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Schwannoma located in the palate: Clinical case and literature review

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Abstract

Schwannoma is a benign tumor that originates from the presence of Schwann cells of the peripheral nerves. They are usually asymptomatic, do not recur, and malignant transformation is rare.

The preoperative diagnosis is often difficult, and although computed tomography and magnetic resonance imaging are very helpful, in the majority of cases, the diagnosis can only be made during surgery and by histological study. The immunohistochemistry reveals that the Schwannoma cells test positive for S-100 protein.

We describe a clinical case of Schwannoma located in the palate of a 15-year-old patient. It is important to highlight that the Schwannoma is usually found in the head and neck, and rarely in the oral cavity. When it does occur in this area, it is more likely to be found in the tongue. Other locations in the oral cavity include: the floor of the mouth, palate, gingiva, vestibular mucosa, lips and mental nerve area, listed from most common to least common.

There has been no sign of recurrence two years after surgery.

Key words: *Schwannoma, neurinoma, neurilemmoma, oral cavity.*

Introduction

Schwannoma is a benign, encapsulated perineural tumor, that originates from the Schwann cells of the nerve sheath deriving from the neuroectoderm (1). It develops from peripheral motor, sensory, sympathetic and cranial nerve sheaths (2). In the parapharyngeal space it usually derives from the vagus nerve and the cervical sympathetic chain. It does not arise from cranial nerves I and II because they lack Schwann cells (3). It is also named neurilemoma, neurinoma and Schwann cell tumor (1). The tumor is solitary, with a smooth surface, and slow asymptomatic growth; although the clinical symptomatology depends on the nerve of origin (1,3).

It does not recur, and malignant transformation is rare (4). Two types are distinguished, central or peripheral Schwannoma, located in bone or in soft tissues respectively (1). It can develop at any age, more commonly in the third and fourth decades (3), there is no predilection for sex (1). Between 25% and 45% of schwannomas occur in the head and neck (3), and 1% only demonstrate an intraoral origin (4); in which case they occur, from greater to lesser frequency, in the mobile portion of the tongue, floor of the mouth, palate, gingiva, vestibule, lips, salivary glands and mental nerve region (1,5).

The etiology of the schwannoma is unknown. It is believed to originate from a proliferation of Schwann cells

in the perineurium causing displacement and compression of the adjacent nerve (1).

The preoperative diagnosis is often difficult and is made by computed tomography and/or magnetic resonance imaging to evaluate the extent and determine infiltration of the surrounding structures (5). A differential diagnosis is needed with other neurogenic tumors such as neurofibroma, neuroma, myoblastoma of granular cells, neurogenic sarcoma, malignant schwannoma, neuroepithelioma and melanoma (2).

The treatment of choice surgical excision. When the tumor is well-encapsulated the exeresis is simple, the difficulty resides in preserving the associated nerve by careful dissection. The prognosis is very good (1).

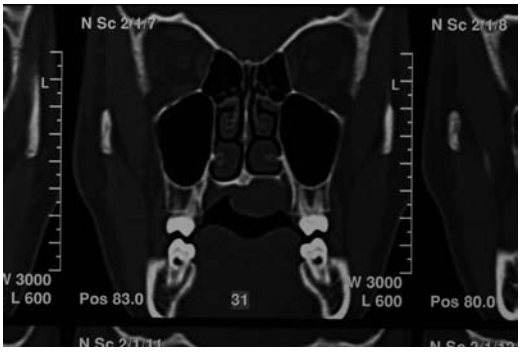

Fig. 1. Computed tomography.

Clinical Case

We present a clinical case of schwannoma in a 15-year-old patient who attended the clinic for a persistent swelling on the left side of the palate, present for three months. The mass was firm and relatively mobile. The mucosa over the tumor was normal. There was no medical history of interest. Computed tomography was requested which revealed a homogeneous mass, slightly less dense than the muscle, with well-defined borders, and without infiltrating the surrounding tissues, which were normal. Slight underlying bone resorption was observed due to pressure (fig.1). Lacking a sure diagnosis a biopsy was decided with local anesthesia (articaine with adrenaline 1:200,000 Ultracaine®). There were no intraoperative complications, however, the appearance of the tumor worsened after the biopsy (fig.2a and 2b). The lesion was sent for histopathological study returning a diagnosis of schwannoma, based on both the histological findings and the immunohistochemical study which was positive for protein S-100. Subsequently, complete extirpation of the lesion was made under general anesthesia, the surgery was simple as the tumor was encapsulated. It was not possible to identify the nerve of origin of the schwannoma, the anterior palatine nerve being a possibility. The patient evolved satisfactorily, with no recurrence two years after surgery (fig.3).

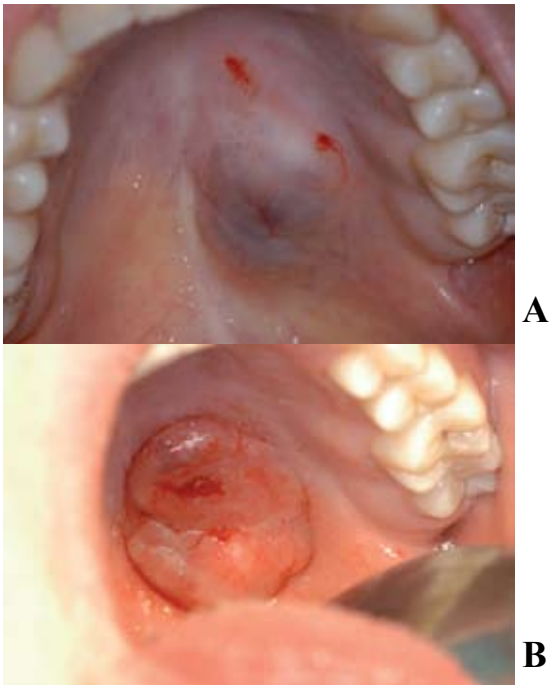

Fig. 2a and 2b. Appearance of the tumor before and after biopsy.

