Case Report

DOI: 10.17354/ijss/2015/140

Neurothekeoma of Oral Cavity: A Rare Case Report

V T Beena[1], P V Deepthi[2], S K Padmakumar[3], R Sivakumar[4]

[1]Professor & Head, Department of Oral Pathology and Microbiology, Government Dental College, Trivandrum, India, [2]Post-graduate Student, Department of Oral Pathology and Microbiology, Government Dental College, Trivandrum, India, [3]Assistant Professor, Department of Oral Pathology and Microbiology, Government Dental College, Trivandrum, India, [4]Assistant Professor, Department of Oral Pathology and Microbiology, Government Dental College, Trivandrum, India

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 (1.8: 1).[3] Three histologic variants include myxoid, mixed and cellular.[4] Histopathologically, these lesions show a circumscribed tumor mass composed of epithelioid and spindle cells, arranged in well-formed micronodules.[5] A recent study of 37 cellular neurothekeoma showed cytological atypia in about 50% of cases.[6] The histogenesis of this tumor is controversial. Earlier it was believed to be a type of nerve sheath myxoma.[7] Gene expression profile study of neurothekeomas have shown that it may be a variant of fibrous histiocytomas.[8]

Oral involvement is extremely rare. The most common intraoral site is tongue.[9] This article describes a case report of cellular neurothekeoma in the tongue of a 51-year-old male patient.

Access this article online

www.ijss-sn.com

Month of Submission : 01-2015
Month of Peer Review : 02-2015
Month of Acceptance : 02-2015
Month of Publishing : 03-2015

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

Corresponding Author: Dr. P V Deepthi, Department of Oral Pathology and Microbiology, Government Dental College, Trivandrum, India. Phone: +91 9496370847. E-mail: deepthipv27@gmail.com

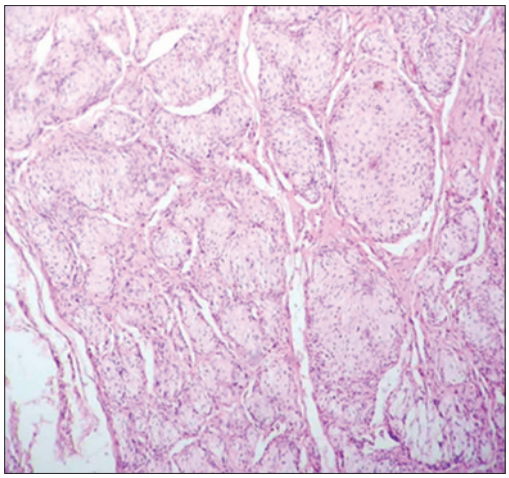

Figure 1: Tumor mass separated from the epithelium by a condensed fibrous connective tissue, H and E stain, under ×4 magnification

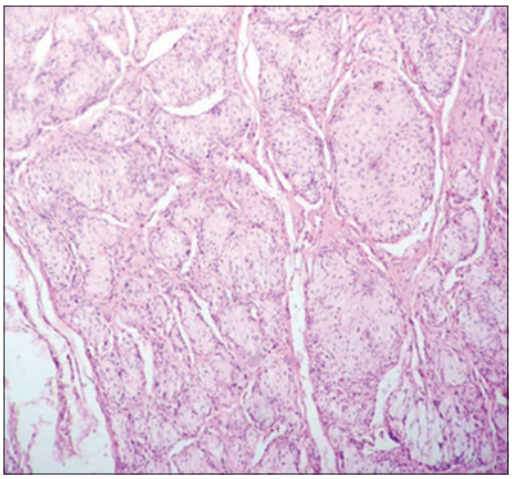

Figure 2: Lobules of tumor cells separated by fibrous connective tissue, H and E stain, under ×10 magnification

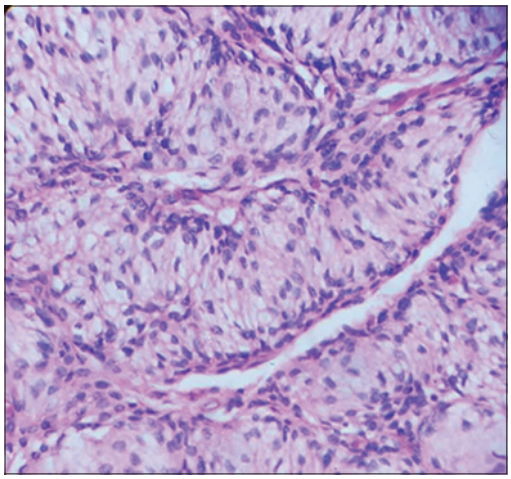

Figure 3: Ovoid to spindle cells with bland nucleus and light eosinophilic cytoplasm, H and E, under ×40 magnification

negative for S100. A diagnosis of cellular neurothekeoma was arrived.

