Case Report

Palisaded Encapsulated (Solitary Circumscribed) Neuroma of the Buccal Mucosa: a Rare Case

Saede Atarbashi-Moghadam [1], Ali Lotfi [1], Saman Salehi Zalani [2], Sepideh Mokhtari [3]

[1] Dept. of Oral and Maxillofacial Pathology, School of Dentistry, Shahid Beheshti University of Medical Sciences, Tehran, Iran.
[2] Student, Research Center, Dental School of Shahid Beheshti University of Medical Sciences, Tehran, Iran.
[3] School of Dentistry, Tehran University of Medical Sciences, Tehran, Iran.

KEY WORDS
Neuroma;
Soft Tissue Neoplasms Mouth;
Mucosa;

Received July 2016;
Received in revised form November 2016;
Accepted December 2016;

ABSTRACT

The rarity of oral soft tissue spindle cell tumors combined with overlapping microscopic patterns can make challenges in their diagnosis and treatment. Oral cavity palisaded encapsulated neuroma is an uncommon lesion which occurs often on the hard palate. It is essential for oral pathologists to be familiar with its histopathology of this lesion is essential since many lesions are probably diagnosed microscopically as neurofibroma or schwannoma. Here, we report a case of oral palisaded encapsulated (solitary circumscribed) neuroma in an unusual site.

Corresponding Author: Mokhtari S., School of Dentistry, Tehran University of Medical Sciences, North Kargar Avenue, Tehran, Iran. Tel: +98-9125173177 Fax: +98-2122403194
Email: mokhtaris@sina.tums.ac.ir

Cite this article as: Atarbashi-Moghadam S., Lotfi A., Salehi Zalani S., Mokhtari S. Palisaded Encapsulated (Solitary Circumscribed) Neuroma of the Buccal Mucosa: a Rare Case. *J Dent Shiraz Univ Med Sci.,* 2017 December; 18(4): 314-317.

Introduction

Although the majority of palisaded encapsulated neuromas (PENs) or solitary circumscribed neuromas (SCNs) occur in the skin, oral cavity is the second most frequent site of involvement [1] and the hard palate is the most common site of occurrence. [2-4] Newman *et al.* [2] reported a case of multiple PENs on the face. The majority of oral PEN/SCN cases present as a slow- growing, painless, well-circumscribed, mobile submucosal mass with small size. Microscopically the lesion is composed of a proliferation of aligned infrequently palisaded Schwann cells with variable number of axons. A delicate layer of perineurium often incompletely wraps the lesion. [5-6] The cells show reactivity to S-100 protein; however, unlike other neural tumors, they are negative for glial fibrillary acidic protein (GFAP). [1] The capsular cells are positive for epithelial membrane antigen (EMA) and collagen type IV. [7]

Here, we present a case of PEN/SCN in an unusual site. Pathologists should be familiar with the histopathology of this lesion to avoid confusion with other peripheral nerve sheath tumors such as neurofibroma and schwannoma.

Case Report

A 58-year-old female was referred to a private oral and maxillofacial pathology center (Tehran, Iran) for evaluation of a slowly growing, painless, pedunculated, submucosal mass with a cylindrical shape in the left posterior buccal mucosa. The lesion had about 15 months duration (Figure 1).

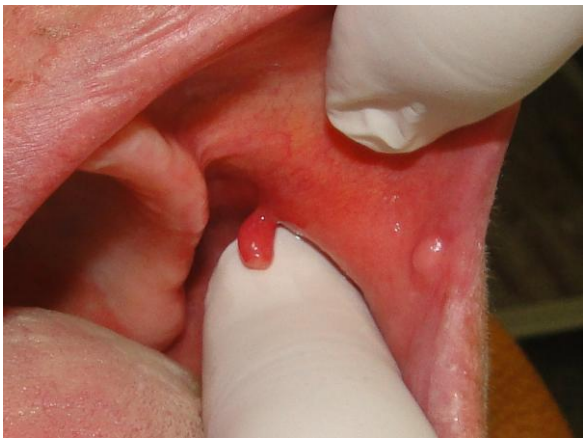

Figure 1: A pedunculated submucosal mass with a cylindrical shape in the left posterior buccal mucosa

