# SPINDLE CELL TUMOR OF NEURAL ORIGIN MANIFESTING IN ORAL CAVITY: A CASE REPORT WITH REVIEW

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**ABSTRACT**

*Tumors composed of neoplastic proliferation of spindle shaped cells can present in oral cavity originating from epithelial, mesenchymal, or even odontogenic tissues. Forty years old female presented with a history of painful slow-growing swelling on lingual aspect of lower left first molar. It was one year old. Radiological findings were inconclusive and excisional biopsy of soft tissue mass was taken. The tissue was sent for histopathology and diagnosis of spindle cell neoplasm of neural origin was established after histopathological and immunohistochemical analysis. This case report also briefly reviews spindle cell neoplasms of neural origin that can manifest in oral cavity.*

**Key Words:** *Spindle cell tumor, neural tumor, oral cavity, S100, schwannoma, neurofibroma, traumatic neuroma, MPNST, GCT.*

## INTRODUCTION

A wide array of both benign and malignant spindle cell lesions can occasionally manifest in oral cavity and present a diagnostic challenge to oral pathologists owing to overlapping histopathological findings. Immunohistochemical analysis is an indispensable aid in differentiating these tumors with similar morphology but a heterogeneous origin and subsequently variable clinical management and prognosis.[1] A case report of spindle cell neoplasm of neural origin developed after extraction of left mandibular first molar is described. It is followed by a brief review of spindle cell tumors of neural origin that may occasionally manifest in oral cavity. The aim of presenting this case report was to highlight the significance of histopathological evaluation, particularly the use of immunohistochemistry in exact characterization of clinically indistinguishable lesions.

## CASE REPORT

A 40 year old female presented in the oral surgery outpatient department at Dow International Medical and Dental College, Ojha campus with the complaint of slowly growing painful swelling on lingual aspect of left mandibular first molar region following its extraction during the past one year. Patient's orthopantomogram (OPG) was acquired and was unremarkable. Upon excisional biopsy, a light brown irregular soft tissue mass was discovered attached to lingual aspect of premolar root. A provisional diagnosis of fibrous tissue overgrowth was made and the excised mass was sent for histopathology.

Biopsy revealed focally ulcerated stratified squamous epithelium lined tissue with abundant blood vessels exhibiting aberrant proliferation of plump spindle shaped cells. No evidence of malignancy or mitotic figures were found. A panel of immunohistochemical profile was obtained for exact categorization. Diagnosis of spindle cell neoplasm of neural origin was established by strong, diffuse positive reactivity against S100 and negative reactivity for both CD68 and smooth muscle actin (ASMA) (Fig 1).

## DISCUSSION

The spectrum of neoplasms that may present as spindle cell tumors in oral cavity encompasses a broad range of both malignant and benign lesions of heterogeneous origin. These tumors, indistinguishable from each other clinically also present a diagnostic challenge to the pathologist because of similar histological picture on routine hematoxylin and eosin stained slides. In the reported case, we faced the same problem. Once we had established the diagnosis of a benign spindle cell lesion, exact characterization of cell of origin could not be made without the aid of immunohistochemistry. Spindle cell neoplasms may be derived from epithelial, mesenchymal, or odontogenic tissues.[2]

A classification based on tissue of origin was proposed by Shamim[3] that categorized spindle cell neoplasms encountered in oral cavity into tumors of neural, myofibroblastic, vascular, epithelial, odontogenic, and miscellaneous origins.

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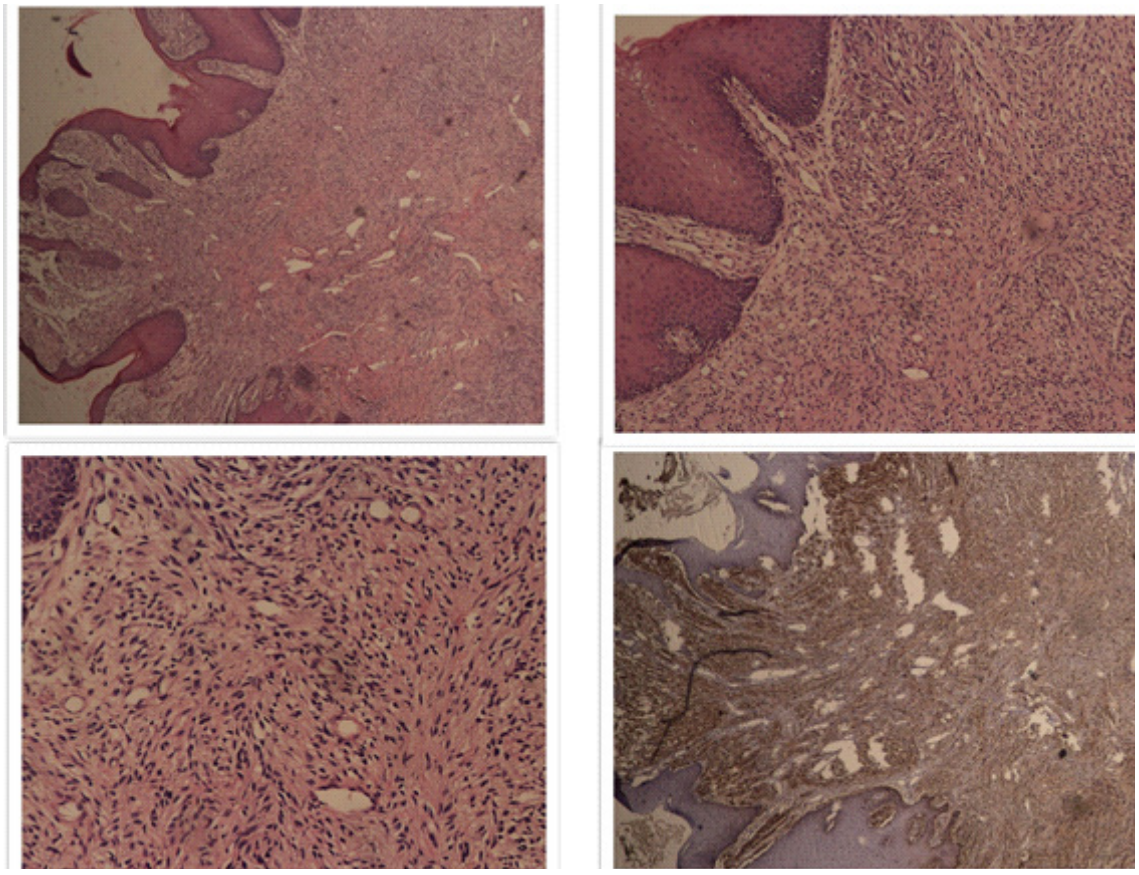

