

Neurothekeoma of Gingiva in a Pediatric Patient: Report of a Case and Literature Review

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ABSTRACT

Nerve sheath myxoma (NSM) or neurothekeoma (NTK) is an uncommon benign tumor of nerve sheath origin. On the basis of the amount of myxoid content it is classified into classical, mixed and cellular variants. Among the 41 reported intraoral cases only six cases have been previously reported in the pediatric age group patients. Out of these six none of the case has been reported in the gingiva. We report a first case of gingival neurothekeoma in an 11-year-old child which was provisionally diagnosed as pyogenic granuloma. We also review the oral neurothekeoma in pediatric age group patients and discuss the histopathological provisional diagnosis.

Keywords: Neurothekeoma, Nerve sheath myxoma, Gingiva, Pediatric age.

How to cite this article: Bajpai M, Rahman F, Girish KL, Pardhe N. Neurothekeoma of Gingiva in a Pediatric Patient: Report of a Case and Literature Review. *Oral Maxillofac Pathol J* 2015;6(1): 575-577.

Source of support: Nil

Conflict of interest: None

INTRODUCTION

Nerve sheath myxoma (NSM) is an uncommon benign soft tissue tumor of peripheral nerves. The lesion was first described in 1969 by Harkin and Reed as nerve sheath myxoma.¹ However, the term neurothekeoma (NTK) was first coined by Helvig.² Although the lesion is commonly seen in the subcutaneous tissue of head and neck region but rarely seen intraorally.³ Oral NTK in children is very unusual and has not been reported in oral location other than tongue, lip, palate and floor of the mouth.⁴ According to our best knowledge, this is the first case of gingival NTK reported in a child.

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CASE REPORT

A healthy 11-year-old child was referred to the department of oral medicine and radiology, for the evaluation of an overgrowth overlying an unerupted maxillary right canine. Intraoral examination revealed a dome shaped sessile mass of 2 × 1.5 cm arising from the maxillary gingival region which was pinkish in color (Fig. 1). An intraoral periapical radiograph revealed no association of the lesion with the bone (Fig. 2).

On the basis of clinical and radiographic features the provisional diagnosis of pyogenic granuloma was given. Excisional biopsy was performed (Fig. 3) and the excised tissue sent to the department of oral and maxillofacial pathology, for histopathological analysis (Fig. 4). Histopathological examination revealed distinct lobules with fibrocellular connective tissue stroma showing



Fig. 1: Dome-shaped sessile mass arising from the maxillary gingival region

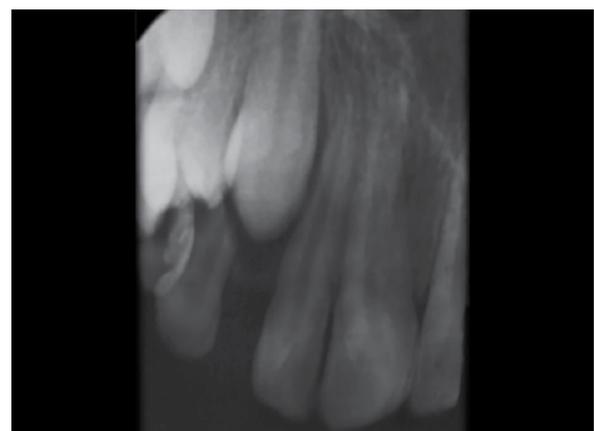


Fig. 2: Intraoral periapical radiograph revealed no association of the lesion with the bone



Fig. 3: Intraoperative view



Fig. 4: Gross tissue

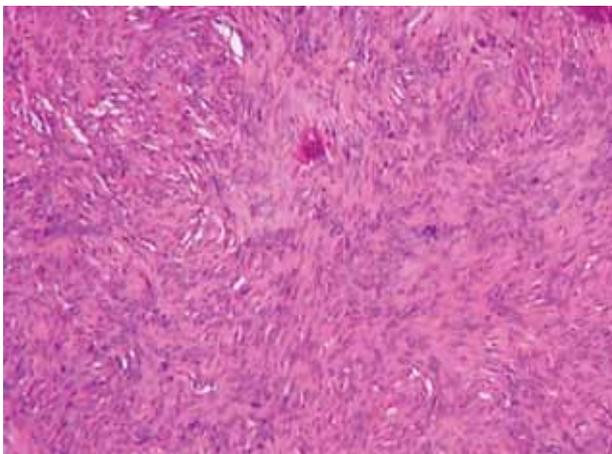


Fig. 5: Lobules of spindle and epithelioid cells in surrounding myxoid stroma (hematoxylin and eosin, 10x)