The nodular mass had soft to elastic consistency measuring 8×4×4mm with an intact overlying submucosa. There was another tiny nonulcerated submucosal

mass in the left commissure. There was no history of previous trauma or other medical problems. Both lesions were completely excised with the clinical diagnosis as a reactive or benign neoplastic soft tissue lesion. Microscopic examination of the posterior buccal mucosa showed a well-circumscribed mass with fascicular proliferation of spindle cells showing tendency toward nuclear palisading. The nuclei did not exhibit any pleomorphism or mitotic activity. Apparent verocay bodies were not observed. The overlying epithelium was intact and chronic inflammatory cells were scattered (Figure 2, 3).

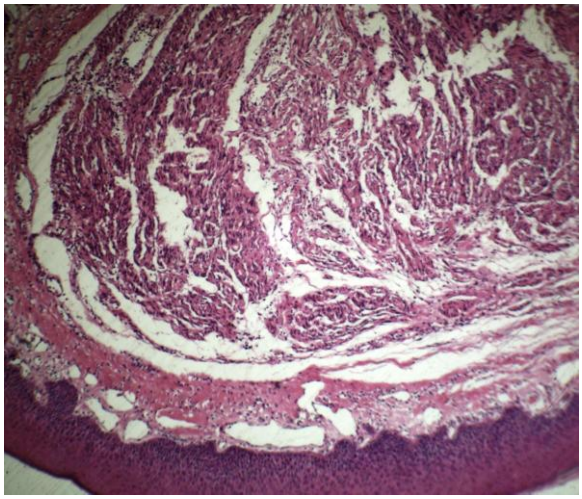

Figure 2: Microscopic examination of the lesion showed a well-circumscribed mass. The overlying epithelium was intact and chronic inflammatory cells were scattered.

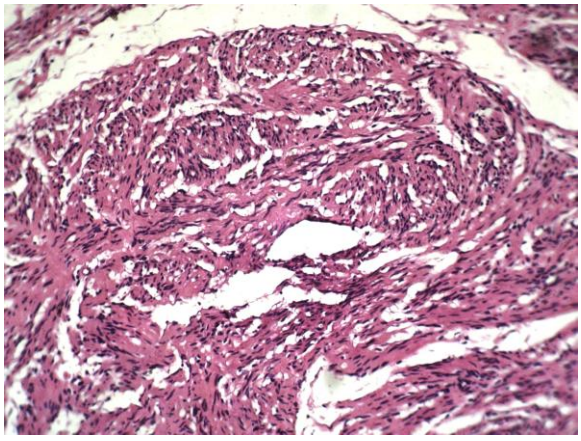

Figure 3: Fascicular proliferation of spindle cells showed tendency toward nuclear palisading. Apparent verocay bodies were not observed.

According to these features, the diagnosis of PEN (lobular pattern) was made. The immunohistochemical (IHC) analysis for S-100 and GFAP was performed to confirm the diagnosis. The lesion cells were strongly positive for S-100 (Figure 4) and negative for GFAP. Lesion of the commissure showed nodular proliferation of fibrous connective tissue. The diagnosis of focal fibrous hyperplasia was made for the buccal lesion. The patient has remained free of tumor for three years postoperatively.