Fig 1: Hematoxylin and eosin stained sections showing stratified squamous epithelium lined tissue with abundant proliferation of plump spindle cells and numerous small blood vessels in underlying lamina propria. Immunohistochemical stain showing diffuse strong positive reactivity against S100.

Tumors of neural origin, as in presented case, are rare in oral cavity.[4] They can be differentiated from other spindle cell tumors by positive reactivity for neural markers, particularly S-100, as done in this case. Exact characterization requires amalgamation of clinical, histopathological, and immunohistochemical information. Due to rarity of their occurrence, spindle cell tumor was not considered a differential upon clinical evaluation and the specimen was sent for histopathology being anticipated a benign fibrous growth. A brief review of spindle cell neoplasms of neural origin is presented below.

**Spindle cell neoplasms of neural origin**

Peripheral nerve tumors that may manifest in oral cavity includes several differentials such as schwannoma (also known as neurilemmoma or peripheral nerve sheath tumor), neurofibroma, neurinoma in association with multiple endocrine neoplasia, palisaded encapsulated neurinoma, traumatic neuroma and granular cell tumor.[2,5] Immunohistochemical reactivity against S-100 and neuron-specific enolase is an essential tool to differentiate these tumors from spindle cell neoplasms of other origin (myofibroblastic tumors, tumors of muscle tissue origin, and fibroblastic tumors).

**Schwannoma**

Schwannoma, also known as neurilemmoma, is a benign, well-circumscribed, encapsulated tumor of myelin forming Schwann cells. The tumor manifests as a solitary, slow-growing, asymptomatic firm mass with smooth surface.[6] Schwannoma is a rare occurrence in oral cavity (1% of all extracranial schwannomas) with tongue being most frequently involved and retro-molar region being the least common site.[7] Treatment of choice for schwannoma is surgical excision.[8]

**Neurofibroma**

Neurofibroma is also a benign nerve sheath tumor of heterogeneous origin (schwann cells and perineural fibroblasts).[9] Unlike Shwannoma, these tumors are poorly circumscribed with potential for local invasion. Neurofibromas may be solitary (6.5% incidence in oral cavity) or multiple as part of neurofibromatosis (NF). Malignant transformation into sarcomas occur in 5-15% of neurofibromas associated with NF-1. Surgical excision is the most frequently used management of solitary oral neurofibromas with rare recurrence.[10] Diode laser has recently been reported to be an effective novel therapeutic alternative to surgical excision.[11]

**Malignant peripheral nerve sheath tumor (MPNST)**

Malignant peripheral nerve sheath tumor (MPNST) is a sarcoma originating from peripheral nerves occurring rarely in oral cavity with only 11 cases reported

in lower labial mucosa to date in English literature.[12] Microscopically tumor comprises of dense fascicles of spindle shaped cells exhibiting evident nuclear pleomorphism and mitotic figures. Surgical excision is the advocated management without metastatic spread. In patients presenting with advanced stage with metastatic deposits radiotherapy improves survival.[13]

### Traumatic neuroma

A reactive proliferation rather than a true neoplasm, traumatic neuroma can arise secondary to trauma to a nerve bundle. The lesion represents frustrated attempt to repair the damaged nerve in an exaggerated manner comprising of nerve tissue hyperplasia.[14] Like other spindle cell neoplasms, traumatic neuroma is rare in oral cavity occurring usually as a solitary nodule involving soft tissue over mental foramen, lower lip, or tongue.[15] A differentiating clinical feature of traumatic neuroma from other neurogenic tumors is that it is symptomatic manifesting as anesthesia, dysesthesia, or pain. Simple excision with electric scalpel is the recommended effective management.[15] In presented case, patient presented with a history of extraction at tumor site and also complaint of pain, features that are shared with traumatic neuroma.

