NOTE CLINICAL-SURGICAL

NEURILEMMOMA OF THE LARYNX: A CASE REPORT

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ABSTRACT

Neuromas or Schwannomas are extremely rare among tumors of the larynx. They are Schwann cell tumors that can be difficult to distinguish from neurofibromas. They present usually as supraglottic masses, since they may arise from the internal branch of the superior laryngeal nerve. Nuclear magnetic resonance imaging is the best diagnostic technique, conferring a high degree of suspicion. We present an exceptional case of a laryngeal neuroma, with a very long evolution, a large tumor volume, dyspnea and vocal cord fixation, with complete resolution through an external approach following surgical removal. The difficulties encountered with its pathological and clinical diagnosis are discussed as well as a review of the literature.

KEY WORDS: Neurilemmoma. Larynx. Surgery.
INTRODUCTION

Laryngeal neuromas are extremely rare in the tumoral pathology of the larynx. For this reason they cause diagnostic and therapeutic problems that cannot be resolved just from personal or institutional experience.

Neurogenic tumors are benign tumors that are divided into:

- Neuromas or Schwannomas: benign tumors with Schwann cells.
- Neurofibromas: mixed benign tumors, made up of axons, Schwann cells and fibroblasts, coated in a collagen or mucoid matrix.

In small biopsy samples it can be difficult to differentiate them because intermediate forms have also been found.

The neuroma is an encapsulated tumor that has two histological areas:

- Antoni A: spindle cells, organized in compact bundles. Their nuclei are generally long and thin and organized in palisade, leaving space between the rows that form the so-called Bodies of Verocay.
- Antoni B: cells with little organization and great pleomorphism.

There are many documented forms of neuroma, amongst them cellular, cystic, hyalinized and pleomorphic neuromas, but this classification does not have any clinical relevance.

A laryngeal neuroma often appears as a lesion, affecting mainly the ariepiglottic fold and the ventricular bands. This is due to the frequent involvement of the internal branch of the superior laryngeal nerve in the development of this tumor. In comparison, the neurofibroma is described as an excrescent extra-mucosal, non-encapsulated tumor. Clinically, neuromas first appear as very slow growing formations that lead to progressive disphonia and an "odd" sensation when breathing. Only in exceptional cases do they reach enough volume to produce dyspnea. Although this is the general rule it is important to bear in mind that symptoms can vary. They can begin acutely (which makes early treatment necessary) or discovery can be purely accidental. Given the rarity of these tumors, the case we present is of particular interest, primarily due to its long evolution and the fact that it has reached enough size to induce a certain degree of dyspnea.

New image techniques, particularly MRI, allow us to establish a diagnosis of suspected cases by showing the existence of a capsulated and non-infiltrated tumor in determined sequences. This enables us to exclude other diagnoses; on the other hand, modern immunohistochemistry allows a clear differentiation between a neuroma and a neurofibroma.

A CASE REPORT

51-year-old male with no previous personal or family history of interest, referred to our department with dysphonia (which had been developing for 13 years) and exercise dyspnea. The previous hospital had diagnosed him of a ventricular cyst and edema of the a vocal band. At that time the patient had signs of progressive dysphonia and intermittent dyspnea at rest.

A fibrolaryngoscopy revealed a submucous neoformation in the right supraglottic area and the right ventricular band, which did not allow to visualize the right vocal cord and the mucosa covering it looks healthy. The arytenoid had no movement.

The cervical CT scan showed a soft mass partially obliterating the right piriform sinus, swelling of the right arytenoepiglottic fold and glottis, extending to the subglottic region. MRI scan of the larynx and neck (Figure 1), using SPIN-ECO sequences boosted in T1 and T2 with and without intravenous contrast in the sagittal and axial planes and STIR sequence in the coronal plane, showed: a tumor in the right hemilarynx, deep in the paraglottic or paralaryngeal space, located between the thyroid cartilage on the outside, and the paralaryngeal fat, which is displaced medially on the inside. The tumor measured 30 x 25 x 17 mm on the cranio-caudal, anteroposterior and transvers axes respectively. The cranial limit of the lesion was the upper border of the thyroid cartilage, producing swelling of the right arytenoepiglottic fold and homolateral ventricular band and glottis, extending to the subglottic region.

