

Spindle Cell Carcinoma of the Tongue: Fallacies in Its Diagnosis



Amrit Kaur Kaler, K Shweta, Smitha Chandra B.C, Naik S

Associate Professor, Pathology; Senior Resident, Pathology; Assistant Professor, ENT; Professor and HOD, ENT

Abstract

Spindle cell carcinoma is a rare aggressive biphasic tumor, composed of neoplastic proliferation of both epithelial (squamous) and spindle cell population. It constitutes about 1% of all oral cavity tumors 2a and is almost rare on the tongue; only few cases have been reported so far. This variant of squamous cell carcinoma, comprises major diagnostic problems due to its varied histomorphology and resemblance to sarcomatous lesion; hence diligent screening and IHC markers are mandatory for its diagnosis.

Keywords: *Spindle cell carcinoma, Biphasic tumor, tongue.*

Introduction

Spindle cell carcinoma (SpCC) was first described by Virchow in 1864; and accounts for 3% of all squamous cell carcinomas in the head and neck region.^[1] It is most commonly seen in larynx, hypopharynx, oesophagus, nasal cavity and trachea.^[2] It is a rare tumour in the oral cavity.^[3] and very rarely seen on the tongue. Spindle cell carcinoma is a variant of Squamous cell carcinoma but various terminologies has been proposed in the past to describe the entity in the past as carcinosarcoma, pseudocarcinoma, pseudosarcoma, pleomorphic sarcoma, malignant mixed mullerian tumor.^[4] These tumors have now been proved to be monoclonal, evolving from conventional squamous cell carcinoma with dedifferentiation associated with sarcomatoid transformation.^[5] Now the WHO has placed this entity under classification of tumors of the oral cavity and oropharynx as malignant epithelial tumors of SCC and labeled it "Spindle cell carcinoma" as proposed by Sherwin et al.^[6] The primary squamous component may or may not be visible in the majority of tumors as they may show ulceration and fibrinoid necrosis.

We present a case of Spindle cell carcinoma of the tongue in a middle aged male, which posed significant diagnostic challenge with remarkable morphological and immunohistochemical overlap with other benign and malignant spindle cell tumors; a rarity in its presentation.

Case report

A 45 year old male came to ENT OPD for the evaluation of the mass on the tongue since 3 months, which was initially small and ulcerated but increased to the present size. He gave a history of pain and odynophagia since 15 days. He gave the history of chewing pan and betel leaves since 5

years and also smoking 3 packs of cigarettes every day since 10 years. Examination revealed 3cm × 1cm tender, whitish pedunculated, mass with the stalk measuring 0.5 cms on the right lateral border of the tongue. The mass didn't bleed on touch and the surrounding area showed no induration. There were no palpable lymph nodes in the neck. A clinical diagnosis of Papilloma with leukoplakia was made. Wide local excision of the lesion with good margin was done under general anaesthesia. The gross specimen showed a portion of the tongue measuring 2.5x1.8x0.2 cms and the ulcerated tumor measured 1.5x0.8x0.7 cms (Fig 1). Cut surface of the tumor is grey white with areas of hemorrhage and necrosis.

Microscopic examination showed a polypoidal tumor covered by dysplastic squamous cells showing areas of ulceration with underlying nuclear debris, fibrinous exudates and sheets of acute inflammatory cells. The stroma shows infiltrating squamous cell carcinoma intermixed with atypical spindle cell proliferation arranged in the form of fascicles and whorls interspersed with granulation tissue composed of proliferating capillaries, acute inflammatory cells and fibrin (Fig 2 & 4). The spindle cells show bizarre morphology with marked nuclear pleomorphism forming binucleate to multinucleate giant cells with mitosis <5/10HPF (Fig 3). All the margins are free of tumor cells. Attached skeletal muscle shows no infiltration of tumour cells. The diagnosis of spindle cell carcinoma, a variant of squamous cell carcinoma was made on histopathology. Immunohistochemistry (IHC) was performed and the cells showed cytoplasmic positive expression for cytokeratin among the squamoid cells (Fig 5). Almost all the spindle cells stained deeply with vimentin (Fig 6). Following the report Supraomohyoid neck dissection was done. None of the nodes showed any metastasis. Since then patient has been under regular follow up with no recurrences.

