Case Report

DOI: 10.7860/JCDR/2016/17522.7261

Dentistry Section

Spindle Cell Carcinoma of the Mandibular Gingiva – A Case Report

SANGEETA R. PATANKAR[1], PRATYUSHA P. GAONKAR[2], PRACHI R. BHANDARE[3], NIDHI TRIPATHI[4], GOKUL SRIDHARAN[5]

ABSTRACT

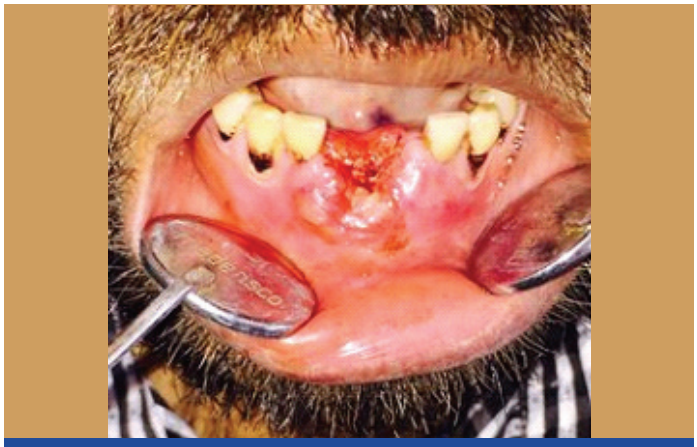
Spindle cell carcinoma is a malignancy of epithelial origin often mimicking its mesenchymal counterpart thus posing a diagnostic challenge. It is a rare biphasic malignant tumour mostly encountered in the upper aerodigestive tract. The chief differential diagnoses of spindle cell carcinoma are true superficial sarcomas and they especially need to be differentiated from fibrosarcoma. This presentation reports a spindle cell carcinoma of the gingiva and highlights the difficulties encountered in the diagnosis. It also emphasizes the importance of accurate and thorough diagnosis of malignant spindle cell lesions to determine the appropriate therapeutic modality.

Keywords: Biphasic tumour, Cytokeratin, Spindle cell lesions

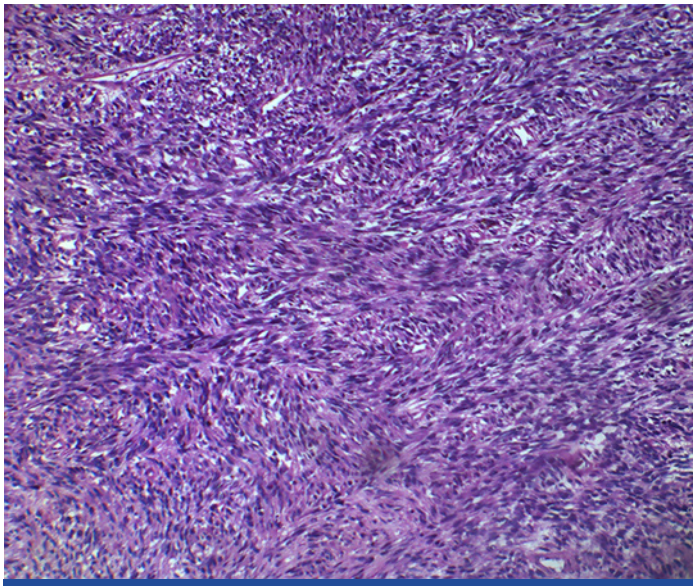
CASE REPORT

A 38-year-old male patient reported with a chief complaint of painful swelling in the mandibular anterior region since two months and mobility of lower incisors. Patient gave history of tobacco quid placement in lower labial vestibule since 7-8 years. Clinical examination revealed a soft, erythematous gingival swelling extending from lower left lateral incisor to right lateral incisor and grade III mobility with the lower central incisors. There was no evidence of regional lymphadenopathy. Radiographic investigations revealed severe vertical bone loss extending till the apex of the lower central incisors. A provisional diagnosis of periodontal pathology was considered. Extraction of both the lower central incisors was performed and soft tissue was curetted from the socket and submitted for histopathological evaluation [Table/Fig-1]. Microscopic examination revealed fascicular arrangement of spindle shaped cells with nuclear hyperchromatism and cellular and nuclear pleomorphism suggestive of malignant spindle cell neoplasm. The lesion grew in size during subsequent follow-up. On the basis of the clinical and histopathological findings, a segmental resection of the anterior mandible was performed under general anaesthesia.The gross specimen was a firm, greyish white mass 35mm x 32mm x 21 mm in size consisting of hard and soft tissue components.

