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CASE REPORT

# CASE REPORT- SPINDLE CELL TUMOR: A RARE ENTITY WITH HISTOPATHOLOGICAL DILEMMA

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**ABSTRACT**

Spindle cell neoplasms are defined as neoplasms that consist of spindle-shaped cells in the histopathology. Sarcomatoid carcinomas are biphasic tumors i.e. epithelial cells and spindle cells. These tumors have been proven to be monoclonal dedifferentiated forms of conventional squamous carcinomas. In the oral cavity, the origin of the spindle cell neoplasms may be traced to epithelial, mesenchymal and odontogenic components. Diagnosis of sarcomatoid squamous carcinoma is challenging because of overlapping histopathological features with other spindle-cell tumors. Spindle cell carcinoma of head and neck is a subtype of squamous cell carcinoma and is a unique and rare neoplasm. It has a more aggressive behavior as compared to classical squamous cell carcinoma warranting surgical interventions with wider surgical margins. It is very difficult to diagnose these neoplasms from routine haemotoxyline and eosin sections of histopathology. Immunohistochemistry along with routine histopathology is essential in establishing the diagnosis of spindle cell carcinoma. Surgery and radiotherapy form the mainstays of treatment.



## INTRODUCTION

### *Case report*

A 37 years old male patient visited to outpatient department of Oral Medicine and Radiology with a chief complaint of growth on the palate since last 2 months. Intraoral examination revealed bluish red coloured dome shaped growth on the left side of palate which was gradually increased to the present size of 3.5 cm × 2 cm. It was well circumscribed and sessile, extending from distal aspect of 27- 28 in the interdental gingiva to the soft palate posteriorly (Figure: 1).

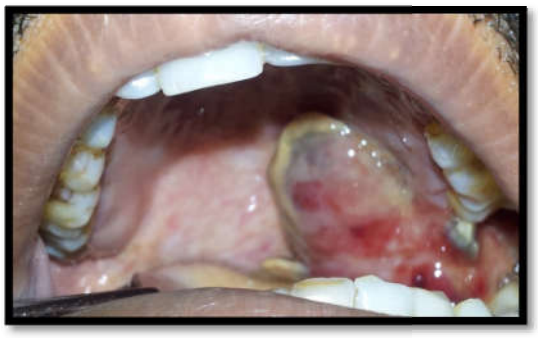

**Figure 1** Intraoral picture showing dome shaped growth on palate

It was associated with dull aching pain, which was continuous in nature. The growth had a smooth intact surface and was firm in consistency. Patient had a habit of chewing tobacco with lime since 15 years, 3-4 times/day. He used to place the quid in the lower labial vestibule. No cervical lymph nodes were palpable. Medical history was unremarkable and hematologic as well as biochemical parameters were within the normal limits. Family history and personal history were not contributory. Based on the above findings the growth was provisionally diagnosed as malignancy of palate and differential diagnosis of squamous cell carcinoma and non-hodgkin's lymphoma was given. The orthopantamograph of the patient revealed no invasion of lesion in the underlying bone. Then incisional biopsy under local anesthesia was performed after taking the patient's consent. Microscopic examination of the incised specimen under 40X (Figure: 2) showed that the tissue had two types of cells. The first type of cells where squamous cells with nuclues in centre suggestive of them being epithelial cells, second type of cell showed spindle arrangement with nucleus in centre suggestive of them being mesenchymal cells. Both cells showed dysplastic features and two types of cells arranged in a biphasic pattern. Under 10X (Figure: 3) showed presence of blood vessels with extravasated Red Blood Corpuscle's, endothelial cells lined by

malignant cells. All the above features were suggestive of spindle cell tumor. Immunohistochemistry using cytokeratin was performed. Immunohistochemistry revealed that spindle cells were strongly positive for cytokeratin (AE1 and AE3). The spindle-cells were focally positive for cytokeratin. (Figure: 4). with this, the final diagnosis of spindle-cell variant of Oral Squamous Cell Carcinoma was made. However, the patient did not return to the center for further treatment.