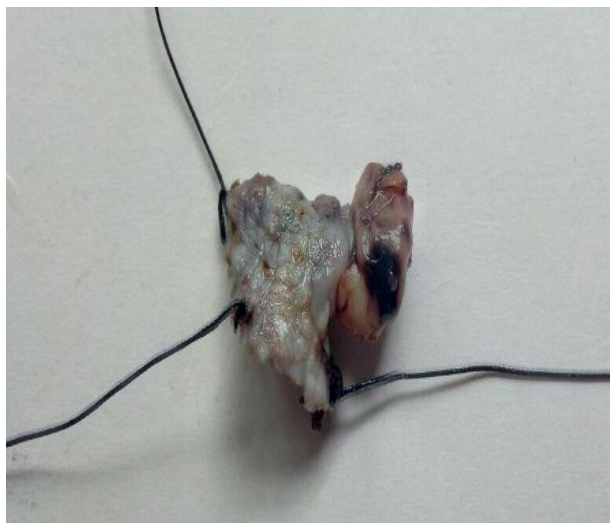


Fig 1: Excision biopsy showed a pedunculated mass attached to tongue.

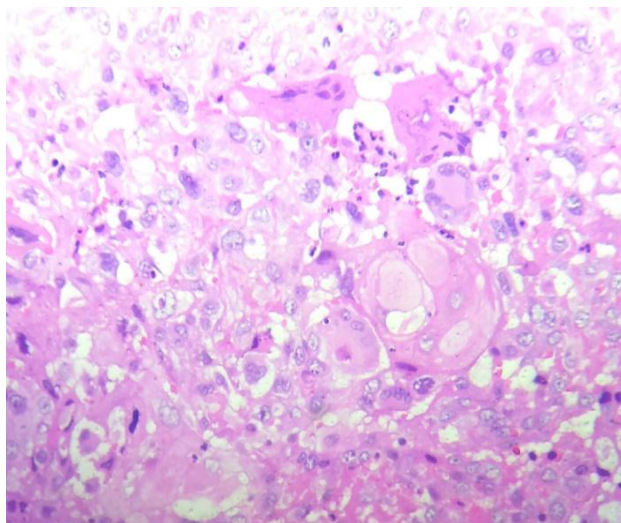


Fig 2: Infiltrating Squamous cell carcinoma in nests and lobules with giant cell reaction.

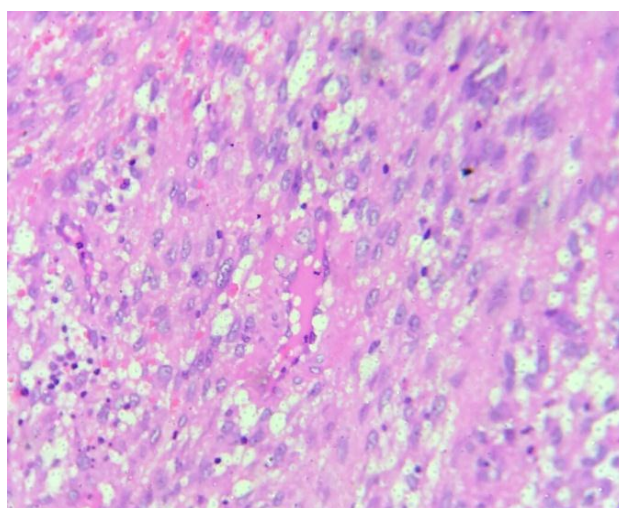


Fig 3: Malignant spindle cells arranged in fascicles and whorling pattern.

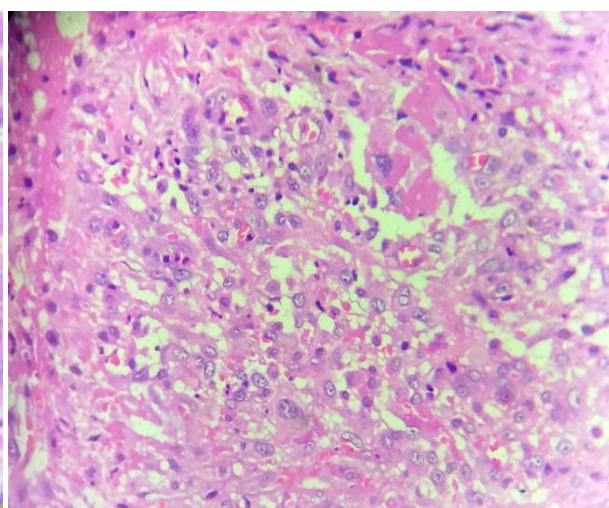


Fig 4: Transition zone showing a transition from malignant squamous to spindle cells.