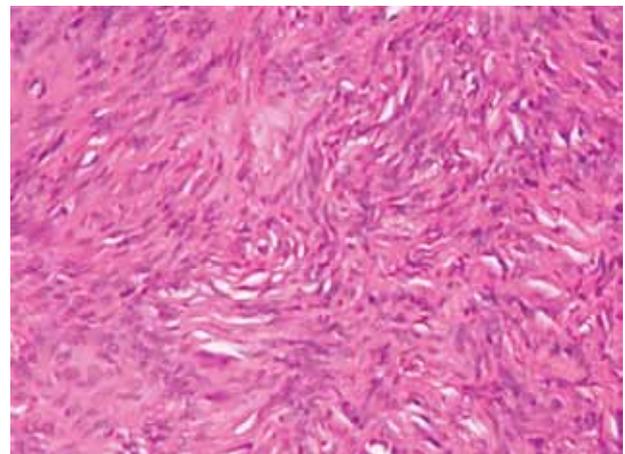


Fig. 6: Spindle/stellate cells with minor atypia

numerous spindle and epithelioid cells with variable amount of myxoid matrix (Figs 5 and 6).

DISCUSSION

Nerve sheath myxoma is a benign peripheral nerve sheath tumor that rarely occurs in the oral cavity.⁵ Among 41 reported cases of intraoral NSM, only six cases have been reported in the pediatric age group patients. Tongue (3/6) is the commonest site of occurrence palate, lip and floor of the mouth accounts for one case each⁶ (Table 1). The present case is the first case reported in gingiva. Based

on the amount of myxoid stroma, NSM is generally classified into classical, mixed and cellular type.⁷ Histopathologically, the classic type present as a multinodular lesion with an abundant myxoid matrix outlined by thin condensed fibrous tissue.⁴ The lesion usually show spindle or stellate-shaped cells typically grouped in cords or nests scattered in a myxomatous area.^{4,8} The cellular type is characterized by spindle and epithelioid

Table 1: Oral neurothekeoma reported in pediatric age group patients⁶

Case	Author	Gender	Age	Location	Size (cm)
1	Breur et al ⁹	F	12	Tongue	1.5 × 2
2	Peñoarocha et al ¹⁰	F	1	Tongue	3 × 2.5
3	Wright et al ¹¹	F	15	Palate	NR
4	Makino et al ⁴	M	8	Tongue	0.8 × 0.5
5	Nishioka et al ¹²	M	2	Lower lip	0.7 × 0.7
6	Emami N et al ¹³	F	15	Floor of the mouth	0.8

F: Female; M: Male; NR: Not reported

Table 2: Histopathological differential diagnosis of neurothekeoma¹⁴

Differential diagnosis	Characteristic features
Neurilemmoma	Encapsulated and shows Antoni A and B architecture with verocay bodies
Neuromas	True neuromas show nerve axons
Myxoid neurofibroma	The cells are more slender and closely aggregated
Mucosal melanoma	Shows positive immunohistochemical expression for HMB 45
Focal mucinosis	Well-circumscribed lesion and does not show spindle and epithelioid cells in lobulated fashion
Soft tissue myxoma	Shows blending in the surrounding tissue in contrast to neurothekeoma which is well-demarcated from the surrounding tissue



cells proliferating in fibromyxoid stroma.⁴ Histology of mixed type frequently shows an aggregate of stellate and spindle-shaped cells with ovoid nuclei scattered in the connective tissue stroma.⁵ The present case belongs to the cellular pattern and shows spindle/stellate and epithelioid cells arranged in a lobular pattern in a fibromyxoid stroma. Immunohistochemically, the cells are found to be positive for S-100.^{3,4,7} Histopathological differential diagnosis of NSM includes neural and non-neural lesions (Table 2).

Local excision is the treatment of choice for NSM. No recurrence has been reported after local excision.⁸ The present case was also treated by the same approach and the 1 year follow-up period was uneventful.

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