Case Report

International Journal of Dental and Health Sciences
Volume 05,Issue 04

SPINDLE CELL CARCINOMA OF BUCCAL MUCOSA: A CASE REPORT OF RARE ENTITY

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ABSTRACT:

Spindle cell carcinoma a rare malignancy of the head & neck regions is an unusual form of poorly differentiated squamous cell carcinoma (PDSCC). It accounts for less than 1% of all tumors of oral regions. Histologically the lesion demonstrates elongated (spindle) epithelial cells that resemble a sarcoma thus posing a challenge towards its diagnosis. Larynx is the most common site of occurrence, but cases have also been reported in nasal cavity, hypopharynx, oral cavity, esophagus, trachea, skin and breast. We report a rare case of spindle cell carcinoma of buccal mucosa, in a 45-year old female patient.

Keywords: Spindle cell carcinoma, Squamous cell carcinoma, Buccal mucosa

INTRODUCTION:

Spindle cell carcinoma (SPCC) is also known by different nomenclatures such as carcinosarcoma, pseudosarcoma, sarcomatoid carcinoma, collision tumor and pseudosarcomatous carcinoma. The different nomenclature of SPCC itself suggests its biphasic nature. SPCC occurs commonly in 6-7th decades of life and represents a male predominance. Smoking, alcohol consumption and previous irradiation of the head & neck region are the predisposing factors.[1] Here we report a case of SPCC in a rare location like buccal mucosa in a 45-year old female patient with habit of Paan chewing.

CASE DETAILS:

A 45 year old female patient reported to the department of Oral Medicine & Radiology of our institution with a chief complaint of pain & swelling on the left side of the mouth with sudden increase in size. History revealed onset of the lesion since twenty days.

No relevant history of any systemic illness and drug allergy was reported. Patient presented a habit history of Paan chewing 6-7 times daily for the past 30 years. On intraoral examination the lesion measured 4 x 4 cm in size (Figure1). It was polypoid, oval in shape, erythematous, tender and fixed to the underlying structures. On extra oral examination left submandibular lymph node was tender, palpable and not fixed. On her first visit patient was advised to quit the habit and an incisional biopsy was done.

Histopathological examination showed connective tissue consisting of sheets of tumour cells arranged in variable patterns like storiform, fasicular and streaming.

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(Figure 2) The tumour cells were predominantly spindle shaped and few squamoid cells. (Figure 3) The spindle cells were monomorphic with blunt pale staining nuclei and scanty cytoplasm. The squamous cells were round to polygonal in shape with vesicular nuclei with prominent multiple nucleoli, altered nuclear cytoplasmic ratio, cellular and nuclear pleomorphism and abnormal mitosis. Connective tissue was minimal consisting of mature bundles of collagen fibers with focal infiltration of chronic inflammatory cells. Areas of necrosis were also noticed.

Immunohistochemistry with Pan Cytokeratin marker showed diffuse strong positivity. (Figure 4) Based on histopathology and aided by Immunohistochemistry, a diagnosis of SPCC was given.

DISCUSSION:

WHO (2015 classification) defined SPCC as a biphasic tumor composed of squamous cell carcinoma, either in-situ and/or invasive, and a malignant spindle cell component with a mesenchymal appearance, but of epithelial origin. In a study by Gupta R et al SPCC is characterized as epithelial in origin and may arise from conventional SCC by sarcomatous transformation. This transformation explains the increased aggressiveness seen in SPCC. [2] Short duration of onset and rapid increase in size of the lesion in the present case also supports the aggressive nature of SPCC.

Battifora in his ultra structural studies demonstrated presence of junctional complexes between tumor spindle cells, with or without pericellular basal lamina and cytoplasmic skeins of intermediate filaments thus postulating epithelial-mesenchymal transformation.[3] It has been documented that the epithelial cells go through a spectrum of progressive phenotypic changes, acquiring a mesenchymal pathway of differentiation metamorphosing to a spindle shape, undergoing a loss of cellular polarity, producing mesenchymal matrix components, and gaining vimentin while losing keratin expression. It has been demonstrated that the phenotypic plasticity of interconversion of epithelium to mesenchyme cells(as seen during embryogenesis)is expressed by a loss of intercellular cohesion, elongation of the cells, loss of basement membrane , production of connective tissue(collagen), and invasion into the stroma.[4]

In a study by Viswanathan et al (2010) the predominant site affected is oral cavity in contrast to larynx of western studies, where smoking of tobacco is reported to be the predominant habit. According to his study the subsite distribution in the oral cavity showed that the most common site involved was buccal mucosa and gingivobuccal sulcus (38.5%) followed by upper or lower alveolus(30.8%), tongue(20%), hardpalate (7.8%) and lip (3.1%).[5] The usual clinical presentation of oral SPCC is

a polypoidal mass with clinical presentation of dysphagia, or bleeding.[6]