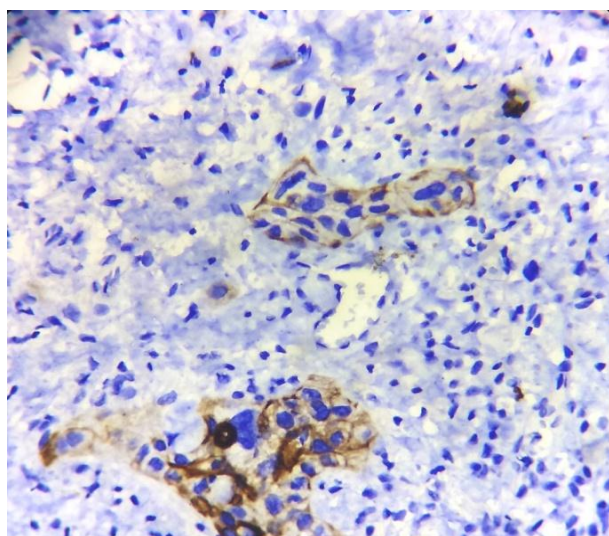


Fig 5: IHC shows cytokeratin positivity among squamous cells

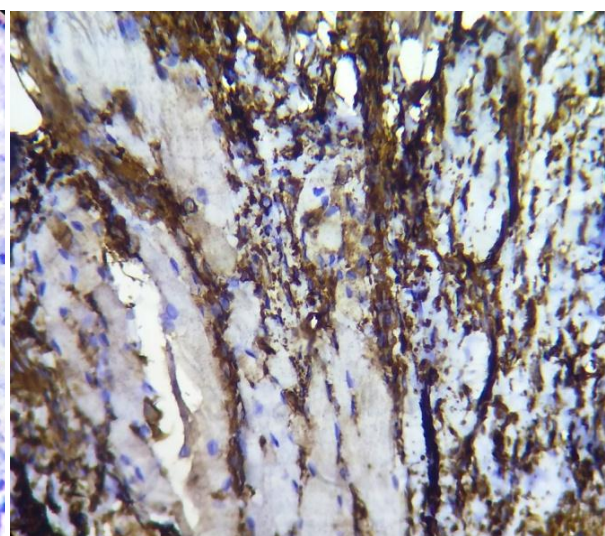


Fig 6: IHC shows vimentin positivity among spindle cells.

Discussion

Spindle cell carcinoma (SpCC) is a rare unique variant of squamous cell carcinoma composed of both malignant epithelial (squamous) and mesenchymal (spindle) cell population. This tumor has been proved to be of monoclonal origin and the spindle cells are merely the dedifferentiated component of squamous carcinoma.^[5] There is also strong evidence that the transformation of the epithelial cells into spindle cells is envisaged to be a part of Epithelial-Mesenchymal Transition (EMT) showing phenotypic plasticity at the microscopic and IHC level.^[4] The spindle shape of the tumor cells consequently shows lack of expression of cell adhesion molecules and alteration of keratin filament network. This theory has been widely accepted and has been strongly supported by some studies.^[7,8]

On histopathological examination, the malignant squamous cells may show varied differentiation from (group I) frank epithelial differentiation in the form of dysplasia, squamous nests, lymph node metastasis to (Group II) with epithelial differentiation at immunohistochemical level and to (Group III) with no epithelial differentiation.^[1,3] The transition of the characteristic polygonal shaped, cobblestone epithelial cells to their mesenchymal elongated and invasive characteristics undergoing a characteristic EMT switch can be identified.^[9] The spindle cells may be arranged in various sarcomatoid patterns and may show component like cartilage formation, bone like calcification, stromal metaplasia; makes it mimic a high grade mesenchymal malignancy.^[3,4,10] The sarcomatoid component is divided into 3 grades i.e. Mild, moderate and severe anaplasia (Grade 1, 2 & 3 respectively).^[1,3]

The present case had malignant squamous component was seen intricately mixed with spindle component in the stroma and the overlying dysplastic epithelium showing ulceration and fibrinoid necrosis while malignant spindle cells were arranged in storiform and fascicular pattern with variable amount of collagen, bizarre pleomorphic cells and giant cells along with high mitosis with abnormal forms. The tumour fell in group I as epithelial differentiation was seen and confirmed at light microscopic level and grade III as the sarcomatous element showed severe dysplasia, as pleomorphism and mitosis was noted in these cells.

Prakash N et al stated that in 60-70% cases, the epithelial differentiation is seen at the stalk of the polypoidal mass at the deepest part of the advancing front.^[7] In the present case also, the malignant squamous cells were seen in the base of the polypoidal mass with the differentiation towards the spindle cells to the periphery. Immunohistochemistry showed cytokeratin positivity among the malignant squamous epithelial cells and dysplastic surface epithelium while vimentin positivity among spindled cells. The change

of immunohistochemistry stain from cytokeratin to vimentin showed complete cytoskeletal protein alteration; proved the dedifferentiation or transformation to spindle cells.

