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Schwannoma de nervo glossofaríngeo em adolescente

Glossopharyngeal nerve schwannoma in an adolescent

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KEYWORDS: Neurilemmoma, Glossopharyngeal Nerve, Parapharyngeal Space, Parapharyngeal Space
PALAVRAS-CHAVE: Neurilemoma, Espaço Parafaríngeo, IX Nervo Craniano, IX Nervo Craniano

INTRODUCTION

Schwannnoma is a benign neoplasm that originates in the myelin sheath of peripheral nerves, spinal nerves, and cranial nerves in Schwann cells. Those of cranial nerves can be seen along the path of the nerves in the intracranial region or in the location of the head and neck. The olfactory and optic nerves are the only cranial nerves that are not affected, as they lack these cells. The most common origin is the eighth cranial nerve, the vestibulocochlear. It has an unknown etiology, and rarely occurs in the oral and maxillofacial region (approximately 1% of cases), requiring surgical treatment of the lesion. In this region, the most common sites are the tongue and oral floor, followed by the oral mucosa, lips, gums, palate and mandible. They are solitary, slow-growing, smooth-surfaced, encapsulated tumors, usually asymptomatic. In more advanced cases, paresthesia and pain due to compression of nerves and adjacent structures may occur.[1,2]

Imaging tests such as computed tomography (CT), magnetic resonance imaging, and ultrasound are useful in diagnosis to estimate tumor margins, structural components, and determine if there is infiltration into surrounding structures. On CT scans, a well-defined mass capable of capturing contrast is observed. Magnetic resonance imaging presents as hypointense or isointense lesions on T1-weighted sequences. On T2-weighted sequences, they are generally homogeneous in moderate signal intensity, although cysts or degenerative changes may be observed within them with higher intensity compared to other compartments, and they present variable enhancement with contrast administration. Histopathological examination allows the identification of possible characteristics compatible with schwannoma. The treatment of this neoplasm consists of surgical excision with a safety margin to ensure a reduction in recurrence rates and favor a good prognosis.

The objective of this report was to describe the case of a young patient with glossopharyngeal nerve schwannoma in the oral cavity who underwent surgical treatment.

CASE REPORT

This report was approved by the Research Ethics Committee of the Hospital de Clínicas, Universidade Federal do Paraná, Curitiba, PR, Brazil – under opinion number 6.224.417.

A young 16-year-old Caucasian woman was admitted to a tertiary hospital after referral to the Emergency Care Unit with a history of a mass in the oral cavity with 2 months of evolution, associated with dysphagia and dysphonia. She denied dyspnea, fever, or recent weight loss. At the initial evaluation, the patient had a protuberance in the posterior region of the oropharynx on the left, with a smooth texture and fibroelastic on palpation. An incisional biopsy of the expansive lesion was performed under general anesthesia, removing whitish fragments that were sent for anatomopathological analysis by immunohistochemistry, which showed low-grade mesenchymal neoplasia with neural phenotype. Magnetic resonance imaging of the skull and neck was requested to evaluate the extent of the lesion and therapeutic planning, which demonstrated a well-defined expansive formation in the parapharyngeal space on the left, posterolaterally displacing the internal and external carotid arteries, and the jugular vein, with radiological characteristics compatible with a low-aggressive primary neoplasm of probable neural origin. Contrast-enhanced CT scan of the neck was also performed, which showed an expansive formation with heterogeneous

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enhancement centered in the parapharyngeal space, measuring 5.8 x 3.0 x 4.2 cm in the largest diameters - longitudinal, anteroposterior and transverse - respectively.

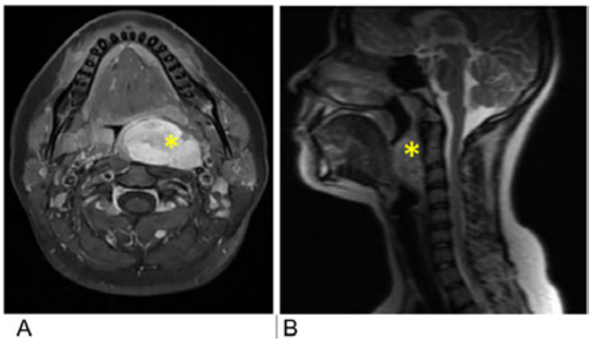

FIGURE 1 — Preoperative T1-weighted magnetic resonance imaging: A) axial section showing the lesion (*); B) sagittal section showing the lesion (*)

Surgical resection was performed with osteotomy of the left lateral mandible, resection of a parapharyngeal tumor, and reconstruction with a microsurgical flap. The patient recovered well after the procedure, complaining of acute pain in the left preauricular region after the first bites of each meal, symptoms compatible with the first bite syndrome, with no other sequelae in the follow-up.

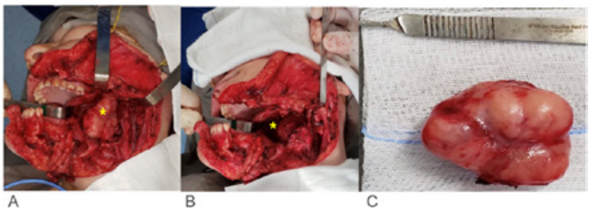

FIGURE 2 — Operative access via mandibulotomy: A) well-defined mass with regular contours visualized in the left parapharyngeal region (*); B) visualization of the operative region after resection of the mass (*); C) appearance of the lesion after resection

DISCUSSION

Schwannoma's is a benign, solitary, slow-growing tumor with rare occurrence in the bucco-maxillofacial region, especially in the oral cavity, as in the case reported.[1,3] In the mouth it is most often seen during the second and third decade of life, with no clear preference for sexes. It mainly affects the soft tissues, with a higher incidence on the tongue.[3] The size of the tumor varies between millimeters and centimeters, rarely exceeding 5 cm.[1] The main symptom reported is painless bulging, and in glossopharyngeal nerve schwannomas the clinical presentation varies from dysphonia, dysphagia and dysarthria to taste changes in the posterior region of the tongue.[2,3] The preoperative diagnosis of schwannoma is difficult, with the differential diagnosis including several benign and malignant causes considering the absence of specific symptoms in the initial phase of the disease.[2] The (CT) images reveal well-circumscribed homogeneous masses that enhance with contrast. However, CT is not very useful in diagnosis due to the limitation of soft tissue differentiation, particularly in small lesions.[1,2] Magnetic resonance imaging is the imaging modality of choice due to the higher tissue contrast and spatial resolution.[1] Histopathological features are used for diagnostic confirmation. Identifying the source nerve can be tricky, as it can be difficult to distinguish between tumors of the lingual, hypoglossal, and glossopharyngeal nerves.[2]

First-bite syndrome should be suspected when the patient describes pain in parotid topography that occurs with the first bite of each meal and improves with subsequent bites. The pathophysiology has been attributed to the unbalanced sympathetic and parasympathetic innervation of the parotid gland and is typically related to the operations of the parotid or parapharyngeal region.[4]

Surgical excision of benign schwannoma with a safety margin is the treatment of choice and favors a good prognosis for the patient, as well as a lower chance of recurrence and malignant transformation of the lesion. Radiotherapy is not indicated. Early diagnosis and the work of multidisciplinary teams are essential for a positive outcome of the condition.

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