Laryngeal paraganglioma

A case report

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Summary

The treatment of a 27-year-old woman with a superior paraganglioma is described. The tumour was excised with a partial supraglottic laryngectomy after repeated biopsies showed chronic inflammatory tissue. The patient has shown no evidence of malignancy during a 4-year follow-up.


Paragangliomas of the head and neck are slow-growing, highly vascular tumours arising from groups of cervical paraganglia. The most common cervical paraganglioma arises from the intercarotid paraganglia and very rarely the laryngeal paraganglia.

The treatment of a patient with a laryngeal paraganglioma is reported.

Case report

A 27-year-old black woman presented with a 2½-year history of intermittent hoarseness, stridor, dysphagia and neck discomfort. Six months before presentation her symptoms worsened. Indirect laryngoscopy at the time revealed a right-sided mucosa-covered supraglottic mass. The right cord could not be seen and the left cord was normal.

On neck examination fullness and widening of the thyrohyoid space was seen on the right. Xerography demonstrated a mass in the right supraglottic region figure (Fig. 1). On direct laryngoscopy a 2.5 cm supraglottic mucosa-covered mass was seen. The rest of the larynx was normal.

Tissue was obtained for histological examination and on two occasions chronic inflammatory tissue was demonstrated. A partial right supraglottic laryngectomy was performed, with excision of the mass and margins.

A histological diagnosis of laryngeal paraganglioma was subsequently obtained. The tumour consisted of a well-circumscribed mass in the submucosa of the false cord. Microscopically the tumour was surrounded by a condensation of connective tissue. Vascularized connective tissue extended into the tumour in a trabecular fashion with fine reticular fibres, and endothelial-lined vessels provided the framework for nests of neoplastic chief cells (Fig. 2). The chief cells were predominantly ovoid in shape with the alveolar pattern resembling the normal paraganglia.

The patient had an uneventful postoperative course and follow-up at 4 years revealed no evidence of local or metastatic tumour.

Discussion

The paraganglionic chief cells arise from neuro-ectoderm derived from the neural crest. The migration of these cells takes place along the paired superior and recurrent laryngeal nerves. The superior paraganglia are situated in the anterior...
false cord while the inferior one is found between the cricoid cartilage and the first tracheal ring.

Tumours of the superior paraganglia are therefore located in the false cord area, while those arising in the inferior paraganglia may present subglottically, intratracheally or adjacent to the thyroid gland. There has been a report of 28 cases of laryngeal paraganglioma with 23 tumours occurring in the superior paraganglia and 2 arising from the inferior paraganglia. These patients, as did our patient, presented with a compromised airway and phonation abnormalities. However, haemoptysis, neck discomfort and a mass in the neck have also been reported. All the reported cases presented with a well-circumscribed submucosal laryngeal mass. Hence laryngeal paraganglioma should be considered in the differential diagnosis of all such masses.

Radiological evaluation includes soft-tissue plain radiograph and xerography which can demonstrate an intralaryngeal mass. This should be followed by computed tomography with contrast medium. If the mass enhances, angiography should be performed.

Pre-operative angiography is essential for the diagnosis and evaluation of paraganglioma and because of the multiple blood supply to these tumours thorough cervical-cerebral angiography is essential. This will demonstrate the blood supply, extent of the tumour and, in addition, other paragangliomas in the region will be identified. If superselective catheterization of the feeding vessels is possible, embolization should be carried out. The resulting devascularization would facilitate the surgical excision.

Surgical treatment of these tumours is indicated. Paragangliomas are radioresistant and the incidence of malignancy is high in laryngeal paragangliomas. Some authors have suggested that laryngeal paragangliomas have the highest incidence of malignancy in the extra-adrenal paraganglioma group. They reported that 24% of laryngeal paragangliomas followed a malignant course, with both local and distant metastases in the form of subcutaneous nodules.

Our patient, and the majority of cases reported, underwent a partial laryngectomy. Hooper reported performing total laryngectomy in 1 patient.

REFERENCES