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 Shamim3 that categorized spindle cell neoplasms encountered in oral cavity into tumors of neural, myofibroblastic, vascular, epithelial, odontogenic, and miscellaneous origins.
Spindle cell tumor of neural origin manifesting in oral cavity

Tumors of neural origin, as in presented case, are rare in oral cavity. 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. 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. 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. Treatment of choice for schwannoma is surgical excision.

Neurofibroma

Neurofibroma is also a benign nerve sheath tumor of heterogeneous origin (schwann cells and perineural fibroblasts). 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. Diode laser has recently been reported to be an effective novel therapeutic alternative to surgical excision.

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 Schwann 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