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CASE REPORT

Ulcerated bleeding palisaded encapsulated neuroma of the tongue

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Abstract

Introduction: Palisaded encapsulated neuroma (PEN) is a benign lesion of Schwann cells and its occurrence in the oral mucosa especially the tongue is very rare. *Case Report:* This article describes a case of a 41-year-old male, a chronic smoker with an actively bleeding, ulcerated, solitary, firm lesion on the lateral border of the tongue which had bled thrice before. A differential diagnosis of pyogenic granuloma, haemangioma, fibroma, nerve sheath tumour, salivary gland tumour and malignancy was made and surgically excised. Histopathology of the excised specimen revealed a well-circumscribed lesion with spindle-shaped cells arranged in interlacing fascicles and with the help of immunohistochemical markers confirmed it to be a PEN. *Discussion:* To our knowledge, this is the first description of an ulcerated PEN presented with an active bleed.

Keywords: Neuroma, cutaneous, tongue

INTRODUCTION

Palisaded encapsulated neuroma (PEN), also called solitary circumscribed neuroma (SCN) is a distinctive, benign neurogenic tumour occurring intraorally, the commonest site being the hard palate.[1] It is also termed as a 'cutaneous tumour' when it occurs on the facial skin and nasal fossae.[2] Rarely they are also reported to occur on the trunk, eyelids, extremities and glans penis with a mean age of occurrence between 37 to 54 years having no sex predilection.[3-6] Tumours arising from peripheral nerves in the oral and para-oral tissues are uncommon (22-25%) and include neurofibroma, schwannoma (neurilemmoma), mucosal neuroma associated with multiple endocrine neoplasia (MEN) IIb, nerve sheath myxoma and granular cell tumour.[7] Most of these tumours clinically mimic a benign fibroma or pyogenic granuloma (PG) and are often painless, superficially situated, solitary, firm and non-pigmented nodules.[8] With adequate surgical excision, which is the treatment of choice, no recurrences have been reported so far.[9]

CASE REPORT

A 41-year-old male patient, a chronic smoker with no comorbidities was seen in the emergency department with an active bleed from a lesion on the left lateral border of the tongue. The bleeding stopped with gentle pressure over the lesion and on enquiry he complained of a slow growing, painless swelling of 7 months duration with 3 episodes of spontaneous bleeding which stopped with firm pressure over the lesion. There was no history of any other similar lesions in the oral cavity, enlargement of neck nodes, symptoms or signs suggestive of a MEN syndrome or neurofibromatosis. General examination was unremarkable. Examination of the tongue revealed a solitary, non-tender, firm, nodular swelling with superficial mucosal ulceration on the left lateral border of the tongue measuring 1x1 cm with no peripheral induration/infiltration (Fig. 1A). Rest of the oral cavity and neck was normal. Considering the nature of lesion, a provisional diagnosis of pyogenic granuloma, haemangioma, fibroma, salivary gland tumour and malignancy was made. Routine blood investigations were unremarkable. He underwent excision of the lesion *in toto*.

Histopathological examination of the lesion showed a tumour completely circumscribed by a thin fibrous capsule and composed of diffuse and dense proliferation of spindle-shaped cells and scantly eosinophilic cytoplasm arranged in interlacing fascicles with indistinct cell borders. The nuclei, showing a parallel and tapering

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orientation within the fascicles was wavy and pointed. There was no atypia or mitotic figures, the stroma was interspersed with few capillaries and fibroblasts and the overlying epithelium being lined with para keratinised epithelium (Figs. 1B and 1C). IHC was positive for S-100 (Fig. 1D) and negative for smooth muscle actin (SMA), glial fibrillary acidic protein (GFAP) and desmin. These findings indicated it to be a palisaded encapsulated neuroma (PEN). He has been kept under close follow-up for the past 3 years and has no local recurrence.

DISCUSSION

PEN is a solitary, benign, cutaneous or mucosal tumour that often occurs on the face, oral cavity or close to the mucocutaneous junction.[1,2] Multiple lesions are rarely found and most of the tumours are nodular or unilobular in appearance, often painless and exhibit smooth contours. In the oral cavity, these tumours are commonly located on the palate, gingiva and the lip since these areas have superficially situated nerve branches compared to the buccal mucosa.[3-5] PEN of the tongue is rare and a bleeding tongue PEN has never been reported in the literature and this rare clinical presentation of our patient brought out plenty of differential diagnoses: Pyogenic granuloma, haemangioma, fibroma, salivary gland tumour and malignancy, but the diagnosis was made on histopathology of the excised specimen.

