Paraganglioma Of The Larynx Presenting As A Neck Mass
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Citation

Abstract
Paragangliomas of the larynx are rare benign tumours that have to be differentiated from other neuroendocrine tumours. We review the literature and present a rare case of a laryngeal paraganglioma that extended into the neck through the thyrohyoid membrane and presented as a neck mass. Cervical extensions of laryngeal paragangliomas have to be differentiated from multifocal tumours. Diagnosis relies heavily on the clinical and radiological features as pre-excision histological diagnosis is not usually available due to the vascular nature of the tumour. Complete surgical excision of both laryngeal and neck components was possible without the need for a partial thyrotomy.

INTRODUCTION
Paragangliomas are uncommon tumours that arise from the numerous paraganglia located throughout the body. In the head and neck, most paragangliomas are, topographically related to the normal occurrence of paraganglia. These paraganglia correspond to the intercarotid, laryngeal, jugulotympanic, orbital, nasal and intravagal groups. The carotid body is the most frequently reported head and neck site in the literature.

Paragangliomas of the larynx are rare tumours. A recent review of the English literature identified 65 cases. Paragangliomas are a subclass of neuroendocrine tumours of the larynx with a neural origin. The other subclass of neuroendocrine tumours are of epithelial origin and include typical carcinoids, atypical carcinoids and small cell neuroendocrine carcinomata.

They show a female to male preponderance of 3:1. Laryngeal paragangliomas are tumours of middle age, with a median age of 44 years.

They are predominantly supraglottic (82%), but may also occur in the subglottis (15%) and the glottic region (3%). These tumours are rarely functional (2.9%) and are seldom associated with other paragangliomas.

CASE REPORT
A 63 year old female presented to the Department of Otolaryngology/ Head and Neck Surgery with a 3 years history of dysphonia, choking and mild dysphagia. She was hypertensive with no history of smoking or alcohol intake. Fibreoptic laryngoscopy revealed a mass apparently arising from the right aryepiglottic fold and occupying most of the supraglottic. (Figure 1) Neck examination demonstrated fullness in the right carotid triangle.

Figure 1
Figure 1: A fibreoptic endoscopic view of the larynx showing a vascular mass occupying most of the supraglottis.

A CT scan of the neck revealed a vascular mass arising from the supraglottis and extending through the thyrohyoid membrane into the neck. The cervical mass appeared separate from the carotid bifurcation with the tentative
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diagnosis of a primary laryngeal tumour. (Figure 2)

**Figure 2**

Figure 2: CT scan of the neck showing an enhancing supraglottic mass extending into the right side of the neck. Coronal reconstruction is shown in inset.

The radiological findings were suggestive of a paraganglioma and a decision was made not to perform a biopsy.

An elective tracheostomy was performed to secure the airway and a carotid angiography with selective embolisation of the feeding vessels was performed prior to surgery to minimise blood loss. This was particularly important as the patient had objected to a blood transfusion on religious grounds.

Surgical excision was performed via an apron incision. The cervical part of the tumour occupied the right paralaryngeal area and was connected to the supraglottic part of the tumour through the thyrohyoid membrane. (Figure 3) The surgical excision commenced with dissection of the cervical component and tracing the connection through the thyrohyoid membrane.

**Figure 3**

Figure 3: Dissection of the cervical component of the tumour (figure 3 a). The excised tumour showing the cervical and laryngeal components connected by a narrow stalk (figure 3 b).

The dissection of the laryngeal component commenced at the superior border of the thyroid cartilage in the subperichondrial plane up to the right ventricle. Both components of the tumour were excised completely and in-continuity followed by repair of the thyrohyoid membrane. The patient had an uneventful postoperative period and was de-cannulated a week following her surgery.

Histologically, the lesion showed the typical organoid arrangement of cells (Zell Ballen) surrounded by sustentacular cells. (Figure 4)
In some areas there were rosette-like structures reminiscent of carcinoid tumours but immuno-histochemically stains for cytokeratin and EMA were negative. Neuroendocrine markers synaptophysin and chromogranin were strongly positive. Other positive markers included, neurone specific enolase (NSE), Vimentin (Figure 5a, b), S100 (neuroectodermal marker) as well as the vascular marker, factor VIII.

**DISCUSSION**

The vascular nature of laryngeal paraganglioma does not allow for a preoperative pathological diagnosis. The presence of a reddish mass in the supraglottis combined with the radiological finding of a vascular mass involving the larynx which enhances with contrast are highly suspicious of this rare tumour.

The neuroendocrine neoplasm that has caused the greatest diagnostic confusion with paragangliomas of the larynx is the atypical carcinoid. The 25 per cent malignancy rate reported for laryngeal paraganglioma is probably based on atypical carcinoid tumours misdiagnosed as paraganglioma.

The ‘Zell Ballen’ pattern is not diagnostic of a paraganglioma as it may also exist in a variety of other tumours including typical and atypical carcinoids, malignant melanoma and medullary carcinoma of the thyroid.

Immuno-cytochemical studies are important in establishing the correct diagnosis as the distinction between paraganglioma and atypical carcinoid may prove difficult on light microscopy. Immunohistochemistry also helps differentiate paragangliomas from primary and metastatic melanomas of the larynx which are chromogranin negative.

Paragangliomas may be multi-centric in origin. There appears to be no previous mention in the literature of laryngeal paragangliomas directly extending into the neck. For the sake of surgical planning, it was important to differentiate between a cervical extension of the laryngeal tumour and the occurrence of two separate tumours; one arising from the supraglottis and the other from the carotid body. This distinction was possible by the absence on the CT scan of splaying of the carotid bifurcation caused by carotid...
body tumours. Stoeckli et al used colour doppler sonography to identify carotid and vagal paragangliomas. Based on the appearance in the B-mode, the hypervascularity, the relationship toward the carotid arteries and the internal jugular vein, and the intratumoral flow direction, colour doppler sonography was able to establish the diagnosis and type of a paraganglioma.\textsuperscript{10} A number of approaches exist for excision of lesions of the supraglottic larynx. The endoscopic approach with or without the use of laser does not lend itself to excising highly vascular lesions such as paragangliomas. The lateral pharyngotomy and anterior (transhyoid) pharyngotomy approaches have been used by various authors to excise laryngeal lesions.\textsuperscript{11,12} However, any approach that involves the pharyngeal mucosa, risks a degree of postoperative dysphagia.\textsuperscript{11} Rubin and Silver, 1992 described the supero-lateral thyrotomy approach for excision of submucosal lesions of the supraglottis including paragangliomas, haemangiopericytomas and saccular cysts.\textsuperscript{10} In our patient, we were able to excise the supraglottic component of the tumour by careful dissection in the subperichondrial plane without the need for excising a window in the thyroid cartilage. The main risk with this approach is injury to the superior laryngeal nerves and great care should be exercised in preserving the internal branches of these nerves as the incision in the thyrohyoid membrane is extended laterally. We intend to follow our patient for the long-term to detect any recurrence at an early stage.

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