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intravenous contrast. There is also a low signal, band around the whole tumor in all its sequences, which could be related to a possible capsule.

Once the radiological study was completed, we have a right hemi-laryngeal tumor, in the paraglottic space, with radiological signs of being non aggressive, which could be a neurogenic tumor, hemangioma or a benign mesenchymal tumor.

An encapsulated tumor of approximately 3 x 2.5 x 1.5 cm was removed through surgery. To do so, we approached the larynx laterally by dissecting a 1 cm piece of thyroid. After dissecting the mucosa from the piriform sinus, we reached the paraglottic space, dissecting a well-encapsulated tumor, which extended from the band to the vocal fold. The whole procedure was extra-mucous. The anatomo-pathological report of the surgical sample defined it as a neuroma (Fig 2) (a neoplasia made up of elongated cells, laid out in fascicles, with a discreet formation in palisade and extensive areas of fibrosis, telangiectasias and sclerosis of the vessel walls, with recent and old bleeding). The post-operative recovery was favorable and the dyspnea disappeared, but three months after surgery the dysphonia persisted and a fibrolaryngoscopy revealed a marked paresis of the right vocal cord with a slightly atrophy, although it was beginning to recover mobility. Six months after surgery the hemi-larynx was completely mobile and the voice was normal (Fig 3).

DISCUSSION

Neurogenic tumors represent 0.1% of all benign laryngeal tumors. Solitary neurofibromas or those related to a neurofibromatosis (Von Recklinghausen) are more common than neuromas. While neuromas are relatively common in the head and neck area, they are extremely rare in the larynx. Massachusetts General Hospital carried out a revision of all the neurogenic tumors of the larynx they had diagnosed in the last thirty years. Only 1 in 9 cases involved a neuroma, the rest being neurofibromas.

There are approximately 140 registered cases of laryngeal tumors of neural origin but it is difficult to know exactly how many are neuromas. The differential diagnosis of neurilemmoma and neurofibroma is difficult. The latter does not tend to be encapsulated and...
it is characterized by a proliferation of axons, fibroblasts and Schwann cells. The neuroma is immune-reactive to protein S-100, Leu 7, calcineurin, components of basal lamina and Vicentina15, also Schwann cells with elongated cytoplasmas and aligned to the basal lamina are observed via electronic microscopy. Collagen fibres can also be included. There are three diagnostic criteria for neuroma according to Enzinger:16

1. Encapsulated tumor
2. Antoni A and Antoni B areas
3. Positive immune reaction to protein S-100

Although the data provided by the imaging procedures, mainly MRI, are very important for diagnosis, without the resection and patology of the tumor it is very difficult, if not impossible, to carry out a definitive diagnosis.

Stanley et al15 published seven cases of laryngeal neuromas and observed that they were usually localized in the ventricular band and the arytenoepiglottic fold, as in our case. This is due to the fact that the internal branch of the upper laryngeal nerve is frequently implicated in the process. In this sense it is worth stressing that although the mobility of the vocal cord is re-established after surgery, the previous paralysis was caused by the pressure effect of the tumor on the arytenoids. In our case recovery of mobility was slow, given that it took six months to return to normal after surgery.

The treatment of this type of tumor is surgery and the results tend to be excellent. While small tumors can be resected by endoscopy, the supraglottic lesions have to be removed by a lateral pharyngotomy, leaving thyrotomy as the best approach6 for glottic tumors. In larger tumors, such as in the case here, endoscopy did not seem adequate for the exposure and resection of the lesion. A lateral approach similar to that used for supraglottic laryngectomies or laryngoceles has the advantage of providing a more precise exposure and dissection and the fact that we are dealing with an extra-mucosal resection means an additional advantage.

REFERENCES