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Recurrent plexiform schwannoma in vestibular mucosa

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SUMMARY: **Recurrent plexiform schwannoma in vestibular mucosa.**

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Schwannoma, also called neurilemmoma, is a benign neoplasm of peripheral nerve sheath. An infrequent location of a multiple intraoral plexiform schwannoma arising on the branches of the facial nerve in the vestibular mucosa of a young male patient is here discussed. Surgical treatment has been discussed on the base of histological findings and diagnosis.

RIASSUNTO: **Schwannoma plessiforme recidivo della mucosa vestibolare.**

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Lo schwannoma, detto anche neurilemmoma, è una neoplasia benigna della guaina del nervo che raramente si presenta nella cavità orale e ancor più raramente nella mucosa vestibolare. Gli Autori presentano un raro caso di localizzazione di uno schwannoma plessiforme multiplo endorale originato su rami del nervo facciale nella mucosa vestibolare di un giovane paziente. Il trattamento chirurgico viene discusso sulla base del quadro istologico e della diagnosi.

KEY WORDS: Plexiform schwannoma - Neural tumors - Oral mucosa.
Schwannoma plessiforme - Tumori neurogeni - Mucosa orale.

Introduction

Whereas the head and neck region is a common site for benign neural tumors (25-37%), the lesions of the facial branches are uncommon, and a small number of these lesions occur in the oral mucosa (1-6).

Schwannoma, a benign neoplasm of peripheral nerve sheath, rarely occur in the oral cavity (1%): the most common form is a unilobular tumour on the dorsum of the tongue whilst the vestibular mucosa is a rare location. Multiple intraoral plexiform schwannoma, as those we reported, is very unusual. The recurrence is rarely reported (3, 4, 7, 8).

Case report

A 16 year old male was referred for a 3 cm mass in the vestibular mucosa, slowly grown in ten years, causing pain during mastication but no weariness during phonation. Clinical examination showed: two intraoral mass (3 cm and 1 cm), little deforming the lower lip, detectable extra-orally, without skin changes (Figs. 1a). Mucosa mobility was conserved, but the masses, not painful, are fixed over the underlying muscle. A slight impairment of the buccal motility (lowering of the right commissure when lips were stretched) was present.

The patient underwent surgical treatment by an intraoral approach under local anaesthesia. A 3 cm long incision evidenced the larger and the smaller tumour as well encapsulated masses, with no adherence to adjacent structures. Two other similar nodules were detected above the formers which required a second, 2 cm long, upper incision. Macroscopically: firm and multinodular masses, individually well-encapsulated with a thin, fibrous thickening (Figs. 1a). Microscopically: spindle, tightly compacted cells with fusiform and tapered nuclei; palisades created by alignment of nuclei alternating with cytoplasm-rich, a nucleate zones produce a staggered pattern (referred to as Antoni A). Histological diagnosis: plexiform schwannoma (Figs. 1b).

No neurological deficit was found after surgery, physical examination failed to reveal signs or symptoms of neurofibromatosis. None of family members presented mucosa nodular pathology.

Six months later, another intraoral nodule arose from the same side of the vestibular mucosa, progressively increases in size and caused pain during mastication (Figs. 1c). In the meantime 3 minor lesions at the lip commissure became evident.

Three year after first surgery a new intraoral incision was performed in local anaesthesia: neoplasm was hard, the cut surface presented smooth contours with no adherence to adjacent structures. The nervous branch at the lip commissure, infiltrated with anaesthetic, was dissected. Four tumour lesions were detected: the biggest one distally, another (0,5 cm) at the back, and two others (0,2 cm) more proximally. All lesions were located within the nerve but in eccentric position: total resection of them was possible, preserving nerve function. Therefore another 4 cm long incision, inferior to the first one, was performed: difficult proximal