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.[5,10] 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.[7] Various immunohistochemical studies showed neural differentiation of myxoid neurothekeoma.[11,12]

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.[2] 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.[8]

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.[13]

Neurothekeomas are variably immunoreactive for smooth muscle actin, PGP 9.5, NKI/C3, CD10, CD68, microphthalmia transcription factor, podoplanin; and negative for S100, glial fibrillary acidic protein, and melan A.[14,15]

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.[13] 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.[16] Melanocytic tumors show positive expression of S100 and melanocytic markers such as HMB45 and Melan-A.[13]

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

1. Gallager RL, Helwig EB. Neurothekeoma – a benign cutaneous tumor of neural origin. Am J Clin Pathol 1980;74:759-64.
2. Fetsch JF, Laskin WB, Hallman JR, Lupton GP, Miettinen M. Neurothekeoma: An analysis of 178 tumors with detailed immunohistochemical data and long-term patient follow-up information. Am J Surg Pathol 2007;31:1103-14.
3. Rosati LA, Fratamico FC, Eusebi V. Cellular neurothekeoma. Appl Pathol 1986;4:186-91.
4. Akhtar K, Zaheer S, Ray PS, Sherwani RK. Myxoid neurothekeoma: A rare soft tissue tumor of hand in a male toddler. Niger J Surg 2013;19:32-4.
5. Emami N, Zawawi F, Wakim RY, Nahal A, Daniel SJ. Oral cellular neurothekeoma. Case reports otolaryngol. 2013. Available from: http://www.dx.doi.org/10.1155/2013/935435 [Last accessed on 23 Dec 2014].
6. Stratton J, Billings SD. Cellular neurothekeoma: Analysis of 37 cases emphasizing atypical histologic features. Mod Pathol 2014;27:701-10.
7. Husain S, Silvers DN, Halperin AJ, McNutt NS. Histologic spectrum of neurothekeoma and the value of immunoperoxidase staining for S-100 protein in distinguishing it from melanoma. Am J Dermatopathol 1994;16:496-503.
8. Sheth S, Li X, Binder S, Dry SM. Differential gene expression profiles of neurothekeomas and nerve sheath myxomas by microarray analysis. Mod Pathol 2011;24:343-54.
9. Nishioka M, Aguirre RL, Ishikawa A, Nagumo K, Wang LH, Okada N. Nerve sheath myxoma neurothekeoma arising in the oral cavity: Histological and immunohistochemical features of 3 *case*s. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2009;107:e28-33.
10. Vered M, Fridman E, Carpenter WM, Buchner A. Classic neurothekeoma nerve sheath myxoma and cellular neurothekeoma of the oral mucosa: Immunohistochemical profiles. J Oral Pathol Med 2011;40:174-80.
11. Laskin WB, Fetsch JF, Miettinen M. The "neurothekeoma": Immunohistochemical analysis distinguishes the true nerve sheath myxoma from its mimics. Hum Pathol 2000;31:1230-41.
12. Fetsch JF, Laskin WB, Miettinen M. Nerve sheath myxoma: A clinicopathologic and immunohistochemical analysis of 57 morphologically distinctive, S-100 protein- and GFAP-positive, myxoid peripheral nerve sheath tumors with a predilection for the extremities and a high local recurrence rate. Am J Surg Pathol 2005;29:1615-24.
13. Goldblum JR, Folpe AL, Weiss SW. Benign Fibrohistiocytic and Histiocyticntumors. In: Goldblum JR, Folpe AL, Weiss SW., editors. Enzinger and Weiss's Soft Tissue Tumors. 5th ed. China: Elsevier; 2013. p. 359-63.
14. Hornick JL, Fletcher CD. Cellular neurothekeoma: Detailed characterization in a series of 133 cases. Am J Surg Pathol 2007;31:329-40.
15. Kaddu S, Leinweber B. Podoplanin expression in fibrous histiocytomas and cellular neurothekeomas. Am J Dermatopathol 2009;31:137-9.
16. Miettinen M. Childhood fibroblastic and myofibroblastic proliferations of variable biologic potential. In: Miettinen M, editor. Modern Soft Tissue Pathology: Tumors and Non-Neoplastic Condition. New York: Cambridge University Press; 2010. p. 285-9.

How to cite this article: Beena VT, Deepthi PV, Padmakumar SK, Sivakumar R. Neurothekeoma of Oral Cavity: A Rare Case Report. Int J Sci Stud 2015;2(12):208-210.

Source of Support: Nil, **Conflict of Interest:** None declared.