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Schwannoma of the Oral Cavity: About Two Cases and Review of Literature

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Case Report

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Abstract: The schwannoma, also called neurinoma, is a benign slow growth tumor from Schwann cells often developing at the expense of the motor and sensory peripheral nervous system. 25% of schwannomas are found in the cervicofacial sphere, but only 1% in the oral cavity; A tongue is the most affected structure. We report two cases of schwannoma of the oral cavity, the first one is in the tongue, and the second case is in the lower lip, discuss the different possible differential diagnoses, and specify the interest of the histopathological examination completed by an immunohistochemical examination, which is the only way to make the positive diagnosis of benign schwannoma. Schwannomas represent clinically a swelling along the peripheral nerves. Spontaneous pain and dysesthesia in the affected sensitive territory are common. Only surgery with histological examination and immunohistochemistry make the diagnosis possible and can differentiate schwannomas from neurofibromas and other differential diagnosis. The most commonly affected organ is the tongue. The malignant transformations of schwannomas are extremely rare. Recurrence after surgical excision is almost never observed for schwannomas. Schwannomas can also appear during the course of neurofibromatosis type I (NFI), as in NF-II. No imaging is necessary. Intraoral location of schwannoma is rare. The treatment is surgical excision. Recurrence is rarely observed.
Keywords: Schwannoma, benign tumor, oral cavity, surgical excision, histological diagnosis.

INTRODUCTION

The schwannoma, also called neurinoma, neurilemmoma or perineural fibro blastoma, represents a benign slow growth tumor from Schwann cells, of neuroectodermal origin, often developing at the expense of the motor and sensory peripheral nervous system. 25% of schwannomas are found in the cervicofacial sphere, but only 1% in the oral cavity; a tongue is the most affected structure.

We report two cases of schwannoma of the oral cavity, discuss the different possible differential diagnoses, and specify the interest of the histopathological examination completed by an immunohistochemical examination, which is the only way to make the positive diagnosis of benign schwannoma.

CASES REPORT

The first patient is a 29-year-old woman, with no particular pathological history, who consults for a limited submucosal swelling of 2 cm long axis in the lower lip, without ulceration or pain. The second patient is a 53-year-old woman, (figure 1) who had a well-defined submucosal swelling of 2 cm long axis at the left free edge of the mobile tongue, with no lingual motility disorder, pain or hypoesthesia.

Both patients had an ultrasound showing a homogeneous hypoechogenic signal. In both cases, the treatment was total excision with no marge under local anesthesia (figure 2).

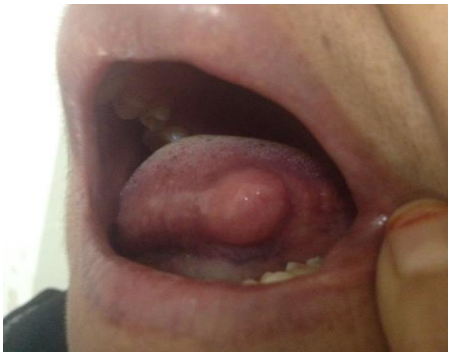

Fig-1: swelling in left free edge of mobile tongue

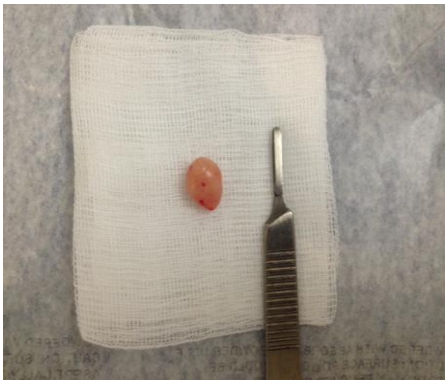

Fig-2: Surgical excision: 2cm encapsulated tumor

The histological examination concluded of the benign schwannoma. No complication or recurrence was observed after a follow-up of 3 years.

DISCUSSION

Schwannomas are tumors originating from Schwann's nerve cells, developing mainly from peripheral motor or sensory nerves. They can occur at any age, but especially between 20 and 30 years (67% of cases) and there is no predominance of sex or race [1]. 25% of schwannomas develop in the cervicofacial region; only 1% of them concern the oral cavity [2-5]; they represent 1 to 9% of all stomatological tumors [2]. According to the literature, oral schwannomas affect the tongue in 50% of cases, especially its mobile part, mainly the lateral edges, whereas it rarely reaches the tip. In the other localizations, we find in decreasing order, the involvement of the palate, the floor of the mouth, the vestibule, the gum, and the lip [2-4, 6, 7]. Bone involvement is very rare. Fawcett and Dahlin report 7 cases of schwannomas on 3,987 bone tumors [1]. Of unknown etiology, certain factors are incriminated in their development: chronic irritations, trauma [8], and treatment by radiotherapy [5, 8]. The lesion is usually unique. Multiple schwannomas (schwannomatosis) can be observed in type II neurofibromatosis, which is characterized, inter alia, by an acoustic schwannoma, bilateral or not [9].

