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Extramedullary plasmacytoma of the larynx presenting with upper airway obstruction in a patient with long-standing IgD myeloma

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Abstract
A rare case of upper airway obstruction due to an extramedullary plasmacytoma of the larynx in a patient with long-standing IgD myeloma is presented. Reports of patients with extramedullary plasmacytomas eventually developing multiple myeloma are common, however, the converse appears to be an extremely rare event. Attention is drawn to the problem of acquiring adequate tissue for diagnostic purposes as well as the use of immunohistochemical staining techniques. The patient required an urgent tracheostomy and was treated with radiotherapy.

Key words: Airway Obstruction; Laryngeal Neoplasms; Pathology; Plasmacytoma

Introduction

Extramedullary plasmacytomas (EMP) are rare soft tissue tumours arising from the malignant proliferation of plasma cells. They primarily occur in the head and neck region, usually presenting as soft tissue masses in the nasopharynx or paranasal sinuses.[1] Laryngeal EMPs have been reported in the literature, but EMPs rapidly leading to upper airway obstruction are an extremely rare phenomenon.

There are multiple reports in the literature of EMPs progressing to multiple myeloma, however the converse is very rarely reported.

The authors present a unique case of a patient with long-standing IgD myeloma presenting with a gross neck swelling and upper airway obstruction due to an EMP of the larynx. The case has a number of unique features and to the best of our knowledge is the first of its kind.

Case report

A 54-year-old man was referred to the otolaryngology department with a history of a left-neck swelling. The onset of the swelling coincided with the insertion of a dialysis permacath into the left internal jugular vein. The swelling was initially thought to be a haematoma, but it subsequently increased in size over six weeks.

The patient had originally presented with end stage renal failure and IgD myeloma some three years previously. The myeloma had been treated with chemotherapy in the form of melphalan and cyclophosphamide.

On examination, the patient had inspiratory stridor. A huge soft tissue mass was evident involving the entire left side of the neck. Flexible nasoendoscopy revealed a large tumour involving the left hemilarynx. The mass was subjected to fine needle aspiration cytology (FNAC), however the samples were inadequate for diagnostic purposes.

Computerized tomography (CT) of the neck revealed a huge left-sided transglottic laryngeal tumour with involvement of the piriform fossa and extension posteriorly to the midline below the level of the cricoid. There was almost complete obliteration of the airway at the level of the true vocal folds. Massive enlargement of the left cervical chain of lymph nodes was noted (Figure 1).

An urgent tracheostomy was performed under local anaesthetic. Direct laryngoscopy revealed a large smooth left supraglottic tumour obscuring the left vocal fold and extending into the adjacent piriform fossa; Multiple biopsies were taken.

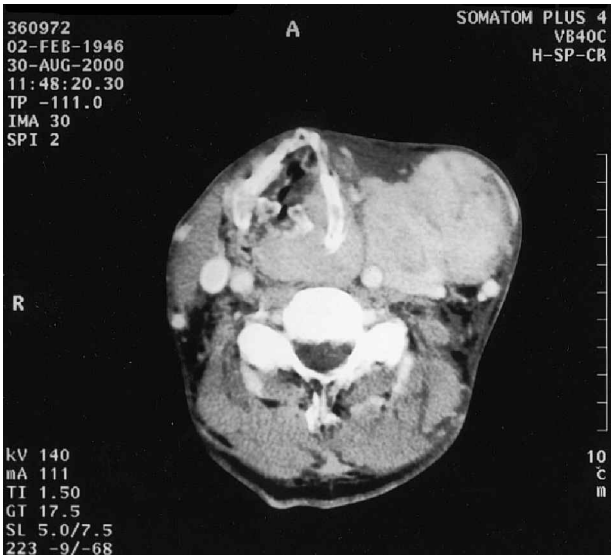

Fig. 1
Axial CT scan of neck showing a large left laryngeal tumour and massive cervical lymphadenopathy.

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Histological examination of the biopsies confirmed an EMP. The patient was treated with radiotherapy. The response to radiotherapy was good with a significant reduction in the size of the mass. However, he succumbed to medical complications within weeks of treatment.

Discussion

Multiple myeloma (MM), medullary plasmacytoma (solitary plasmacytoma of bone) and EMPs are all tumours occurring as a result of the malignant proliferation of plasma cells. Multiple myeloma represents the diffuse form of the disease, it is characterized by the disseminated proliferation of malignant plasma cells with involvement of multiple organ systems. It is the most common of the three and is essentially incurable. Medullary plasmacytoma may be solitary or multiple, they most commonly occur in the longbones, spine and pelvis. EMPs may also be solitary or multiple, and occur in submucosal tissues. They represent less than 10 per cent of all plasma cell malignancies and approximately 80 per cent occur in the head and neck region.[2] However they account for less than one per cent of all head and neck malignancies.[3]

The most common sites for EMPs in the head and neck are the paranasal sinuses, the nasal cavity and the oral cavity.[4] Approximately six to 18 per cent of all EMPs are found in the larynx.[5] Within the larynx the most frequently involved sites are the epiglottis, ventricles, vocal folds and ventricular folds.[6]

Hoarseness remains the most common presenting symptom,[4] stridor and dysphagia are late features. Pain is an uncommon symptom unless secondary infection or bone erosion occurs.[7] Acute airway obstruction requiring intubation is an extremely rare event and may represent haemorrhage within the tumour or secondary bacterial infection.[8]

The diagnosis of EMP relies on examination of pathological specimens with immunohistochemical staining techniques. In cases where there is no previous history of plasma cell malignancy it is important to exclude disseminated disease in the form of multiple myeloma, this should include a skeletal X-ray survey and bone marrow biopsy.

Most lesions are accessible for FNAC, however as in this case, FNAC often fails to yield tissue samples adequate to make a definitive diagnosis. FNAC can, however be useful in excluding other pathologies such as squamous cell carcinoma. For a definitive diagnosis a larger tissue sample is usually required, often in the form of an incisional biopsy. The biopsies should be deep as the lesions are submucosal and may be covered by a thickened mucosa due to a reactive inflammatory infiltrate.[9]

The diagnosis may be difficult even with larger biopsies since there are similarities with other small cell tumours e.g. malignant melanoma, aesthesioneuroblastoma, undifferentiated carcinoma and lymphoma. Immunohistochemical staining reveals the presence of kappa or lambda light chain restricted-immunoglobulins which can be used to distinguish plasmacytomas from other tumours.[10]

Plasmacytomas are very radiosensitive and radiotherapy is the main treatment modality. Surgery can be used effectively for small lesions or as a salvage procedure. Chemotherapy is reserved for locally advanced, recurrent or disseminated disease.

Our case has a number of unusual features, firstly the development of an EMP in a patient with long-standing MM. This appears to be an extremely rare event, we were only able to find one other similar case, an EMP of the neck in a patient with IgG myeloma.[1] Our patient also had an IgD myeloma which is rare, representing only 0.6–three per cent of all cases of MM.[11] The EMP grew at an unusually rapid rate for what is typically a slow growing tumour, eventually leading to airway obstruction. It also developed in a background of a relatively indolent case of MM. It has been suggested that such a series of events may be explained by the development of a subpopulation of plasma cells that have been further mutated.[1] In patients with MM and plasmacytoma the prognosis is most probably determined by the activity of the MM and the patient's general medical condition.[1] It also appears that the outlook for patients that develop MM after the diagnosis of EMP is better than those who have MM as the initial diagnosis.[12]

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