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CASE REPORT
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Palisaded encapsulated neuroma in tongue – A commonly misdiagnosed peripheral nerve sheath tumor

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Abstract

Palisaded encapsulated (solitary circumscribed) neuromas are benign neural tumors, rarely found in oral mucosa. This case reports a 24-year-old man that presented a unique soft nodule at the left side of the tongue. An excisional biopsy was performed under local anesthesia and histopathological examination of the surgical specimen revealed a well circumscribed mass composed of spindled Schwann cells, often aligned and fasciculated, forming occasional nodules embedded in a fibrous stroma. Histopathological analysis showed the presence of positive cells for S-100, EMA, CD57, and collagen IV. Based on clinical and microscopical features, the diagnosis established was of palisaded encapsulated neuroma. This case report aims to discuss the differential diagnosis among palisaded encapsulated neuroma and the other neural tumors that affect the oral mucosa.

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Full Text

Introduction

The palisaded encapsulated neuroma (PEN) is a distinct form of true neuroma, consisting of benign proliferation of Schwann cells and axons, often aligned and fasciculated, roughly in equal amounts and without propensity for recurrence or malignant transformation.[1],[2],[3]

The PEN and the traumatic neuroma share the same origin; however, some clinical and histopathological features are distinct.[2],[4] Traumatic neuroma represents a proliferative response following an extrinsic damage to a nerve fiber.[2] Like PEN, this lesion is composed of nerve fascicles separated by clefts, but the fascicles are arranged haphazardly and embedded in fibrous scar tissue, signifying a prior injury.[2]

Although PEN is more commonly found in the skin, the oral mucosa remains as the second most frequent site of this tumor where it usually occurs as small superficial and asymptomatic nodules on hard palate, tongue, and lips.[5],[6],[7]

The aim of this paper is to report a case of a 24-year-old man presented for removal of a unique soft nodule at the dorsum of tongue and to discuss the differential diagnosis of the PEN with others neural tumors that affect the oral mucosa.

Case Report

A 24-year-old man presented a unique soft nodule at the dorsum of the tongue, which has been present since he was a teenager. His medical history was non-contributory. Ophthalmological and thyroidal exams were normal. Intraoral examination revealed a unique sharply demarcated, coalescent, pink, and pedunculated nodule, measuring approximately 1.0 × 1.0 cm, located in the dorsum of the tongue. An excisional biopsy was performed under local anesthesia and the histopathological examination revealed a non-encapsulated but well-delineated, submucosal tumor covered by oral mucosa. Neoplastic irregular Schwann cells were observed, forming nerve fascicles of various sizes and shapes, embedded in a collagenous background [Figure 1]. A slight chronic inflammatory reaction was observed subjacent to oral epithelium. Blood vessels were inconspicuous and most of them had thin walls. Immunohistochemical stains for S-100 protein, EMA, CD57, and collagen IV were performed using a standard streptavidin-biotin-peroxidase method on deparaffinized tissue sections [Table 1]. Immunoreactivity was graded with a semiquantitative method and the number of positively stained cells was evaluated in five high-power fields (x400), as shown in the [Table 1]. Spindle to stellar shaped cells, which were compatible with Schwann cells, showed intense positivity for S-100 protein [Figure 2]a and [Figure 2]c and moderate positivity for CD57 (Leu-7). Peripherally, nerve fascicles were circumscribed by perineurium presenting EMA positivity [Figure 2]b, however, there was a lack of cells with immunopositivity for EMA in the center of the tumor. There was no immunoreactivity to collagen IV into the lesion [Figure 2]d. The definitive diagnosis was of palisaded encapsulated neuroma, also known as solitary circumscribed neuroma. One-year after the surgical excision, the patient had no signs of recurrences.{Figure 1}{Table 1}{Figure 2}

Discussion

Oral PEN represents 0.05% of all intraorally peripheral nerve sheath tumors and less than 8% of PEN cases occur in the tongue.[5] The present case report seems to be the second patient, in English literature, with PEN located in dorsum of the tongue; however, it is important to consider that this tumor is frequently misdiagnosed, especially when its occurrence is not in the usual oral sites, such as the palate.[8]