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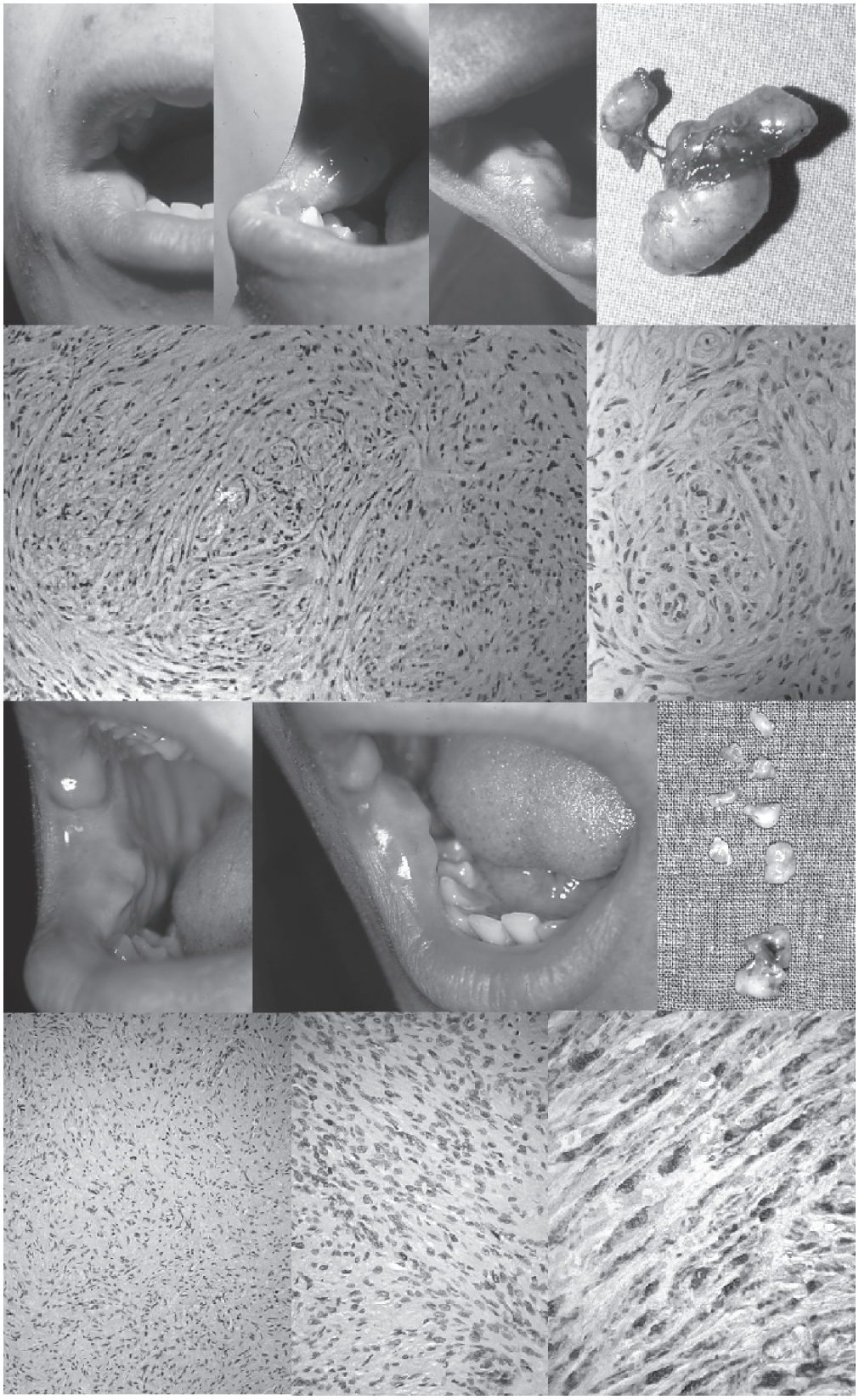

a)

b)

c)

d)

e)

Fig. 1 - a) First clinical observation: nodule in vestibular mucosa at the lip commissure and resected tumors.
b) Microphotograph of surgical specimen (haematoxylin-eosin) at the first operation (x10 HPF and x20 HPF): the tumor shows fascicular pattern and intracytoplasmatic vacuoles.
c) Three years after: clinical observation and resected tumors (multinodular).
d) Microphotograph of surgical specimen (haematoxylin-eosin) at the second operation (x10 HPF and x40 HPF): the tumor spindle cells with elongated nuclei and palisades arrangement.
e) The immunoprofile shows widespread (nuclear and cytoplasmatic) S-100 protein reactivity: microphotograph of surgical specimen at the second operation (x50 HPF).

TABLE 1 – GUIDELINES FOR THE DIFFERENTIAL DIAGNOSIS OF SCHWANNOMA (MODIFIED BY KOIZUMI - ref 5).