Various growth patterns like the epithelioid, fungating, plexiform, nodular and those in MEN type IIb have been observed among which the nodular type is most frequently seen as in our patient.[8] PEN shares histopathological features with neurofibroma and schwannoma and hence IHC markers help single out PEN from other neural tumours. Moreover absence of mast cells

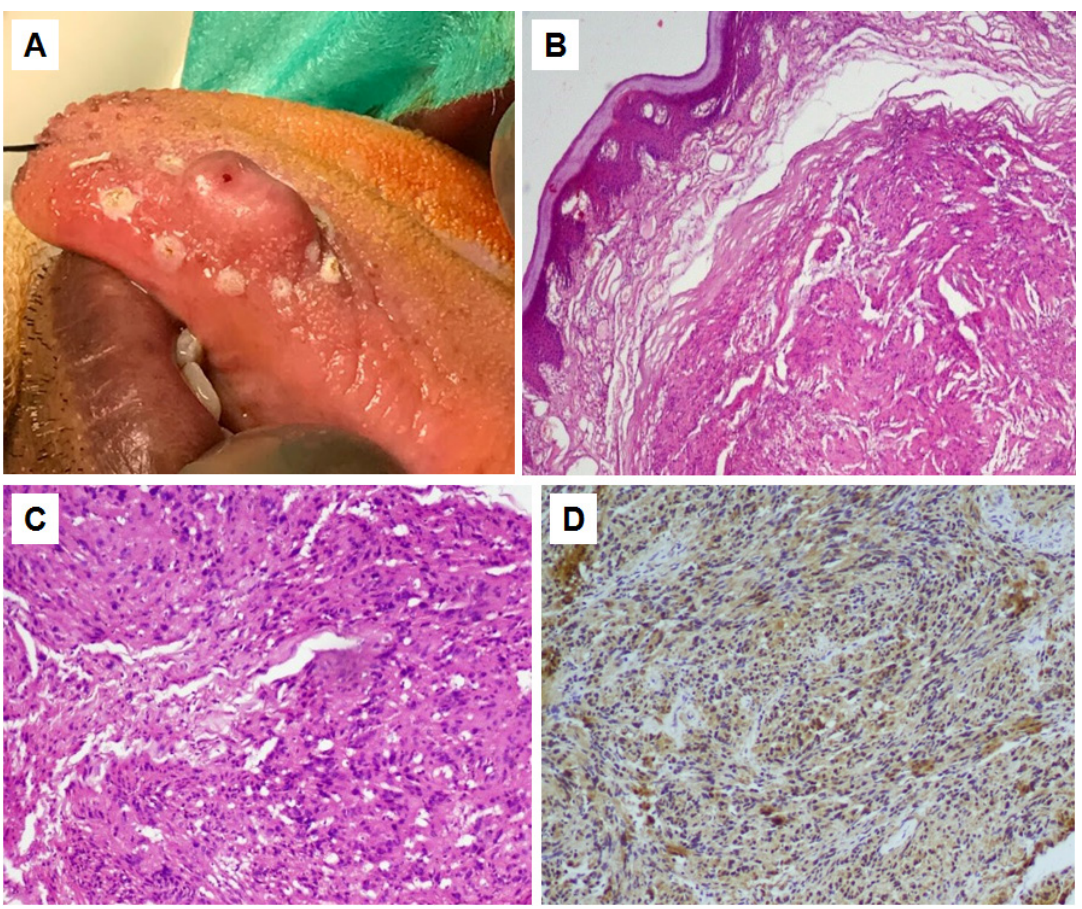

FIG. 1: A) Nodular, ulcerated swelling on the left lateral border of the tongue with recent bleed. B) Photomicrograph showing circumscribed well-encapsulated proliferating neoplastic cells (H&E, x40). C) Interlacing fascicles of neoplastic tumour cells (H&E, x100). D) Tumour cells are positive toward S-100 protein (x100).

and verocay bodies are additional features which aids in the diagnosis.[7-9] Differentiating features from these neural and smooth muscle tumours has been discussed in Table 1.

The pathogenesis of PEN remains a matter of debate. Though a traumatic aetiology has been proposed, in the vast majority of patients there is no history of trauma. Unlike traumatic neuroma in which myelination is prominent, PEN features only focal and poorly preserved myelin.[9] The absence of GFAP staining in PEN is intriguing and may highlight an altered Schwann cell phenotype. PEN appears to be a GFAP deficient non-myelin producing intraneural Schwann cell.[10] Thus the proposed pathophysiology of PEN was suggested to be a hamartomatous growth of Schwann cells which outgrow the axons with no atypical features.[11] PEN is an underdiagnosed and misdiagnosed tumour that can be easily excised (the treatment of choice) without recurrence.[7]

TABLE 1: Differential diagnosis and tumour characteristics of nodular lesions of the oral cavity.

