WK Ho 何偉權

Schwannoma of the larynx—an uncommon cause of vocal cord immobility

喉部神經鞘瘤:一種造成聲帶癱瘓的罕見原因

We present a rare case of schwannoma of the larynx in a 26-year-old Chinese patient. The tumour was excised using a lateral thyrotomy approach, with satisfactory restoration of laryngeal function. The presentation, management, and outcome of treatment for this type of tumour are discussed.

本文報告喉部神經鞘瘤的一個罕有病例。患者為一名26歲的華裔男性;腫瘤以側 面甲狀軟骨切開術切除,喉部功能恢復至令人滿意的水平。本文亦討論了這類腫瘤 的病徵、處理方法以及其成效。

Introduction

A schwannoma is a neurogenic tumour arising from nerve sheaths of peripheral and cranial nerves. Between 25% and 45% of benign schwannomas occur in the head and neck region.^{1,2} Just over 100 cases of laryngeal schwannoma have been reported in the English literature to date.²⁻⁶ We report the first case of a schwannoma of the larynx in a Chinese patient. The tumour was excised using a lateral thyrotomy approach, with satisfactory restoration of laryngeal function postsurgery. The presentation, imaging findings, and management of these tumours are discussed, and the relevant literature is reviewed.

Case report

A 26-year-old Chinese man presented to the Ear, Nose and Throat clinic at Queen Mary Hospital with a 12-month history of hoarseness. He did not have dysphagia or other upper airway symptoms. No palpable lymph nodes were present. Indirect laryngoscopy revealed a submucous bulge in the left aryepiglottic fold, extending to the false vocal cord, which obscured the true vocal cord. Elective microlaryngoscopy confirmed the findings. Translaryngeal fine-needle aspiration cytology was inconclusive, although it demonstrated no evidence of malignancy.

A computed tomography (CT) scan showed a dumbbell-shaped, soft-tissue mass in the left supraglottis, which extended from the aryepiglottic fold superiorly to the false vocal cord inferiorly, pushing the latter medially (Fig 1). The imaging features of periphery enhancement and central necrosis were suggestive of a schwannoma.

Subsequently the patient underwent surgical excision using a lateral thyrotomy approach. Intra-operatively, the tumour mass was found arising from the left aryepiglottic fold, resulting in medial bulging of the left false vocal cord. The mass was completely excised and the thyrotomy closed by primary intention. A prophylactic tracheostomy was performed. Postoperative recovery was uneventful and the patient was decannulated 11 days after surgery.

Macroscopically, the surgical specimen consisted of a well-circumscribed, encapsulated, tan-coloured tumour mass, measuring 3.5 x 2.5 x 1.5 cm. Microscopic sections showed densely cellular areas, composed of compact elongated cells with pallisading (Antoni A pattern) and a less cellular, loosely textured

Key words:

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關鍵詞:

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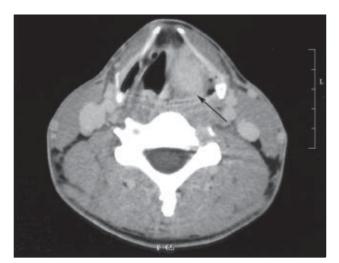


Fig 1. Post-contrast computed tomography scan showing a well-defined, hypodense mass in the supraglottic area. The features of periphery enhancement and central necrosis are suggestive of a schwannoma

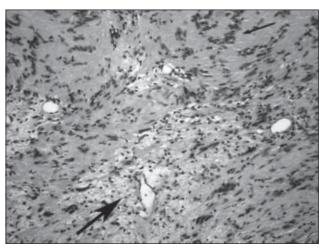


Fig 2. Routine H&E (medium powered, x60) photomicrograph of a schwannoma, showing characteristic Antoni A (small arrow) and Antoni B (large arrow) areas

pattern in which cells often contained lipid (Antoni B pattern) [Fig 2]. Immunohistochemical studies demonstrated reactivity for S100 protein. The overall features were consistent with schwannoma. Four months after the operation, the patient's voice quality had improved and there was full mobility of his vocal cords.

Discussion

The first neurogenic tumour of the larynx was described by Vérocay in 1908.⁷ Since then, there have been few reports documented in the literature, accounting for less than 0.1% of all benign tumours of the larynx. There are two types of neurogenic tumours: schwannomas and neurofibromas. A schwannoma is a solitary, encapsulated, slow-growing benign tumour. More common in women, there is an increased incidence in the sixth and seventh decades of life. Laryngeal schwannomas most commonly occur in the aryepiglottic folds in the supraglottis (80%). The remaining 20% arise from the false vocal cords and the true vocal cords.⁸⁻¹⁰ It is postulated that the internal branch of the superior laryngeal nerve is the most likely nerve of origin.11 Symptoms are related to the mass effect of a slow-growing tumour in the larynx, and include sore throat, odynophagia, dysphagia, stridor, dyspnoea, dysphonia, hoarseness, and a foreign body sensation in the throat. In this case, the tumour occurred in the left aryepiglottic fold and the patient complained of hoarseness only.

Schwannomas are diagnosed histologically by immunohistochemical study for S100 protein reaction and by electron microscopic Antoni A and B patterns. The Antoni A pattern is characterised by compact, spindle-shaped cells, with their nuclei aligned in parallel rows resembling a palisade. The Antoni B type is less cellular, loosely organised, with vacuoles and spindle-shaped nuclei. The

proportions of these two components are variable, and may change abruptly or blend imperceptibly. Enzinger and Weiss¹² suggest that the diagnosis of schwannoma can be made in the presence of three features:

- (1) encapsulation;
- (2) Antoni A and B areas; and
- (3) a positive S100 reaction.

All three features were seen in this case.

On CT scan, small schwannomas are seen as a homogenous, enhancing mass. ¹³ Tumours larger than 3 cm in size often exhibit heterogenic density on contrast enhancement, with centrally distributed areas of low attenuation, surrounded by a peripheral enhancing ring. Cystic changes have also been observed. ¹⁴ On magnetic resonance imaging (MRI) studies, T1-weighted imaging of a schwannoma shows variable intensity, and it is well-enhanced after gadolinium injection. ^{13,15} However, CT and MRI appearances are not diagnostic and cannot always differentiate schwannomas from other benign tumours of the larynx.

Diagnosis can be achieved by endolaryngeal fine-needle aspiration or incision biopsy, although it may be difficult to distinguish between schwannoma and neurofibroma.⁶ In addition, the capsule tends to hinder biopsy.¹⁵ However, these investigations do not alter management. Surgical removal is the treatment of choice. Endoscopic excision is recommended for small lesions, especially for a true vocal cord tumour.² For larger tumours, an external approach with median thyrotomy, lateral pharyngotomy, or lateral thyrotomy have been described.⁵⁻⁷ The lateral thyrotomy approach was used for this case, as it offered the most direct route for tumour excision, avoiding the risk of injury to the vocal fold and laryngeal mucosa.^{4,6} Recurrence or malignant sarcomatous changes following surgical treatment are extremely rare.

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