### Granular cell tumor

Granular cell tumor (GCT) is a soft tissue neoplasm originally believed to arise from muscle cell precursors (myoblasts) but currently the cell of origin is recognized to be Schawnn cells. GCTs can occur anywhere in the body with a predilection for occurrence on tongue. Histopathologically, the tumor is well-circumscribed but lacking a true capsule. Pseudoepitheliomatous hyperplasia in overlying epithelium is well-established phenomenon in GCTs but underlying pathogenesis is still poorly understood.16 Treatment of choice is surgical excision with safe margins.[17]

## REFERENCES


1 Viswanathan S, Rahman K, Pallavi S, Sachin J, Patil A, Chaturvedi P, et al. Sarcomatoid (spindle cell) carcinoma of the head and neck mucosal region: a clinicopathologic review of 103 cases from a tertiary referral cancer centre. Head and neck pathology. 2010;4(4):265-75.

2 Shamim T. The Spindle Cell Neoplasms of the Oral Cavity. Iranian journal of pathology. 2015;10(3):175.

3 Shamim T. A simple working type classification proposed for the spindle cell neoplasms of the oral cavity. Journal of cytology/ Indian Academy of Cytologists. 2013;30(1):85.

4 Venkataswamy A, Ranganath S, Challapalli S, Mohanthy L. Nonsyndromic solitary neurofibromas in the oral cavity: Case series and literature review. Journal of Indian Academy of Oral Medicine and Radiology. 2016 January 1, 2016;28(1):52-56.

5 Baranović M, Macan D, Begović E, Lukšić I, Brajdić D, Manojlović S. Schwannoma with secondary erosion of mandible: case report with a review of the literature. Dentomaxillofacial Radiology. 2006;35(6):456-60. PubMed PMID: 17082339.

6 Nishijima Sakanashi E, Sonobe J, Chin M, Bessho K. Schwannoma Located in the Upper Gingival Mucosa: Case Report and Literature Review. Journal of Maxillofacial and Oral Surgery. 2015;14(1):222-25.

7 Rathore AS, Srivastava D, Narwal N, Shetty DC. Neurilemmoma of Retromolar Region in the Oral Cavity. Case Reports in Dentistry. 2015;2015:4.

8 Handschel J, Heikaus S, Depprich R, Kübler NR, Yekta SS, Smeets R, et al. Intraoral schwannoma: review of the literature and presentation of a rare case. CRANIO®. 2012;30(2):150-53.

9 SS, Shashikumar P, H.S S, Kumar GS. Solitary Plexiform Neurofibroma of the Gingiva: Unique Presentation in the Oral Cavity. Journal of Clinical and Diagnostic Research : JCDR. 2013 Sep;7(9):2090-2. PubMed PMID: 24179954.

10 Pawar B, Tejnani A, Sodhi NK, Pendyala G. A silent tumor of the gingiva: An unusual case report and surgical management with 1 year follow-up. Journal of Indian Society of Periodontology. 2013 Jul-Aug 12/01/received 04/30/accepted;17(4):510-3. PubMed PMID: PMC3800417.

11 Angiero F, Ferrante F, Ottonello A, Maltagliati A, Crippa R. Neurofibromas of the Oral Cavity: Clinical Aspects, Treatment, and Outcome. Photomedicine and laser surgery. 2016;34(2): 56-60.

12 do Amaral TL, Valiati R, de Andrade BAB, Piña AR, Torres SR, Romañach MJ, et al. Malignant peripheral nerve sheath tumor of the lower labial mucosa: case report and literature review. Oral surgery, oral medicine, oral pathology and oral radiology. 2015.

13 Arshi A, Tajudeen BA, John MS. Malignant peripheral nerve sheath tumors of the head and neck: Demographics, clinicopathologic features, management, and treatment outcomes. Oral oncology. 2015;51(12):1088-94.

14 Jham BC, Costa NL, Batista AC, Mendonça EF. Traumatic neuroma of the mandible: A case report with spontaneous remission. Journal of Clinical and Experimental Dentistry. 2014 07/0105/01/accepted04/23/received;6(3):e317-e20. PubMed PMID: PMC4134868.

15 Eguchi T, Ishida R, Ara H, Hamada Y, Kanai I. A diffuse traumatic neuroma in the palate: a case report. Journal of medical case reports. 2016;10(1):1.

16 van de Loo S, Thunnissen E, Postmus P, van der Waal I. Granular cell tumor of the oral cavity; a case series including a case. Medicina Oral, Patología Oral y Cirugía Bucal. 2015 Jan;20(1):e30-3. PubMed PMID: 24880452.

17 Costa NCS, Bertini F, Carvalho YR, Almeida JD, Cavalcante ASR. Granular cell tumor presenting as a tongue nodule: two case reports. Journal of medical case reports. 2012;6(1):1.


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