Types of tumour	Clinical features	Histopathological features	Immunohistochemistry
Neurofibroma	- Soft, non-ulcerated asymptomatic nodules usually of the same color as the oral mucosa - Usually associated with neurofibromatosis or Von Recklinghausen's disease of the nerve	Wavy elongated cells, dark staining nuclei. Lack a capsule, contains mucopolysaccharide ground substance and have fewer axons with myelin sheath	S-100 (Positive) GFAP (Varies; usually positive)
Schwannoma	- Single circumscribed, slow growing nodule - usually asymptomatic	It is found subcutaneously, contain Antoni A and B type tissue with the presence of verocay bodies and do not have axons in the core. Perineural encapsulation is complete	S-100 (Positive) GFAP (Positive)
Traumatic neuroma	- Extremely rare, appear as a small nodule - Usually on the lip or tongue - Surface may be smooth and non-ulcerated	Axonal and Schwann cell proliferation with the addition of inflammatory cells	S-100 (Positive) EMA (Positive)
Mucosal neuroma	- Multiple yellowish white sessile painless nodule - Associated with MEN IIb syndrome	Partially encapsulated aggregation or proliferation of nerves. Individual nerves flow in fascicles of 2-3 fibres and are histologically normal. Inflammatory cells and dysplasia is usually absent	S-100 (Positive) Positive for collagen type IV, vimentin, NSE and neural filaments
Leiomyoma	- A slow growing painless lesion, superficial and pedunculated - Resembles normal mucosa in colour and texture	Well-circumscribed tumours, consisting of interlacing bundles of spindle-shaped smooth muscle cells with fibrous connective tissue	Positive for desmin, myosin, smooth muscle actin and vimentin

CONCLUSION

PEN is a relatively rare entity which can occur in any part of the body and is usually slow growing and painless. This lesion being a clinical and histopathological mimicker of various benign tumours, and immunohistochemical study will aid in the diagnosis. However, there are no reports of malignant potential, transformation or recurrence following complete excision. The surgeon and the pathologist should keep a diagnosis of PEN in mind while dealing with nodular lesions of the oral cavity.

REFERENCES

1. Lombardi T, Samson J, Kuffer R. [Solitary circumscribed neuroma (palisaded encapsulated neuroma) of the oral mucosa]. Ann Dermatol Venereol. 2002; 129(2): 229-32.
2. Lanoue J, Singh R, Chen C, Phelps R. Multiple cutaneous linear neuromas. JAAD Case Rep. 2015; 1: 315-8.
3. Beutler B, Cohen PR. Palisaded Encapsulated Neuroma of the Trunk: A Case Report and Review of Palisaded Encapsulated Neuroma. Cureus. 2016; 8: e726.
4. Surapaneni KR, Phelps PO, Potter HD. Palisaded Encapsulated Neuroma of the Eyelid. Ophthalmology. 2015; 122: 1554.
5. Omori Y, Tanito K, Ito K, Itoh M, Saeki H, Nakagawa H. A pediatric case of multiple palisaded encapsulated neuromas of the palms and soles. Pediatr Dermatol. 2014; 31: e107-9.
6. Navarro M, Vilata J, Requena C, Aliaga A. Palisaded encapsulated neuroma (solitary circumscribed neuroma) of the glans penis. Br J Dermatol. 2000; 142: 1061-2.
7. Misago N, Toda S, Narisawa Y. The relationship between palisaded encapsulated neuroma and the mucocutaneous neuroma seen in multiple endocrine neoplasia 2b syndrome: a histopathologic and immunohistochemical study. Am J Dermatopathol. 2014; 36(7): 562-9.
8. Mortazavi N, Gholami A, Amini Shakib P, Hosseinkazemi H. Palisaded Encapsulated Neuroma of the Tongue Clinically Mimicking a Pyogenic Granuloma: A Case Report and Review of Literature. J Dent (Tehran). 2015; 12(7): 537-41.
9. Hall LD, Ferringer T. Palisaded encapsulated neuroma. Cutis. 2013; 92: 167, 177-8.
10. Newman MD, Milgraum S. Palisaded encapsulated neuroma (PEN): an often misdiagnosed neural tumor. Dermatol Online J. 2008; 14: 12.
11. Dover JS, From L, Lewis A. Palisaded encapsulated neuromas. A clinicopathologic study. Arch Dermatol. 1989; 125: 386-9.