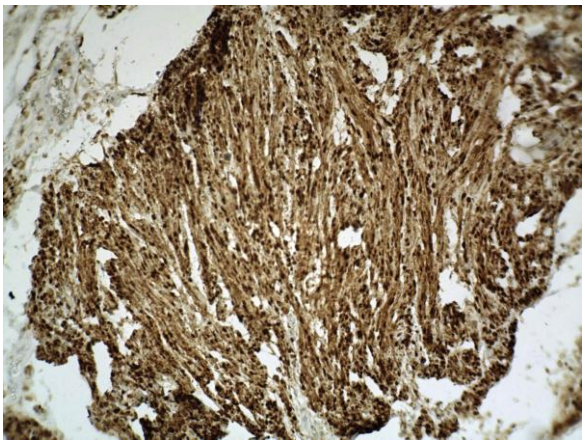

Figure 4: The lesion cells were strongly positive for S-100 protein.

Discussion

PEN/SCNs comprise 4.5% of oral soft tissue neoplasms. [8] Most patients are in middle age [7] and no sex predilection is observed. [9] However, in Koutlas *et al.* [1] study with large number of cases, the male to female ratio was 2.4. The vast majority of cases involve the palatal and gingival mucosa followed by tongue and labial mucosa. To date, about five cases of PEN in the buccal mucosa are reported in the literature. [1, 10] Palate, gingiva, tongue and lips have more superficial nerve branches than buccal mucosa. Therefore, buccal area is a very rare location of involvement. [10] Macroscopically, the lesions are usually unilobular and the overlying epithelium is atrophic. [1] However, the oral mucosa in the present case was normal. The differential diagnosis of PEN contains neurofibroma, schwannoma, amputation neuroma, and mucosal neuroma. [4] Differentiation from neurofibroma is critical since the latter is often associated with neurofibromatosis, and has a tendency for malignant conversion. [2] Neurofibroma is not encapsulated and shows hypocellular sheets, mucoid matrix with delicate collagen and mast cells in significant amounts. [1] Schwannoma has a complete capsule and reveals cellular fascicular Antoni A (with verocay bodies) and more definite palisading in the nuclei than that in PEN/SCN. [4] Traumatic (amputation) neuroma has a

history of trauma and the lesion is commonly painful with occasional burning sensation or paresthesia. [3,11] Traumatic neuroma shows perineural cells rimming discrete microfascicles, the larger quantity of interstitial collagen, mucoid matrix and myelin elements. [1] Moreover, absence of inflammatory cells helps to distinguish PEN from traumatic neuroma. [12]

Mucosal neuroma does not have capsule and palisading nuclei. It demonstrates nerve bundles in different sizes surrounded by normal connective tissue and is frequently associated with multiple endocrine neoplasia (MEN) type 2B. [4] The lesion cells are negative for GFAP. GFAP negative immunostaining may be useful to separate PEN/SCN from other peripheral nerve sheath tumors. GFAP might contribute to produce macro-complexes to start effective nerve renewal. [13]

A presentation of linear papules and nodules on the neck and forehead, similar to collagenoma, is also reported. [14-15] Therefore, clinicians should include PEN in the differential diagnosis of fleshy papules presenting on the nose and forehead for obtaining better detection of the tumor, which responds well to surgical excision. Generally, PEN/SCN is considered as a reactive hyperplastic course but the pathogenesis of this lesion remains unclear. Some authors suggest that it has a traumatic etiology. This concept is supported by the fact that incompletely removed PEN/ SCN does not recur. [1] Intraoral PEN/SCNs have an excellent prognosis; complete surgical excision is a treatment of choice and recurrence is rare. [2, 4] Laser therapy has also been used to manage the PEN. [6] This technique allows for adequate histological examination [16] and is ideal for most soft tissue excisions. [17]

Conclusion

In conclusion, PEN/SCNs may be clinically similar to reactive soft tissue lesions. Its presence, particularly near mucocutaneous junctions, necessitates definitive histologic diagnosis because it can mimic other tumors that have underlying systemic disease. [2] Moreover, since there are similarities and overlap in features with other neural tumors, IHC staining is needed to arrive at a definitive diagnosis in some cases. [7]

Conflict of Interest

None declared.

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