacytomas are nondisseminating but locally aggressive; 10 to 20% metastasize to cervical lymph nodes. Approximately one-third of extramedullary plasmacytomas disseminate to multiple myeloma within 2 years of presentation.

The typical patient presenting with a plasma cell tumor is male and 50 to 60 years of age. Common symptoms include hoarseness, stridor, hemoptysis, and dysphagia, depending on tumor size and position at presentation.

The diagnosis of extramedullary plasmacytoma is made with biopsy. If the biopsy findings are consistent with plasma cell tumor, further investigations are required to exclude multiple myeloma. These would include: (1) no histologic evidence of plasmacytoma/plasmacytosis in bone marrow; (2) negative blood investigations and no clinical signs and symptoms of myeloma (e.g., bone pain, anemia, renal failure); and (3) no urine monoclonal gammopathy (Bence-Jones proteins) or osteolytic lesions on plain films.

Occasionally, extramedullary plasmacytoma may be associated with a small monoclonal gammopathy (<0.03g/L), which usually disappears after treatment. Persistent monoclonal gammopathy can be interpreted as possibly suggestive of disease recurrence or dissemination to myeloma.

In extramedullary plasmacytoma, immunohistochemical staining of tissue biopsies shows monoclonal immunoglobulin. Congo red stain is often positive for amyloid as a result of free light-chain deposition in soft tissue.

Staging of extramedullary plasmacytoma can be defined as:

- Confirming disease is confined to one site (stage 1)
- Excluding local extension of tumor or lymph node involvement (stage 2)
- Excluding metastatic spread (stage 3)

Computed tomography and MRI can both be used to further evaluate local extension of soft-tissue lesions and can demonstrate the presence of additional, clinically occult, lesions or cervical node involvement.
The treatment for extramedullary plasmacytoma is surgery, radiotherapy, or a combination of both. Plasmacytomas are generally very radiosensitive. The radiotherapy field should include cervical lymph nodes because of the risk of local/regional spread. Surgical removal is effective for localized tumors in positions associated with a low risk of intraoperative damage or proximity to vital structures. It should be noted that even with adequate treatment, there is a 6 to 10% local recurrence rate. Chemotherapy can be used in cases of locally advanced, recurrent, or disseminated disease. Therefore, follow-up is important, especially in younger patients for whom there is a higher long-term risk of dissemination to malignant myeloma.

References