Spindle cell carcinoma is common in 5-7th decade of life with a profound male domination (M: F- 11:1).^[11] It constitutes about 1% of the tumors in the oral cavity tumors^[3] and is almost rarely present on the tongue. The duration of the lesion is usually less than 1 year in 95% of patients^[2] and presents as exophytic, firm and non-tender mass.^[8] The present case had a rare presentation on the lateral side of tongue in a 45-year-old male as a polypoidal, tender, soft tissue mass with surface ulceration which grew rapidly to the present size in a period of 3 months. The patient was a chronic smoker with history of eating betel leaves and pan since 5 years; the possible reason to present this growth early in life. The potential risk factors described in the literature include like tobacco use, alcohol abuse, previous irradiation exposure, poor oral hygiene.^[2,8]

The spindle cell carcinoma has metastatic rate of 26-75% to the lymph nodes; regional metastasis is more common than distant.^[11] SpCC is more aggressive than the more usual squamous cell carcinoma, and hence has lower overall survival.^[5] The present case was diagnosed in early stage within 3 months and hence did not present with nodal metastasis; hence carried a good prognosis.

The SpCC bears a close differential diagnosis with both benign and malignant spindle cell tumors at microscopic level due to the arrangement of sarcomatoid cells in storiform pattern like leiomyosarcoma, malignant fibrohistiocytoma, fibrosarcoma and melanoma.^[4] A detailed clinical data with diligent analysis of different patterns on histopathology with IHC correlation may be useful in diagnosing the entity. Tumor cells admixed with extensive inflammation and granulation tissue might simulate inflammatory myofibroblastic tumors. Fibrosarcomas are rare in head and neck region, synovial sarcoma is deep soft tissue in young adults with SYT-SSX1 translocation, leiomyosarcomas show cigar shaped nuclei with cytoplasmic vacuolations, rhabdomyosarcomas show tadpole like cells, melanomas may not be pigmented with fusiform cells and IHC marker HMB-45 may be required for diagnosis.^[2,3,8]

Conclusion

The diagnosis of spindle cell carcinoma is challenging among the mucosal sites with much diverse presentation as with areas mimicking benign and malignant spindle cell tumors. These features emphasize that the tumour can provide fallacies in its diagnosis. Hence, a systemic approach towards understanding the clinical, microscopic and immunohistochemistry features are critical in

diagnosing the disease and hence better patient management.

References

- [1] Radhika M Bavle, Girish Govinda, Padmalatha G.V. et al. Spindle Cell Carcinoma of Mandible. *Journal of Clinical and Diagnostic Research*. 2016; 10(7): 05-86.
- [2] Ravindran R, Mohan V M. Saji A. Spindle cell carcinoma of maxilla: case report of a Rare entity and review of literature. *OMPJ*; 2013; 4:0976-1225.
- [3] 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 Neck Pathol*. 2010; 4(4):265-75.
- [4] Thompson LD, Wieneke JA, Miettinen M, Heffner DK. Spindle Cell (sarcomatoid) carcinomas of the larynx: a clinicopathologic study of 187 cases. *Am J Surg Pathol*. 2002; 26:153–70.
- [5] Lewis JE, Olsen KD, Sebo TJ. Spindle cell carcinoma of the larynx: review of 26 cases including DNA content and immunohistochemistry. *Hum Pathol*. 1997; 28:664–73.
- [6] Sherwin RP, Strong MS, Vaughn CW Jr. Polypoid and junctional squamous cell carcinoma of the tongue and larynx with spindle cell carcinoma ('pseudosarcoma'). *Cancer*. 1963; 16:51–60.
- [7] Prakash N, Kumar MS, Sharada P, Pradeep GL. Spindle cell carcinoma of the oral cavity: A case report of a rare entity and review of literature. *World Journal of Dentistry*. 2010; (1):55-58.
- [8] Oktay M, Kokenek-Unal TD, Ocal B, Saylam G, Korkmaz MH, Alper M. Spindle cell carcinoma of the tongue: a rare tumour in an unusual location. *Pathol Res Int*. 2011:1-6.
- [9] Krisanaprakornkit S, Iamaroon A. Epithelial-mesenchymal transition in oral squamous cell carcinoma. *ISRN Oncol*. 2012; 681469(10).
- [10] Katase N, Tamamura R, Gunduz M, Murakami J, Asaumi J, Tsukamoto G, et al. A spindle cell carcinoma presenting with osseous metaplasia in the gingiva: a case report with immunohistochemical analysis. *Head Face Med*. 2008;4:28.
- [11] Ramamurti A, Venkataraman M, Narasimhan M, Rao SR. Spindle cell carcinoma of the gingiva: A rare occurrence. *Contemp Clin Dent*. 2013; 4(4):500-03.
- [12] Parikh N, Desai N. Spindle cell carcinoma of the oral cavity: A case report of a rare entity and review of literature. *J academy Adv Dental Research*. 2011; 2:3136.
- [13] Prakash N, Kumar MS, Sharada P, Pradeep GL. Spindle cell carcinoma of the oral cavity: A case report of a rare entity and review of literature. *World Journal of Dentistry*. 2010; 1:55-58.