Microscopically SPCC shows two distinct epithelial-derived components: A carcinomatous or OSCC component and a sarcomatoid or dysplastic spindle cell component.[7] A minor portion of the tumor mass is carcinomatous while the greatest portion is spindle cell component, presenting in storiform and fasciculated pattern, as also revealed in our case. However confirmatory diagnosis was done after using pan CK immunohistochemical marker. IHC improves diagnosis by including a larger panel of antibodies. However in daily practice pan CK (including intermediate and LMWCK) and EMA are most useful and were positive in about 61.3% of cases of sarcamatoid carcinoma.[5]

Differential diagnosis includes fibromatosis, reactive epithelial proliferations, inflammatory myofibroblastic sarcoma, low grade myofibroblastic sarcoma, myoepithelial carcinoma, and fibrosarcoma.[8]

Treatment of choice is surgical excision and controlling local and distant metastasis. The most common sites of distant metastasis being lymph node and lungs. Prognostic features like low stage, polypoid rather than endophytic growth, relatively shallow depth of sarcamatoid invasion, absence of prior radiation favour the patient. The reported 5- year survival is between 65 and 95%.[8]

CONCLUSION:

To conclude SPCC are rare biphasic tumors with few cases being reported in literature on buccal mucosa. It is difficult to discern the lesion even histopathologically because of its overlapping features with other spindle cell tumors. Thus SPCC should always be considered if clinicians encounter with any polypoid and aggressive lesion aiding in better patient management and prognosis.

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FIGURES:

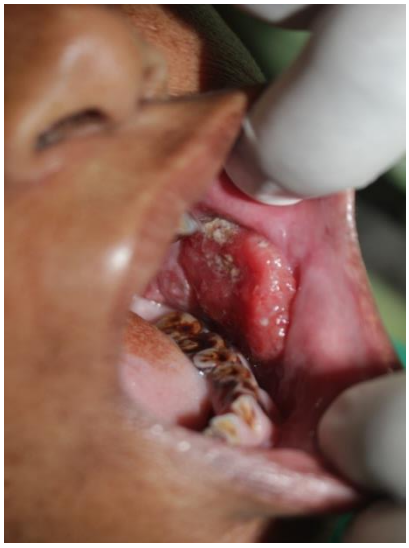

Fig 1:Photomicrograph showing lesion on the left side of buccal mucosa

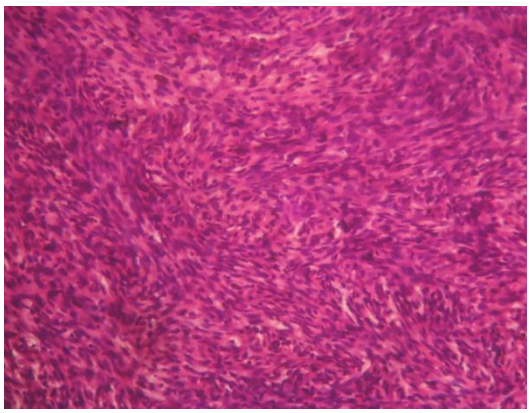

Fig 2:Photomicrograph demonstrating sheets of tumour cells forming variable pattern like storiform and fasicular. (H &E Stain, X200)

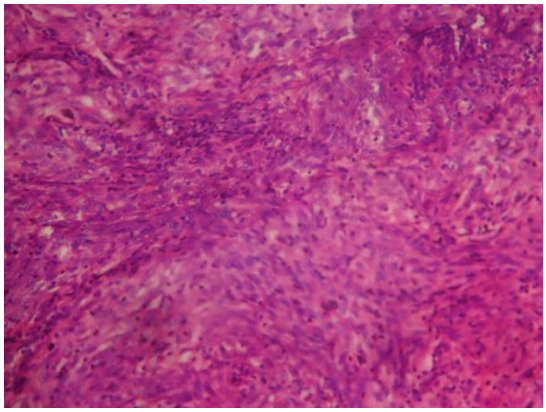

Fig 3:Photomicrograph demonstrating tumour cells, mainly spindle shaped and few squamoid cells (H&E Stain, X200)

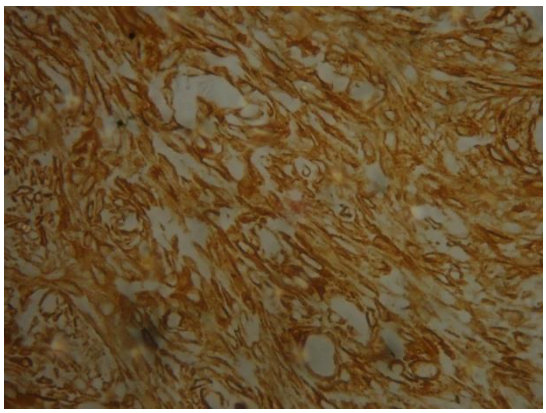

Fig 4:Photomicrograph demonstrating diffuse strong positivity immunostaining with Pan Cytokeratin marker.(X200)