Benign schwannoma poses the diagnostic problem with other nerve tumors, in particular with neuromas, consisting of a proliferation of axons and Schwann cells, most often induced by local trauma, for example, during a Caldwell Luc's procedure. But also with Abrikossof granular cell tumors, gliomas, neurothecomas, extracranial meningiomas, chondromas, melanotic neuroectodermal tumors [10]. Apart from nerve tumors, the differential diagnosis arises with lipomas, leiomyomas, hemangiomas, lymphangiomas, and benign or malignant tumors of the salivary glands.

Clinically, schwannomas are most often asymptomatic, slow growth, but they can be accompanied by pain and/or paresthesia in 50% of cases. Other symptoms have been reported depending on the tumor localization, we can note the appearance of dysphagia and a change of voice in schwannomas of the base of tongue, neurological signs such as facial paralysis if it develops in facial nerve, decreased auditory acuity in acoustic neuromas, dyspnea if there is nerve damage IX, X, XI, or loss of lingual sensitivity if it affects the lingual nerve). The most affected nerves are the V, VII, VIII, IX, XI, XII. For the lingual localization, the examination shows a firm and regular swelling, the mucosal ulcerations are extremely rare.

Radiologically, the images obtained are not very specific. Ultrasound shows a homogeneous, hypoechogenic signal with posterior reinforcement. The

CT scan shows a heterogeneous or homogeneous mass, isodense respecting the soft tissues, with heterogeneous enhancement [4,6]. MRI shows isointense lesion in T1 muscles and hyperintense T2, with enhancement after gadolinium injection [11]. Imaging makes possible to understand the extension of the tumor.

The treatment is surgical; it consists of a complete excision of the tumor without safety margin, if it is well encapsulated. Histopathological examination of the piece shows a tumor surrounded by a fibrous capsule, consisting of two cell populations, Antoni A cells (majority) and Antoni B cells [4, 7]. Fusiform cells A, embedded in connective tissue high in collagen, are oriented parallel, grouped in compact bundles around an acellular central zone. This set forms the corpuscles of Verocay. The nuclei of these cells are aligned in palisade. B cells are less ordered, crosslinked, within loose connective tissue and fibers surrounding microcysts. The tumor is not very vascularized; the vessels dilated, with thickened and hyalinized walls. The histopathological examination makes it possible to rule out the diagnosis of malignant schwannoma [12, 13], whose frequency is of the order of 5 to 10% of all soft tissue sarcomas, by showing suggestive but less specific characters. : presence of hyaline strips and nodules around which the cells can be arranged in rosettes, peri and intra-neural extension of the tumor, cell proliferation around vessels herniated in the vascular lumen, heterotopic elements (approximately 15% of cases), most often in the form of islets of cartilage or mature bone, more rarely in the form of striated muscle fibers (Triton tumor) or mucosecreting glands (glandular malignant schwannoma). The appearance of cells is an important argument for diagnosis. Spindle-shaped cells have irregular contours with asymmetric curved, wavy or comma-shaped nuclei. The cytoplasm is poorly colored, with poorly visible boundaries. The immunohistochemical study, whatever the nature of the schwannoma, found a positive reaction for the S100 protein [14].

Old schwannoma is a rare variant of benign schwannoma characterized by degenerative histological changes: calcifications, hyalinization, cystic formations. There are also areas of cellular atypia that misdiagnosis malignancy [15].

There is a risk of malignant transformation of benign schwannomas, which varies between 8 and 13.9%, by increasing its size; the vascularization becomes insufficient, encouraging the malignant transformation [15]. Despite this risk of malignant transformation, benign schwannomas keep a good prognosis if the surgical excision is complete, the recurrences are exceptional. On the other hand, malignant schwannoma is a tumor of bad prognosis with an overall survival of 20 to 25% in case of Recklinghausen's disease and of 50% in case of isolated tumor. The poor prognosis in Recklinghausen disease is related to the location of tumors (more often at the trunk or proximal), the size and the higher grade, and the fact that some patients develop several sarcomas simultaneously. Local recurrences are common, and metastases (lung, liver, skin, and bone) usually appear within two years of diagnosis [12, 13].

CONCLUSION

Schwannoma is a rare histological type of tumor, the treatment is only surgical, we rarely observe recurrences, malignant transformation or regional complications. No predisposing factors are known actually. The future researches must defined etiologies, factors of malignant transformation and recurrence in order to more highlighting this tumor.

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