PLEXIFORM (MULTINODULAR) SCHWANNOMA OF SOFT PALATE.
REPORT OF A CASE

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ABSTRACT
Plexiform schwannoma is a rare benign neoplasm of the neural sheath characterized by a multinodular plexiform growth pattern. Only 5% of schwannomas have a plexiform or multinodular growth pattern. Schwannoma apparently derives from the Schwann cells. Extracranially, 25% of all schwannomas are located in the head and neck region, but only 1% show an intraoral origin. The intraoral lesions show a predilection for the tongue, followed by the palate, buccal mucosa, lip and gingival. Microscopic examination is necessary to confirm the diagnosis. Characteristic histological signs are the palisading of the spindle-shaped Schwann cells around the central acellular area, so called Verocay bodies.

We report a case of a 21-year-old woman with a smooth mass of the soft palate that was gradually increasing. Surgical excision of the mass was done and the histopathology and immunohistochemistry study of the excised lesion revealed a multinodular plexiform schwannoma of the soft palate. The patient is under regular clinical control, with no signs of recurrence after 17 months.

Plexiform schwannomas of the soft palate are mentioned very rarely in the English literature. This rare benign tumor is worthy of recognition because it can be misdiagnosed as plexiform neurofibroma.

Key words: neural sheath tumor, plexiform schwannoma, neurilemmoma, oral cavity, anti S-100 protein

INTRODUCTION
Schwannoma, also known as neurilemmoma or perineural fibroblastoma is a rare benign, encapsulated perineural tumor of neuroectodermal derivation, arising from the neural sheath Schwann cells of the peripheral, cranial or autonomic nerves. The etiology is unknown, but it is postulated that the lesion arises by the proliferation of Schwann cells at one point inside the perineurium.1,2

We report the clinical, histopathologic and immunohistopathologic features of a very rare case of a plexiform (multinodular) schwannoma of the soft palate.

CASE REPORT
A 21-year-old female patient presented to the ENT Department with swelling on the soft palate, for 14 months. The swelling mass gradually increased in size but there was no pain. Intraoral examination revealed a yellowish, 1.5-2-cm well circumscribed mass of the soft palate. There was no tenderness to palpation and no evidence of paresthesia. There was no ulceration and the overlying mucosa was normal. The rest of the oral cavity was normal. Macroscopically, the lesion was grossly similar to a lipoma and clinically appeared to be a benign soft tissue neoplasm.

Excision of the mass was done with adequate surgical margins of resection and the tissue was sent for histopathological examination. The histopathological findings revealed that the mass was well encapsulated, had plexiform structure and the tumor cells had a fusiform aspect and were organized forming arrangements of Antoni type B. Rare areas with Antoni type A aspect, forming Verocay bodies, were observed. The immunohistochemical examination for S-100 protein was done and revealed intense positivity in the cells of the tumor.
Based on the clinical behavior, histological and immunohistochemical aspects the final diagnosis was of a plexiform schwannoma of the soft palate.

DISCUSSION

Schwannoma is a benign encapsulated nerve sheath neoplasm composed of Schwann cells in a poorly collagenized stroma. They were first described by Verocay in 1908. In 1935 it was proposed that these tumours arose from nerve sheath elements and they were termed neurilemmomas. Embryologically, Schwann cells arise during the fourth week of development from a specialized population of ectomesenchymal cells of neural crest.2,3

Most reports suggest that the majority of tumors are present between the ages of 10 and 40 years2,4 and are equally distributed between the two sexes. Extracranially, 25% to 45% of all schwannomas are located in the head and neck region. However, the intraoral lesions are very rare.2-5 Only 1% show intraoral origin. It is usually asymptomatic, commonly appears as a single, slow-growing encapsulated nodule, but sometimes can cause displacement and compression of the surrounding normal nerve tissue. Occasionally there may be pain and paresthesia (commonly affected VIII acoustic cranial nerve).5,6 In the oral cavity, the lesion is usually presented in soft tissue, more commonly the tongue, followed by hard and soft palate, buccal mucosa, lip and gingival.7-12 The lesion may have clinical features similar to other benign lesions like fibromas, lipomas, mucocele, epithelial hyperplasia and benign salivary gland tumors.8,11

Immunostaining analysis is critical in the diagnosis of these neoplasms. Histologically, schwannomas show two different components, Antoni type A and Antoni type B tissue. Antoni type A consists of Schwann cells arranged in compact, twisted bundles, associated with delicate reticulin fibres and spindle-shaped nuclei aligned in parallel rows forming a typical palisading pattern. Between the rows there are fine cytoplasmatic fibrils with acellular, eosinophilic masses called Verocay bodies. Antoni type B tissue is formed by irregularly arranged masses of elongated cells and fibers similar in appearance to neurofibroma, with areas of cystic degeneration and edema.7,9,12 The tumoral cells with Antoni A show greater intensity scores compared to Antoni B tumor pattern.13,14

Plexiform schwannoma which represent 4.3% of all schwannomas is a rare variant of Schwann cell tumor.7,14 Plexiform schwannomas of the soft palate are particularly rarely mentioned in the relevant literature.7,8,14

In this case, the plexiform schwannoma presented as a slow growing, circumscribed swelling of the soft palate without any particular features to distinguish it from other benign lesions of soft tissue. The initial diagnosis after clinical examination was a benign neoplasm of mesenchymal origin or minor salivary gland neoplasm. No specific assumption was formulated concerning schwannoma because it is a rare oral lesion, particularly at the soft palate. To establish the diagnosis, the lesion was totally excised under local anesthesia. The histological findings in this case revealed a multinodular growth pattern, a majority of Antoni B pattern for the whole specimen and a nonorga-

Figure 1. Photomicrograph shows multinodular growth pattern and the plexiform pattern (original magnification x 20; hematoxylin-eosin stain).

Figure 2. Photomicrograph reveals that tumor cells show intense positivity for S-100 protein (original magnification x 20; immunostaining for S-100 protein).
nization of the tumoral Schwann cells forming a few Verocay bodies which are classical structures of the plexiform schwannoma (Fig. 1). Immunohistochemical study for S-100 protein was done and it was intensively positive (Fig. 2).

CONCLUSIONS

In conclusion, benign schwannoma represent a lesion that is often not even considered in clinical practice. Plexiform schwannoma of the soft palate is extremely rare and in many cases is misdiagnosed. No more than 2 cases of plexiform schwannoma of soft palate have so far been reported in English literature.7,15 It is very useful for the clinicians to know the exact origin of the lesion because the therapy differ in every case.

REFERENCES