

Spindle Cell Carcinoma of the Buccal Mucosa: A Rare Tumor in a Rare Site

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Abstract: Spindle cell carcinomas are biphasic tumors of monoclonal origin with mesenchymal component proved to be the dedifferentiated forms of conventional squamous cell carcinomas. This unusual variant of squamous cell carcinoma usually presents in the larynx, but are also infrequently reported in the gingiva, tongue, lips, hypopharynx and nasal cavity. We report a spindle cell carcinoma of buccal mucosa in a 56-year-old male patient.

Keywords: spindle cell carcinoma, oral squamous cell carcinoma, Lane tumor, oral cavity

I. Introduction

Spindle cell carcinoma (SPCC), a poorly differentiated variant of squamous cell carcinoma (SCC), is a rare malignancy accounting for less than 1% of all tumors of oral regions. The diagnosis of this variant is important because the tumor exhibits an aggressive course and high incidence of metastasis than conventional SCC^[1]. This biphasic tumor, with both malignant epithelial and spindle cell components, can sometimes be challenging since they can resemble spindle cell lesions ranging from benign reactive to malignant ones like fibrosarcoma. Since larynx is the most common site for the tumor, literature regarding SPCC arising in the upper aerodigestive tract is limited, with very few cases pertaining to its localisation in the buccal mucosa. We report a case of spindle cell carcinoma of buccal mucosa, in a 56-year-old male patient.

II. Case history

A 56-year-old male patient presented to the department with a complaint of pain and a non healing wound on the right cheek since 2 months. He had consulted a dentist elsewhere for the same and did selective grinding of the lower posterior teeth and was prescribed some vitamin tablets. No reduction in symptoms was noticed. Medical history was unremarkable except for the presence of diabetes mellitus, for which he is under oral medication. He reported habits of tobacco chewing (betel leaf, arecanut, slaked lime and tobacco), pan chewing (commercial preparation) and occasional alcohol consumption for more than 12 years. On examination, an ulceroproliferative lesion on the right posterior buccal mucosa, extending 1 cm from the angle of mouth to the maxillary tuberosity region anteroposteriorly and to the alveolus and gingiva of tooth 13 to 17 superiorly, was noticed. The surface of the lesion was granular and was covered by a whitish necrotic slough in some areas (Fig 1). On palpation, it was mildly tender. Induration was positive. There was a single palpable right submandibular lymph node which was mobile and mildly tender. Incision biopsy was performed and histopathological examination revealed an ulcerated stratified squamous epithelium infiltrating into underlying moderately collagenous connective tissue stroma in a streaming pattern (Fig 2). The tumor cells exhibited a biphasic pattern and were seen as strands, nests and individually. The cells in the upper portion were round to ovoid with high nuclear cytoplasmic ratio, hyperchromatism and pleomorphism. Numerous mitotic figures were noted (Fig 3). The advancing tumor front consisted of proliferating spindle cell population infiltrating into skeletal muscle fibres (Fig 4). Moderate diffuse collection of inflammatory cells, chiefly macrophages were seen surrounding the tumor cells. Immunohistochemical analysis revealed the tumor cells to be positive for both cytokeratin (AE1/AE3) and vimentin (Fig 5 and 6). With this, a diagnosis of spindle cell carcinoma was given. The patient was referred to Regional Cancer Centre, Trivandrum for further management and was lost for follow up.

III. Discussion

Spindle cell carcinoma, as defined by WHO, is a biphasic tumor composed of a squamous cell carcinoma, either in-situ and/or invasive, and a malignant spindle cell component with a mesenchymal appearance, but of epithelial origin^[2]. It account for 3% of all squamous carcinomas in the head and neck region^[3].

The histogenesis of SPCC remains controversial. The differences in opinion have paved way for authors to coin a plethora of terms like carcinosarcoma, pseudosarcoma, sarcomatoid carcinoma, collision tumor, Lane tumor and pseudosarcomatous carcinoma. The currently accepted terminology 'spindle cell carcinoma' was coined by Shervin et al. [4]. The concept that the sarcomatous part is of epithelial origin and results from metaplasia of squamous cells was put forth by Krompecher as early as 1900 [5]. Several immunohistochemical, electron microscopic, and genetic studies have now proved that the epithelial and spindle components are of monoclonal origin. Despite their evident divergence at the phenotypic level, they share a common pathway of tumorigenesis [6]. The epithelial cells undergo a loss of cellular polarity and keratin expression while acquiring a mesenchymal pathway of differentiation by producing mesenchymal matrix components and gaining vimentin, thus metamorphosing to a spindle shape [3].

The tumor occurs predominantly in males of 6th and 7th decades. Cigarette smoking, alcohol consumption and radiation exposure are considered as the main contributory factors. 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. In our case, though the sex and age was typical for SPSC, the site of the tumor was buccal mucosa which is very rare. This may be attributed to his habit of tobacco chewing and alcohol consumption. It can be assumed that oral cavity becomes the predominant site affected, in areas where tobacco chewing is prevalent in contrast to Western countries where tobacco smoking is reported to be the predominant habit and larynx the predominant site. A study by Vishwanathan S et al [3] support this finding.