Discussion

Schwannoma has a predilection for head, neck and surface flexors of the upper and lower extremities (6). In the oral cavity it is relatively uncommon, being found principally in the tongue, more frequently in the anterior portion (7). The location in the palate, as in the case presented, is quite rare. Schwannoma can occur at any age, although when present in the oral cavity it tends to appear more often in adults than in children (1). In our case this was a very young patient, 15 years old. These are typically slow-growing, solitary tumors. In some cases there may be a sudden increase in size, possibly due to an intralesional hemorrhage.

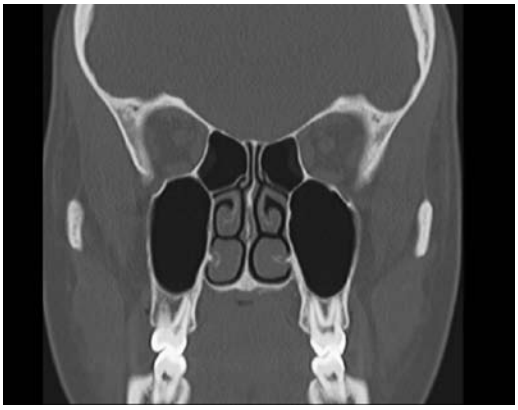

Fig. 3. Postoperative computed tomography (at two years).

Clinically, two forms of oral schwannoma can occur: the most frequent is the encapsulated in which the tumor is surrounded by dense fibrous connective tissue; the other is pediculate, resembling a fibroma (7). The preoperative diagnosis is quite difficult, among other reasons because this is an infrequent tumor and is not usually suspected in the oral cavity. Fine needle aspiration biopsy usually gives negative results (6-8). It is especially difficult to identify the nerve of origin of neurogenic tumors in the cervical region (9). Imaging techniques such as computed tomography and magnetic resonance are very helpful. In the first, the schwannoma appears as a well-defined tumor. Cystic changes can appear probably associated with mucinosa degeneration, hemorrhages and necrosis. In the magnetic resonance the tumor can present as solid, cystic or mixed, this technique is able to determine not only the tumor but also the nerve from which it develops (10). If the nerve of origin is identified, as in the case described by Yamazaki et al. (11) of schwannoma associated with the mental nerve, it is possible to diagnose a peripheral nerve sheath tumor, which at least allows the patient to be given appropriate information regarding the risk of nerve lesion during surgery. Kun et al. (6), were able to make a correct preoperative diagnosis in only 4 cases located in the neck of the 49 cases studied, they concluded that it is very difficult to make the diagnosis based on diagnostic imaging techniques. Kawakami et al. (12), in the case of a tumor in the floor of the mouth, with computed tomography and magnetic resonance a malignant tumor of the sublingual gland. On excision it was found to be a tumor deriving from the lingual nerve, which the histological exam classified as schwannoma. Another diagnostic, although worthless, method is ultrasound (5). The differential diagnosis (by means of histological study) should be made with traumatic neuroma, solitary neurofibroma, granular cell tumor, neurofibromatosis, malignant schwannoma (also called neurogenic sarcoma), nerve sheath myxoma and ganglioneurofibroma, mucosal neuroma associated with multiple endocrine neoplasia 3, neuroepithelioma and melanoma (1,2,11). Although these tumors share a common neurological origin there are differences (13,14). In our case differential diagnosis was also made with lipoma, mucocele and adenoma.

Regarding anatomic pathology, classically two histological patterns are defined, Antoni A (with hypercellularity) and Antoni B (with hypocellularity). The first are formed by fusiform cells with elongated nuclei arranged in a well-organized palisading pattern. Groups of fusiform nuclei, known as Verocay bodies, can frequently be seen. Antoni B tissue shows a disordered arrangement of cells distributed in a loose fibrillar matrix with areas described as microcysts (1,7). The immunohistochemical tests reveal the schwannoma cells to be positive for the protein S-100, a marker for the nervous system (3,5,12,13).

The treatment of choice is excision. The encapsulated form is enucleated easily, whereas the nonencapsulated requires normal tissue margins to avoid relapse. If the nerve of origin is visualized, an attempt should be made to separate carefully to preserve function, although this is sometimes not possible (1,7). In our case the connection with the nerve could not be seen, as occurs in other locations of the oral cavity such as the upper lip (5). The schwannoma should be extirpated in its entirety to avoid tumor recurrence, even if the nerve of origin cannot be preserved, because of the possibility of malignant transformation, which although very rare has been described in the literature (6) and because although a partial exeresis is made, neurological sequelae can remain (2).

The prognosis is very good since it does not usually recur, and malignant transformation is rare. Since it usually develops in the extremities, malignant schwannoma is very rare in the oral cavity, although Hamakawa et al. (15) described a case in the mandible with parotid and lung metastasis. Kun et al. (6), described six cases in the maxillofacial region, two of these had malignant transformation.

Conclusions

Schwannoma should not be discarded when observing a tumor in the oral cavity, as in the case presented.

The definitive preoperative diagnosis could only be carried out with a biopsy and anatomopathologic study.

The treatment consists of surgical excision.

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