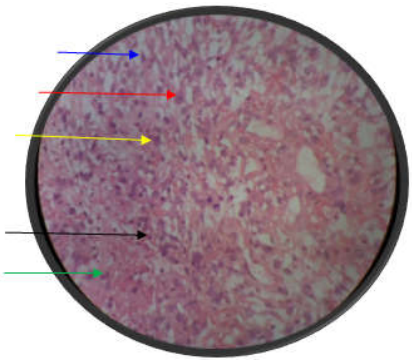

**Figure 2** Photomicrograph under 40X Shows Spindle cells (Blue arrow), Squamous epithelial cells (Red arrow), Hyperchromatic nucluei (Black arrow), Altered nuclear cytoplasmic ratio (Green arrow), Increased mitotic activity (yellow arrow)

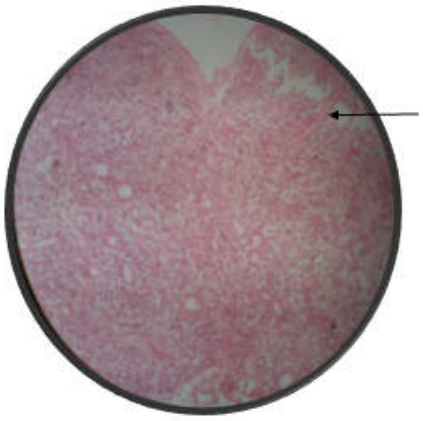

**Figure 3** Photomicrograph under 10X arrow showing presence of Blood vessels with extravasated red blood corpuscle's

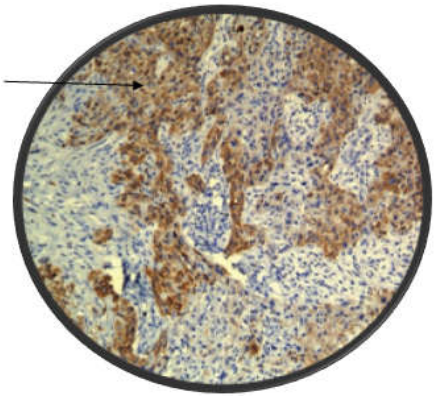

**Figure 4** Immunohistochemistry revealed that spindle cells (Black arrow) were strongly positive for cytokeratin (AE1 and AE3)

## DISCUSSION

Squamous cell carcinoma (SCC) is that the most ordinarily occurring oral malignancy. Spindle Cell carcinoma (SpCC) is uncommon form of poorly differentiated SCC consisting of elongated (spindle) epithelial cells that match a malignant neoplastic disease. This rare variant of SCC has malignant squamous cells and spindle- cells.[1] SpCC is taken into account to be a biphasic tumour composed of a SCC either in place or invasive Spindle Cell carcinoma. There has been confusion over the nature of the sarcomatoid element: whether or not it's benign or malignant, and mesenchymal or epithelial tissue in origin. Thus, to determine the right identification, any clue of the epithelial tissue element thought to be sought after in suspected lesions.[1,3] Mainly 3 theories are projected to clarify the histogenesis of spindle-cells. Initial theory states that the spindle-cells and epithelial cells arise at the same time from separate stem cells. Thus, the tumour worth the name of a "collision" tumour. Second theory explains the character of the spindle-cell element as Associate in atypical reactive proliferation of the stroma and thus the tumour being known as "pseudo sarcoma." consistent with the last theory, each spindle and epithelial tissue elements have constant being origin, and "dedifferentiation" or "transformation" of epithelial cells to spindle-cells has occurred.[1,3]

The third theory is greatly supported lately by the evidences that: (1) they occur within the sites that unremarkably have squamous epithelial tissue and a Preponderance of SCC instead of sarcomas. (2)A polypoid or ulcerated lesions almost like SCC. (3) The direct continuity and transition of the spindle-cells with areas of squamous epithelial tissue. (4) Immunoreactivity with epithelial tissue antigens. (5) A twin expression of epithelial tissue and mesenchymal differentiation with double labeling techniques in some growth spindle-cells. The spindle-shape of the tumour cells has been thought of to be caused by the lack of expression of cell adhesion molecule comparable to cadherins and therefore the consequent alteration of keratin filament network. It's been stated that development of the spindle-cell constitution involves purposeful loss of genes that managed epithelial tissue differentiation which leads conversion to spindle morphology may be a recessive entry. [3,5]