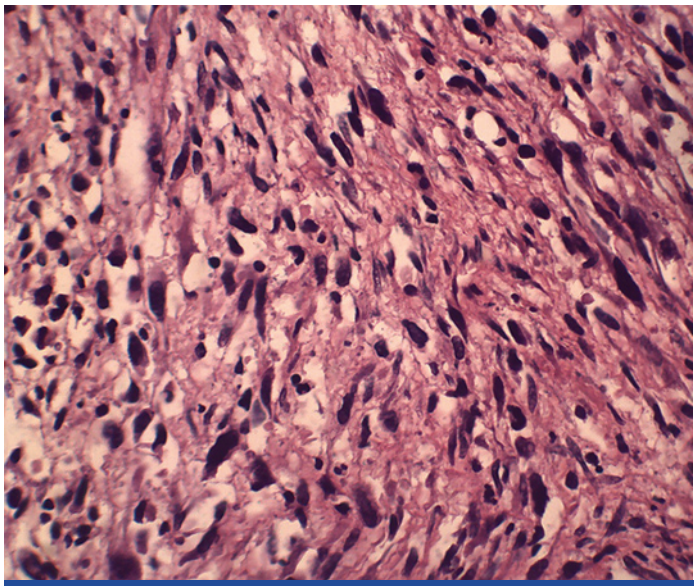
Microscopic examination of the excised specimen revealed a cellular connective tissue stroma with fascicular arrangement of elongated, spindle shaped cells exhibiting nuclear and cellular atypia [Table/Fig-2,3]. Based on these findings, diagnosis of



[Table/Fig-1]: Photograph of soft erythematous swelling on the anterior mandibular gingiva (published with the patient's consent).



[Table/Fig-2]: Photomicrograph showing cellular connective tissue stroma with malignant cells which are spindle, elongated, round in appearance and arranged in fascicular pattern (haematoxylin and eosin, original magnification x100).



[Table/Fig-3]: Photomicrograph showing malignant cells with cellular atypia in the form of nuclear hyperchromatism, pleomorphism, many abnormal mitotic figures (haematoxylin and eosin stain, original magnification x400).

spindle cell malignancy was given. The differential diagnoses considered were spindle cell carcinoma, spindle cell melanoma and mesenchymal neoplasms like fibrosarcoma. A panel of immunohistochemical markers were used to confirm the diagnosis. The tumour cells were positive for AE1/AE3, focally positive for EMA while negative for S-100, myogenin, desmin and p63. Based on the immunohistochemistry, a final diagnosis of spindle cell carcinoma was given. The patient was not available for follow-up and no further treatment was possible.

DISCUSSION

The variants of squamous cell carcinoma arising within the mucosa of the upper aerodigestive tract represent about 15% of squamous cell carcinomas in this region [1]. Spindle cell carcinoma, is an unusual and highly malignant variant of squamous cell carcinoma comprising upto 3% of squamous cell carcinomas in the head and neck region with propensity for occurrence in the larynx followed by the oral cavity and the nasal cavity [1]. The incidence rate of spindle cell carcinoma is about 0.59 percent of all upper aerodigestive tract neoplasms. Spindle cell carcinoma is an invasive carcinoma composed of malignant pleomorphic spindle cell component mimicking true sarcoma but of epithelial origin. The tumour has been described by various terminologies which reflect the controversy associated with its histogenesis [2]. The WHO classification of tumours of the oral cavity and oropharynx has placed this entity under a highly malignant variant of squamous cell carcinoma and labelled it as spindle cell carcinoma [3].

