CASE STUDY

Schwannoma of the Larynx. An Infrequent Laryngeal Tumour

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Abstract Schwannomas are benign tumours, rare among tumours of the larynx. They normally present as supraglottic masses (because they may arise from the internal branch of the superior laryngeal nerve), most commonly involving aryepiglottic folds or false vocal folds. Most patients present with a globus sensation, dysphagia or hoarseness. Conservative surgery is the treatment of choice. We report a case of a laryngeal asymptomatic neurona that was diagnosed accidentally in an imaging test. Complete excision of the tumour was performed through a transoral CO2 laser microsurgery without resorting to a tracheotomy. We discuss the clinical, pathologic and imaging findings and the management of this neoplasm. We also try to update the knowledge on the management of these tumours.

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PALABRAS CLAVE

Schwannoma; Laringe; Neoplasia laringea

Schwannoma laringeo. Un tumor de laringe poco común

Resumen Los schwannomas laringeos son tumores benignos infrecuentes, localizados generalmente en la región supraglótica. Suelen originarse a partir de la rama interna del nervio laringeo superior. La mayoría de los pacientes presentan sensación de globo faringeo, disfagia o disfonía. La cirugía conservadora es el tratamiento de elección. Se presenta un caso de un schwannoma de laringe asintomático, el cual es diagnosticado incidentalmente en una prueba de imagen. La extirpación completa de la lesión se realizó a través de un abordaje endoscópico transoral con láser CO2 sin requerir la realización de una traqueotomía. Revisamos los procedimientos diagnósticos, el diagnóstico diferencial y las opciones terapéuticas de esta infrecuente entidad.

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Introduction

About 50% of neurogenic tumours originate in the head and neck region and are primarily located in the parapharyngeal space.1 However, they are extremely rare among tumours of the larynx, representing less than 0.1% of laryngeal tumours.2 Schwannomas of the larynx are frequently

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located in the aryepiglottic folds and in the false vocal folds. Their origin appears to be the internal branch of the superior laryngeal nerve after penetrating the thyroid membrane. We report a case of laryngeal schwannoma in an asymptomatic, 28-year-old female who was diagnosed incidentally by imaging studies and review her clinical–pathological characteristics.

Case Report

In October 2006, a 23-year-old female was referred to our department by her rheumatologist after observing a cervical mass at the level of the laryngeal vestibule in a magnetic resonance imaging (MRI) scan. The patient was being examined due to neck pain and had no significant medical or surgical history.

The anamnesis included no reports of dyspnoea, dysphonia, dysphagia, cough or any other ENT symptoms.

A cervical MRI scan (T1- and T2-weighted sequences) showed the presence of a well-defined, soft tissue mass, with no signs of infiltrative growth, measuring 30 mm × 40 mm in diameter and located in the supraglottic region. The lesion appeared to originate from the epiglottis and blocked the entrance to the larynx. The mass was hyperintense to muscle on T2-weighted images and isointense on T1-weighted images (Fig. 1).

Nasofibroscopy revealed a large and well-defined polypoid mass which originated in the laryngeal surface of the suprahypoid epiglottis and which prevented visualisation of the true vocal cords. The mucosa covering the tumour was normal and the arytenoid joint was not affected. During breathing movements, the tumour acted as a valve which obstructed the airway.

Given the location and size of the lesion, which could affect the airway, and due to the need to obtain a histopathological diagnosis, the patient urgently underwent direct microlaryngoscopy under general anaesthesia.

Exposure of the lesion was optimal and the exploration confirmed the findings described by nasofibroscopy, so we decided to excise the lesion through CO2 laser microsurgery. The lesion was completely excised in a single block (Fig. 2) and sent for anatomopathological analysis. After resection of the tumour, we verified that both the glottis and subglottis were normal. The procedure was uneventful and the patient was extubated immediately and discharged from hospital after 24 h.

The histopathological study revealed a proliferation of spindle cells forming a well-circumscribed palisade located below a coating of squamous epithelium and with some Verocay bodies. There were no images of mitosis, atypia or necrosis. The cells showed intense immunoreactivity for protein S-100 (Fig. 3). These findings were consistent with the diagnosis of benign, laryngeal schwannoma.