The precise reason behind Spindle Cell carcinoma isn't better-known, it's powerfully related to a history of tobacco intake, smoking and alcohol abuse as found in our case. SpCC is additional predominant in men compared to females (1.2:1 ratio).[6] The subsite distribution within the mouth showed that the foremost common site concerned was buccal mucosa and gingivobuccal fissure, followed by upper or lower alveolus, tongue, hard palate and lip.(4) The tumors sometimes mature rapidly. As ascent was seen during this patient, the lesion expressed classical polypoid and ulcerated appearence. The identification of Spindle Cell carcinoma needs microscopic anatomy demonstration of each the epithelial cell and the spindle-shape cells with sarcomatous appearances. Histopathology shows the presence of SCC at the surface or deeper at intervals the tumour. Though this is often rare, particularly within the oral cavity tumors, Lesion shows a mixing of squamous cells and spindle-cells which might be completely differentiated by their different arrangement which has storiform, solid, and fascicular look in regarding 1/2 cases there's additionally a desmoplastic stromal pathology, and since the epithelial tissue cells are capable of reverting into sarcomatoid spindle-cells.[4,6]

In addition to microscopic studies, immunohistochemical studies of epithelial and mesenchymal markers are helpful to diagnose a tumour. Epithelial markers consists of keratin (AE1/AE3), epithelial tissue membrane antigens. Mesenchymal markers are vimentin, desmin, S-100,

Osteopontin, and bone morphogenetic protein [7,8]. These cases are tough to differentiate histologically from different spindle-cell lesions, particularly atypical fibroxanthoma (AFX). The growth cells0 of AFX are extremely atypical and are variably cigar-shaped with few eccentric multinucleated cells. The predominant cells in AFX are plump, fusiform, and occur in poorly organized fascicles. The cells have a distinguished nucleus that is usually vesicular. Mitotic activity is typically brisk. Usually AFX cells obscure at the fringe to mix with the encircling dermal fibroblasts; this feature will facilitate identification.[9,10] Chaudhary *et al* reported a case in which an ulcerative lesion on lower front region of jaw since last 3 months. In this the microscopic examination of the incised specimen showed the tumor consisted of malignant epithelial and mesenchymal components. The bulk of the tumor was mainly basophilic, hyperchromatic, pleomorphic spindle-cells accompanying small areas of neoplastic epithelial cells arranged in sheets. Immunohistochemistry with cytokeratin revealed polygonal lesional cells which are strongly positive for cytokeratin. Moreover the spindle cells were strongly positive for vimentin. [1]

The differential diagnosis includes both benign and malignant neoplasm like SCC, malignant fibrous histiocytoma, fibrosarcoma, leiomyosarcoma, rhabdomyosarcoma, malignant peripheral nerve sheath tumor, osteosarcoma, malignant melanoma, kaposi's sarcoma, angiosarcoma, synovial sarcoma, leiomyoma, fibromatosis, reactive epithelial proliferations.[10] SpCC in oropharynx and oral cavity is seems to be more aggresive, which recurs frequently and metastasized. Treatment modality includes surgical excision as same as for SCC. The role of chemotherapy has not been reported, but it may be needed to reduced rate of recurrence or metastasis of primary sarcomatous tissue. After radiotherapy the poor prognosis has been reported in patients who have nodal metastasis during time of diagnosis. [8,9]

## CONCLUSION

SpCC have been proven to be monoclonal dedifferentiated forms of conventional squamous carcinomas. In this case site of occurence of SpCC was rare. In the oral cavity, the origin of the spindle cell neoplasms may be traced to epithelial, mesenchymal and odontogenic components. Diagnosis of spindle cell tumor is challenging because of overlapping histopathological features with other spindle-cell tumors. Spindle cell carcinoma of head and neck is a subtype of squamous cell carcinoma and is a unique and rare neoplasm. SpCC shows spindle cells in transition from SCC which is considered as sarcomatous transformation of SCC. It has a more aggressive behavior as compared to classical squamous cell which is seen due to transformation. It is very difficult to diagnose these neoplasms from routine haemotoxyline and eosin sections (H and E) on histopathology. Immunohistochemistry along with routine histopathology is essential in establishing the diagnosis of spindle cell carcinoma. Immunohistochemistry revealed that spindle cells were strongly positive for cytokeratin (AE1 and AE3). The spindle-cells were focally positive for cytokeratin. Surgery and radiotherapy form the mainstays of treatment. After radiotherapy the poor prognosis has been reported in patients who have nodal metastasis during time of diagnosis.

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