Characteristic	Conventional schwhannoma	Cellular schwhannoma	Plexiform schwhannoma
Encapsulation	Common	Variable	Common
Necrosis	None	Occasional patches of necrosis but no gross necrosis	None
Pathologic formation	Antoni A and B areas with Verocay bodies	Mainly hypercellular Antoni A tissue	Antoni A
Cellular differentiation	High to moderate	High to moderate	High
Tumor necrosis	None	Minimal	None
Atypical mitosis	None	None	None
S-100 protein	Positive	Mostly	Diffusely and strongly positive, nuclear and cytoplasmatic

dissection, due to prior surgery, were accomplished. Five other lesions (0,2 - 0,4 cm) were found within the branch and were resected preserving most of the nerve fibres (Figs. 1c). The resection of a smaller lesion (0.2 cm) located under the muscle fascia was not attempted: the local anaesthesia did not allow to evaluate correctly the resection effects on the nerve integrity.

Histology confirmed that all lesions were plexiform schwannomas: the tumours show spindle cells with fusiform nuclei and palisades arrangement; the proliferation of Schwann cells clearly lies within nerve (Figs. 1d). The cells show nuclei and cytoplasm diffusely and strongly immunoreactive for S-100 protein (Fig. 1e).

Seven years later the patient did not show any sign of recurrence, no neurological deficit, no pain or difficulty during mastication.

Discussion

The oral peripheral nerve tumors include schwannoma, neurofibroma, nerve sheat myxoma, palisaded encapsulated neuroma, mucosal neuroma associated with multiple endocrine neoplasia III, traumatic neuroma and granular cell tumor. Although these tumors share a common neural origin, they exhibit microscopic and pathogenetic heterogeneity (9, 10).

Schwannoma is a benign encapsulated nerve sheath tumour composed entirely of Schwann cells, that may develop in peripheral cranial or autonomic nerves (6). While most conventional schwannoma are well-encapsulated, firm, globular to multinodular masses lying eccentric to their parent nerve mucosa conventional schwannoma are rare and unencapsulated. The majority of conventional schwannomas exhibit two distinct histological pattern: Antoni A and B. The former consists of elongated, tightly compacted cells with fusiform and tapered nuclei: palisades created by alignment of nuclei alternating with cytoplasm-rich, nucleate zones (Verocay bodies) produce a staggered pattern, which is either regimented over several low-power fields or jumbled in distribution. Antoni B tissue consists of an ultrastructural meshwork of loosely disposed cells with rounded hyperchromatic nuclei and short, disordered processes. All schwannomas are diffusely immunoreactive for S-100 protein.

In addition to conventional schwannoma, there are two other morphologic variants: cellular schwannoma (a hypercellular schwannoma composed largely or exclusively of Antoni A tissue) and plexiform schwannoma (Tab. 1) (5). This form of schwannoma exhibits a plexiform or multinodular pattern of growth. Although the parent nerve is not always evident, the lesion does involve superficially situated nerves (dermal and/or subcutaneous). Histologically, the proliferation of Schwann cells clearly lies within nerves. A small proportion of plexiform schwannomas recur if incompletely excised (11).

The schwannoma is indistinguishable from other encapsulated benign tumours on the basis of clinical findings. Consequently, occasional diagnosis when the neoplasm are removed is not unusual (1, 12).

Preoperative diagnosis is important: in a benign lesion a resection preserving the nerve function is the treatment of choice whilst in a malignant neoplasm an extended resection and adjuvant chemotherapy are recommended (12-14). Malignant transformation of a benign schwannoma is extremely rare, as opposed to the transformation of a neurofibroma in neurofibromatosis (13, 14).

The cranial nerve involvement and the evaluation of the pain caused by the lesion are no useful criteria for a histological classification. Tomography and CT scans disclosing a well-circumscribed capsule are not helpful. Neither aspiration needle biopsy nor simple biopsy of the suspicious area are reliable techniques. Histology during operation is not a certainty. Only DNA flow cytometry during operation would have given useful indications on the histological diagnosis (14).

In our case significant unjustifiable neurological lesions of the facial nerve branches were expected if a complete resection has been attempted. Therefore, one little lesion was not resected when the second surgery was performed, and regular follow-up was performed.

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