At 5 years of follow-up, the patient is asymptomatic, the exploration of the larynx is normal and the MRI scan shows no recurrence of the disease (Fig. 4).
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Discussion

Schwannomas of the larynx can be diagnosed at any age, but most often occur in the fourth or fifth decade of life. They affect females more frequently and are usually benign tumours whose malignant transformation is rare. Clinically, they are slow-growing lesions, so they usually generate non-descript symptoms for months or years, leading patients to attend consultation much later, once the tumour acquires a considerable size. Patients may report symptoms such as pharyngeal foreign body sensation, sore throat, odynophagia, dysphagia or dysphonia, which persist over time. Only in exceptional cases do the tumours reach sufficient volume to produce severe dyspnoea, stridor and even asphyxia symptoms. Dysphonia may be caused by a decreased mobility of the vocal cords, in the event that the schwannoma originates in them, or more frequently, through an impairment of laryngeal mobility by the effect of the tumour mass, which may mimic cricoarytenoid joint fixation. However, sometimes the lesion is asymptomatic and only discovered upon routine examinations performed for other reasons, as in the present case.

Neurogenic tumours are divided into: (1) schwannomas: well-encapsulated, benign tumours arising from Schwann cells, which grow next to the originating nerve, but extrinsically to nerve bundles and (2) neurofibromas: non-encapsulated, mixed, benign tumours arising from perineural fibrocytes, which usually grow intertwined with nerve bundles of the originating nerve. This differentiating characteristic is important from the surgical point of view because the surgical removal of the tumour from the originating nerve is theoretically possible in schwannomas, but impossible in neurofibromas. In the larynx, schwannomas are more common than neurofibromas and may appear as sporadic lesions or within the spectrum of clinical syndromes, such as neurofibromatosis. Malignant transformation into neurofibrosarcoma and malignant schwannoma has been described in patients with von Recklinghausen disease.

Supraglottic involvement is the most common type among these tumours since they generally originate either in the aryepiglottic folds (80%) or in the false vocal cords (15%), and the originating nerve of these tumours is usually the internal branch of the superior laryngeal nerve. Nevertheless, there have been reports of cases in the true vocal cords, requiring a differential diagnosis with other benign lesions thereof. In our case, the tumour originated from the laryngeal surface of the epiglottis and completely obstructed the laryngeal vestibule, so the possibility of causing sudden asphyxia was high and surgery was urgent. In general, schwannomas tend to grow in the submucosa, but there are some cases, like ours, which present polyoid growth.

Diagnosis of laryngeal schwannomas by endoscopic visualisation is difficult or even impossible due to the rarity of the lesion and the lack of specificity of the endoscopic findings. In computed tomography (CT) scans, schwannomas appear as submucosal, well-defined, hypodense masses, without signs of invasive growth or destruction of adjacent structures. On MRI scans, the lesions appear isointense or slightly hyperintense in T1-weighted sequences, hyperintense in T2-weighted sequences and hyperintense after administration of gadolinium. A differential diagnosis must be conducted with laryngeal cysts, internal laryngoceles and other tumours of the larynx, such as adenomas, chondromas and malignant tumours.

Since exploration and imaging studies cannot rule out a malignant tumour, definitive diagnoses can only be obtained histologically, after a biopsy by direct laryngoscopy. The typical histopathological finding is a proliferation of spindle cells with a fibrillary, elongated, eosinophilic cytoplasm, and with bipolar extensions, similar to Schwann cells. Immunohistochemical staining helps to identify the cellular origin. In our case, we used antibodies against protein S-100, which is characteristic of benign tumours of the nerve sheath. Enger and Weiss established 3 histological criteria for the diagnosis of schwannoma: (1) the presence of a capsule, (2) the presence of a stromal Antoni A and/or Antoni B histological pattern, and (3) positive staining with S-100 (characteristic of Schwann cells). In the Antoni A pattern, cells and fibres are grouped compactly, intertwining and forming rows or
spirals, forming Verocay bodies in a palisade shape. In the Antoni B pattern, cells and fibres are arranged irregularly and there are areas of intercellular oedema forming Luse bodies, which are unique of schwannomas.

Due to the location of the lesion, its excision normally has a diagnostic and therapeutic effect. Treatment is based on complete surgical removal of the tumour. The surgery should be planned according to the symptoms of each patient, as well as the location and extent of the tumour. At present, provided that laryngeal exposure is adequate, the majority of tumours located in the supraglottis can be completely resected by transoral CO\textsubscript{2} laser microsurgery. This type of surgery avoids the need for a temporary tracheotomy, shortens the time until swallowing is restarted and allows patients to be discharged after a shorter period.\textsuperscript{11} However, some authors support the use of external approaches (medial/lateral thyrotomy or medial/lateral pharyngotomy) without prior tracheotomy for extensive submucosal tumours. Moreover, Kayhan et al. recently defended the advantages of robotic transoral surgery.\textsuperscript{12} In our opinion, the effectiveness and safety of laryngeal transoral CO\textsubscript{2} laser microsurgery make it the technique of choice, whenever possible, for the resection of these tumours.\textsuperscript{13}

**Conflict of Interests**

The authors have no conflict of interests to declare.

**References**