The usual clinical presentation of oral and oropharyngeal SPSC is a polypoidal mass with nonhealing ulcer, dysphagia, or bleeding [1,3]. Histologically, the bulk of the tumor is usually formed by the spindle cell component in various patterns and evidence for squamous epithelial derivation is seen as either in-situ carcinoma or as invasive SCC. Mitotic figures are usually prominent. Patients with a history of radiotherapy may show foci of osteosarcomatous, chondrosarcomatous, or rhabdosarcomatous differentiation. Regional lymph node metastasis is positive in up to 25% of cases and distant metastasis is uncommon (5-15%). Our patient did not show any lymph node metastasis. The metastatic foci usually contain SCC alone or both SCC and spindle cell component, and rarely, only the spindle cell component. Tumor cells can express both epithelial and mesenchymal markers. The most useful epithelial markers are AE1/AE3, CK1, CK18 and epithelial membrane antigen (EMA). Spindle cells express vimentin and cytokeratin (40-85% cases) and less frequently, smooth muscle actin, muscle specific actin, and desmin. We did immunohistochemistry with AE1/AE3 and vimentin, both of which showed positivity for tumor cells.

The diagnosis of SPCC generally requires the demonstration of both malignant spindle cells and squamous cell carcinoma, either in-situ or invasive. When a SCC component is inconspicuous, then the spindle cells should be investigated for evidence of epithelial differentiation. Differential diagnosis include fibromatosis, nodular fasciitis, reactive epithelial proliferations, inflammatory myofibroblastic sarcoma, and low-grade myofibroblastic sarcoma, myoepithelial carcinoma, mucosal spindle cell melanoma, leiomyosarcoma, fibrosarcoma, malignant fibrous histiocytoma, rhabdomyosarcoma, malignant peripheral nerve sheath tumor, mesenchymal chondrosarcoma and malignant melanoma [2,3,7].

In the oral region, surgery is accepted as the best choice of treatment [1]. Since tumors of the oral cavity and oropharynx are potentially aggressive and seem to recur and metastasize easily, management should aim at controlling local and distant recurrence. Favorable prognostic features are: lowstage, polypoid rather than endophytic growth, relatively shallow depth of sarcomatoid invasion, absence of prior radiation and minimal immunoreactivity for polyclonal cytokeratin. The reported 5-year survival is between 65 and 95% [2]. A large number of cases show distant metastasis, the most common sites being lymph node and lungs.

IV. Conclusion

Spindle cell carcinoma of oral cavity is a rare biphasic tumor. An accurate diagnosis of these tumors is essential as they differ in their clinical management and outcome. Since true sarcomas of the head and neck are extremely rare, spindle cell carcinoma should always be considered during evaluation of the polypoid lesions of this region especially when the epithelial component is sparse or absent. Its undisputed poor prognosis and high rate of recurrence and distant metastasis when compared to conventional SCC should always be kept in mind.

References

- [1]. Oktay M, Kokenek-Unal T.D, Ocal B, Saylam G. Spindle Cell Carcinoma of the Tongue: A Rare Tumor in an Unusual Location. Pathology Research International. 2011.
- [2]. Fletcher C.D.M., Unni K.K., Mertens F. (Eds.): World Health Organization Classification of tumours. Pathology and genetics of tumours of soft tissue and bone. IARC Press: Lyon 2002
- [3]. Viswanathan S, Rahman K, Pallavi S, Sachin J 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 Pathol. 2010; 4:265–275.
- [4]. Prakash N, Harishkumar M.S, Sharada P and Pradeep G.L. Spindle cell carcinoma of the oral cavity: a case report of a rare entity and review of literature. World J Dentistry. 2010; 1(1): 55-58.

- [5]. Agha F.P and Keren D.F. Spindle cell squamous carcinoma of the esophagus: a tumor with biphasic morphology. *AJR*. 1985; 145: 541-45.
- [6]. Su H.H, Chu S.T, Hou Y.Y, Chang K.P and Chen C.J. Spindle Cell Carcinoma of the Oral Cavity and Oropharynx: Factors Affecting Outcome. *J Chin Med Assoc*. 2006; 69(10).
- [7]. Sarma A, Das R, Sharma J.D and Kataki A.C. Spindle cell carcinoma of the head and neck: a clinicopathological and immunohistochemical study of 40 cases. *J Can Therapy*. 2012; 3: 1055-59.

