Neurothekeoma of Oral Cavity: A Rare Case Report

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Abstract

Neurothekeoma is a benign soft tissue tumor with a clinical presentation as a solitary slow-growing painless mass. It is seen most commonly in the central area of the face, neck, and upper extremities. The mean age of occurrence is 25 years with a slight female predilection. Three histologic variants include myxoid, mixed, and cellular. The histogenesis of this tumor is controversial. The occurrence in the oral cavity is extremely rare. This article describes a case report of cellular neurothekeoma in the tongue of a 51-year-old male patient. The lesion was excised during biopsy and has shown no recurrence to date. This is the 7th case reported in the literature on cellular neurothekeoma presenting in the oral cavity.

Key words: Nerve sheath myxoma, Neurothekeoma, Oral cavity, Tongue

INTRODUCTION

Neurothekeoma is an uncommon benign soft tissue tumor. Gallagher and Helwig coined the term neurothekeoma.¹ The term cellular neurothekeoma was coined by Rosati et al. in 1986.² It is seen most commonly in the central area of the face, neck, and upper extremities. The mean age of occurrence is 25 years with a slight female predilection. Three histologic variants include myxoid, mixed and cellular.³ Histopathologically, these lesions show a circumscribed tumor mass composed of epithelioid and spindle cells, arranged in well-formed micronodules.⁴ A recent study of 37 cellular neurothekeoma showed cytological atypia in about 50% of cases.⁵ The histogenesis of this tumor is controversial. Earlier it was believed to be a type of nerve sheath myxoma.⁶ Gene expression profile study of neurothekeomas have shown that it may be a variant of fibrous histiocytomas.⁷

Oral involvement is extremely rare. The most common intraoral site is tongue.⁸ This article describes a case report of cellular neurothekeoma in the tongue of a 51-year-old male patient.

CASE REPORT

A 51-year-old male patient presented with the chief complaint of swelling in the right side of the tongue since 1 year. The swelling was asymptomatic, insidious in onset, first noticed 1 year back, and has slowly increased to its present size. No relevant medical or family history was present.

On examination, a swelling of size 1 cm × 1 cm × 0.4 cm was noticed on the right side of the dorsum of the tongue. It was firm in consistency, non-fixed with limited mobility. The overlying mucosa appeared relatively normal. The provisional diagnosis was fibroepithelial hyperplasia or granular cell tumor. The swelling was excised during biopsy.

Microscopic examination of hematoxylin and eosin stained section showed stratified squamous epithelium and an underlying lamina propria with tumor mass (Figure 1). The proliferating tumor mass was arranged as lobules (Figure 2). Under ×40 magnification, ovoid to spindle cells and epithelioid cells with bland, vesicular nuclei, and a light eosinophilic cytoplasm, arranged like staves of a barrel were seen (Figure 3). Immunohistochemical analysis showed positivity of tumor cells for NKI/C3, vimentin; and were
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DISCUSSION

Neurothekeoma is a benign cutaneous tumor with rare mucosal involvement. Oral involvement of neurothekeoma is extremely rare with only six reported cases of cellular neurothekeomas. A slight female predilection was reported. Tongue was the most common site.

In earlier days, the term nerve sheath myxoma and neurothekeoma were used interchangeably. Husain et al. considered these tumors as either ends of the morphologic spectrum of neurothekeoma. Various immunohistochemical studies showed neural differentiation of myxoid neurothekeoma.

The histogenesis of cellular neurothekeoma is controversial. Fetch et al. proposed an origin from fibroblastic cells with the ability to differentiate into myofibroblasts and a tendency to recruit histiocytic cell. Sheth et al. studied microarray-based gene expression profile of dermal schwannomas, dermal nerve sheath myxomas, cellular fibrous histiocytomas and myxoid/mixed/cellular neurothekeomas. They found that neurothekeomas and cellular fibrous histiocytomas showed upregulation of genes encoding various metalloproteinases and glycoproteins involved in growth and remodeling of extracellular matrix; whereas genes encoding neuronal cell intercellular signaling were differentially expressed between nerve sheath myxomas and schwannomas.

Histopathologically, neurothekeomas are seen as multinodular, lobular, or plexiform patterns surrounded by bands of dense collagen. Cells are epithelioid or spindle-shaped with light eosinophilic cytoplasm, and contain bland, ovoid nuclei. Myxoid areas are frequently seen in the stroma which may mimic nerve sheath myxoma. Osteoclast like giant cells may be seen.

Neurothekeomas are variably immunoreactive for smooth muscle actin, PGP 9.5, NK1/C3, CD10, CD68, microphthalmia transcription factor, podoplanin; and negative for S100, glial fibrillary acidic protein, and melan A.

Complete excision is the treatment of choice. Recurrence may occur with incomplete removal. The differential diagnosis includes plexiform fibrohistiocytic tumors, reticulohistiocytoma, epithelioid fibrous histiocytoma, and melanocytic tumors. Plexiform fibrohistiocytic tumors show diffuse, nodular, and plexiform growth pattern of either spindle or epithelioid cells, but plexiform pattern being the predominant. Giant cells are more in number and with more nuclei than cellular neurothekeoma. Expression of microphthalmia transcription factor...
helps in differentiating neurothekeoma from plexiform fibrohistiocytic tumors. Reticulohistiocytoma lacks the plexiform or whorling growth pattern seen in neurothekeoma and the epithelioid cells are CD163 positive. The epithelioid variant of fibrous histiocytoma shows a diffuse pattern of epithelioid fibroblasts rather than the multi-nodular pattern of neurothekeoma. Melanocytic tumors show positive expression of S100 and melanocytic markers such as HMB45 and Melan-A.

CONCLUSION

The present case was that of a male patient with tongue involvement. Histopathology of our case was that of a clear-cut cellular neurothekeoma. The lesion was excised during biopsy. Patient is on follow up and has shown no recurrence to date. This is the 7th case reported in the literature on cellular neurothekeoma presenting in the oral cavity.

REFERENCES


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