The clinical hypothesis of the lesion in our case was of peripheral nerve sheath tumor, particularly, the traumatic neuroma. The hypothesis of traumatic neuroma was ruled out as the patient reported to have the lesion for a long time (around 10 years), without symptoms, and no previous history of trauma. The histopathological analysis in hematoxylin and eosin confirmed the neural origin of the lesion. Although some authors described that, microscopically, PEN present areas that resembles traumatic neuroma, some features including the absence of inflammatory cells and the parallel arrangement of the nuclei, resembling palisading pattern or Verocay bodies, as observed in the case reported [Figure 1], contributed to establish the diagnosis of PEN.[7],[9],[10] Other histopathological differences residues in the fact that while PEN are characterized by well circumscribed nodules, composed of Schwann cells arranged in interlacing fascicles separated by cleft-like spaces surrounded by a thin fibrous capsule, the traumatic neuromas show nerve fascicles arranged haphazardly and embedded in fibrous scar tissue.[2],[10] The pathogenesis of PEN remains unsettled and the proliferation of axons and Schwann cells confirms a reactive or hyperplastic nature of the tumor suggesting its hamartomatous origin.[7],[10],[11]

Although PEN and traumatic neuroma present the same tissue origin, the microscopic patterns could be confusing and not sufficient to establish the diagnosis.[12],[13] An useful tool to differ such lesions in this tumor group is the immunohistochemistry.

The immunohistochemical analysis of PEN in the present case revealed an intense positive expression of S-100 and collagen IV, reinforcing the neural origin, with moderate positivity to EMA, peripherally to the tumor, and to CD57 (Leu-7) by Schwann cells. Then, others neural tumors as neurofibroma and schwannoma was also included in our differential diagnosis of PEN. The neurofibromas are distinct of PEN because they are often associated with neurofibromatosis and have a tendency for malignant transformation.[14] Microscopically, neurofibromas lack a capsule, usually have a myxoid stroma with delicate fibrillary pattern of collagen deposition and have fewer axons with myelin sheaths.[3],[14],[15] Although Schwannomas have a complete capsule, they differ from PEN because of the fascicular Antoni A cells (with Verocay bodies) and schwannomas usually show a more definite palisading in the nuclei than PEN.[9]

Additionally, in the present case, the positive and intense S-100 staining in Schwann cells of PEN [Figure 2]a helped to differ it from neurofibromas and Schwannomas.[15] The S-100 protein is found predominantly, but not exclusively, in the nerve system. This protein is expressed by Schwann cells, but not by perineural cells and endoneural fibroblasts and all the oral benign neural tumors present an intense expression of S-100 protein, except neurofibromas.[12],[13],[16],[17] The immunohistochemistry pattern presented in our case reported is in accordance with other authors who found a positive and consistently staining for S-100 in Schwann cells of most palisaded encapsulated neuromas cells.[4],[5],[10],[11],[14],[16],[18]

Another distinguishing feature was the moderate positive expression of EMA by the perineural capsule of the surrounding lobules of PENs [Figure 2]b, which was described by others.[11],[16],[18] This lack of EMA positive cells within PEN, as observed in our case reported, was confirmed by Magnusson *et al.* (1996) as a differential diagnostic feature of traumatic neuromas.

Furthermore, in the present case, we also observed a diffuse and moderate expression of CD57, suggesting an inconsistent production of myelin sheath proteins by Schwann cells [Figure 2]c. Similar studies[12],[18] had found a variety of 0–25% of positive CD57 Schwann cells in palisaded encapsulated neuromas. Differently, Schwann cells of traumatic neuromas usually show intense reactivity for CD57 (more than 50% of cells) while a low expression of CD57 is observed in schwannomas, which contributes to distinguish these lesions and others peripheral nerve sheath tumors.[12],[18]

Palisaded encapsulated neuromas, as the case presented here [Figure 2]d, have high expression of collagen IV due to the presence of many Schwann cells and perineurium. This high expression of collagen IV can be also identified in schwannomas,[10],[19] suggesting that this marker could be helpful to identify lesions of neural origin, but it may not be of great value in separating the various neural subtypes.

As a benign peripheral nerve sheath tumor, the conservative surgical excision of PEN is the recommended treatment and recurrences are not expected.[3] In our patient, there was no signs of recurrence after 1-year of surgical excision.

In conclusion, this case report highlights the importance to ally the histopathologic and immunohistochemical features to distinguish peripheral nerve sheath tumors. The careful analysis avoids misdiagnosing PEN and the incorrect treatment.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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