Figures:

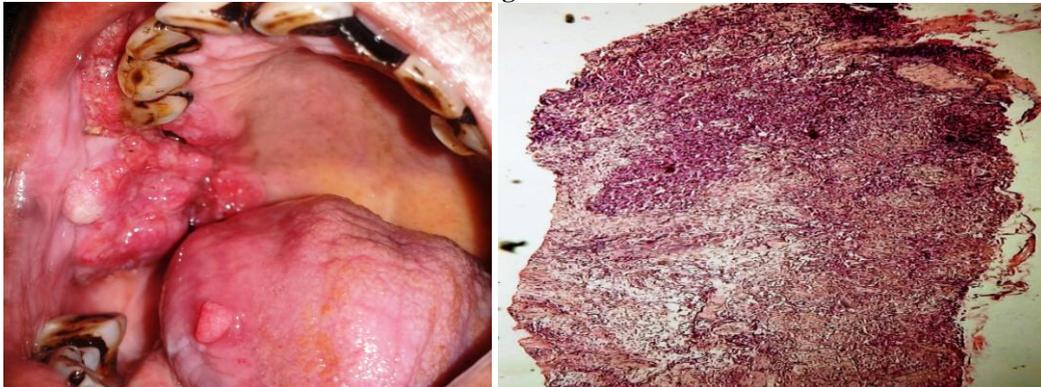


Figure 1: Intraoral photograph of the patient.

Figure 2: H and E stained section showing ulcerated stratified squamous epithelium infiltrating into underlying connective tissue stroma in a streaming pattern (4x magnification)

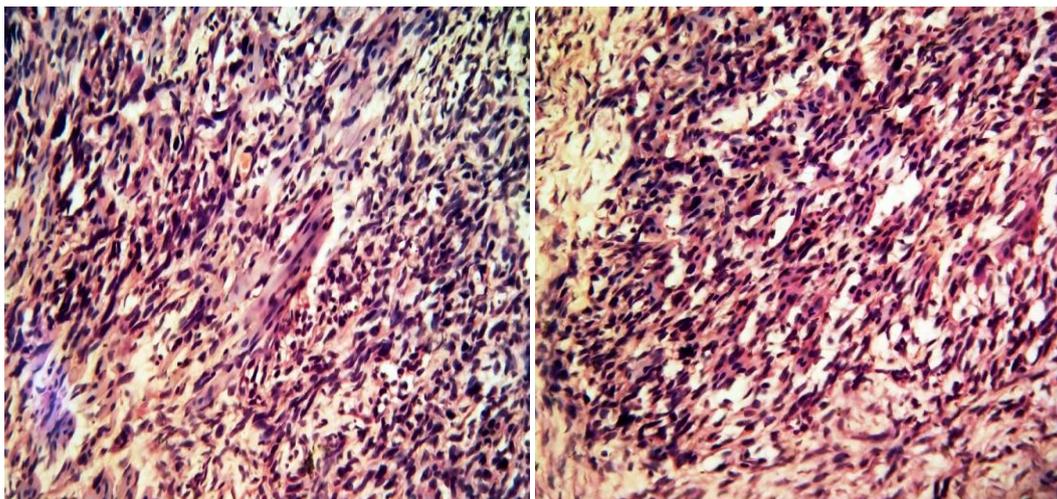


Figure 3: infiltrating bits of stratified squamous epithelium and round to ovoid tumor cells with high nuclear cytoplasmic ratio, hyperchromatism and pleomorphism. Numerous mitotic figures also noted (H/E 40x magnification).

Figure 4: advancing tumor front consisting of spindle cells in a streaming pattern (H/E 40x magnification).

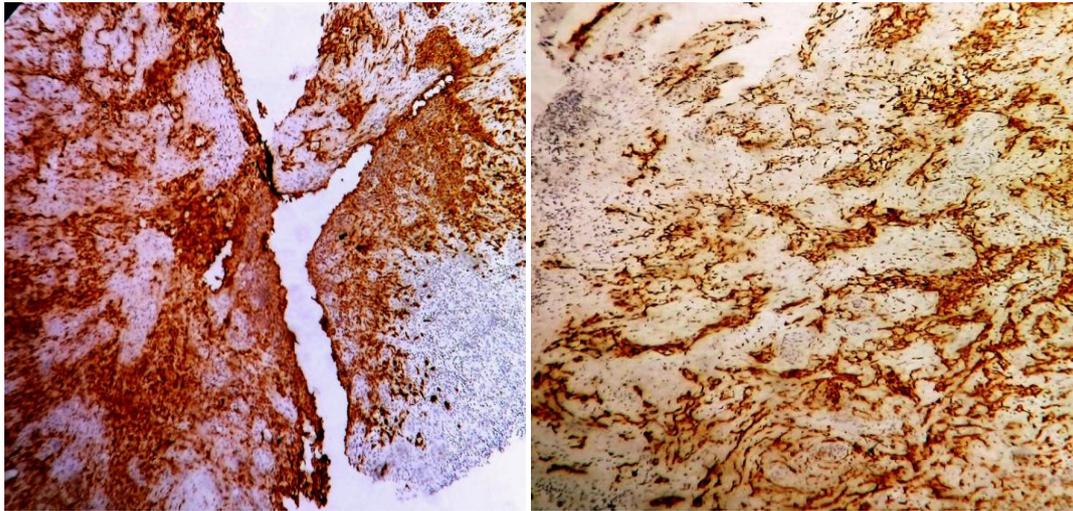


Figure 5: Tumor cells showing positivity for cytokeratin (AE1/AE3). Cytokeratin positive epithelium also seen (10 x magnification).

Figure 6: spindle shaped tumor cells showing positivity for vimentin (10 x magnification).