Spindle cell carcinoma of the oral cavity presents with a profound male to female predilection (11:1) and the mean age of occurrence is 57 years; it can be diagnosed in younger age group and very old age group (range 29-93 years). It has a site predilection for the lower lip, tongue and alveolar ridge or gingiva. In our case, the growth was seen in the gingiva of the anterior mandible. The growth often presents as an exophytic polypoid mass, but sessile, nodular or endophytic presentations have also been described in the literature. Prior history of radiation, trauma, poor oral hygiene, tobacco use or alcohol abuse are the potential risk factors [4]. Viswanathan et al., in an extensive clinicopathologic review of 103 cases of sarcomatoid carcinoma of the head and neck demonstrated that tobacco chewing (63.8%) was more frequently observed than smoking (21.3%) in these patients and none of them had pasthistory of radiation exposure [5]. In our case the patient was a habitual tobacco user in the form of tobacco quid placement in lower labial vestibule.

The different theories of histogenesis of spindle cell carcinomas have earned them an array of labels such as carcinosarcoma, sarcomatoid carcinoma, pleomorphic carcinoma, metaplastic carcinoma, polypoid carcinoma and pseudosarcomatous carcinoma.

The histogenesis of spindle cell carcinoma has been a very controversial and debatable topic for many years. With numerous histogenetic hypothesis having been put forth over the years, the three theories that are most accepted are: 1) Collision tumour or a carcinosarcoma; wherein the spindle and the epithelial cells are arising from separate stem cells; or 2) Pseudosarcoma – a squamous cell carcinoma with an atypical reactive stroma; 3) Spindle cell carcinoma or sarcomatoid carcinoma – wherein both epithelial and spindle cell components have epithelial origin with de-differentiation to a spindle cell morphology. The third theory has been lately reinforced by the following facts: their occurrence in the sites that normally contain squamous epithelium and a prevalence of carcinomas more than sarcomas; a location that is superficial in nature; simultaneous immunoreactivity of epithelial and mesenchymal markers with double labelling techniques in some neoplastic spindle cells and lastly the molecular studies showing its monoclonal origin [6-8].

Histopathological identification of squamous cell carcinoma in some parts of the tumour is essential for the diagnosis of spindle cell carcinoma. In various cases, however, malignant epithelium is frequently scarce and may be absent from the surface in many areas as was encountered in our case. The spindle cell component occupies the major portion of the neoplastic mass, often extending over the entire tumour surface, making recognition of the biphasic process challenging [9]. The chief differential diagnoses of spindle cell carcinoma are true superficial sarcomas and theyespecially need to be differentiated from fibrosarcoma [10]. As opposed to spindle cell carcinoma, fibrosarcoma often presents in young patients. The prognosis of fibrosarcoma is poor as compared to spindle cell carcinoma with a ten year survival rate less than thirty percent in high grade primary fibrosarcoma. In our case, the histopathological picture revealed only spindle shaped cells which represented sarcoma like tissue. It is prudent to consider a malignant spindle cell lesion a nonsarcomatous malignancy before it is accepted as a sarcoma.

Owing to the difficulties associated with the histopathological diagnosis, it is imperative to use epithelial and mesenchymal immunohistochemical markers to diagnose the tumour. The panel of epithelial markers include keratin (AE1/AE3, CK1, 8, 9), epithelial membrane antigens, KI, and K18 whereas mesenchymal markers include vimentin, desmin and S-100 [10,11]. The epithelial cells undergo phenotypic change to a spindle cell component in these tumours. These changes are generally associated with a variation in cytokeratin expression. Inspite of this, cytokeratin, is an essential aid in the diagnosis and identification of spindle cell carcinomas. Cytokeratins are proteins that comprise of the cytoskeleton of the epithelial cells and hence valuable in the demonstration of epithelial phenotype. Ramamurti et al., reported a case of spindle cell carcinoma wherein expression of podoplanin in the spindle cells was studied [12]. Podoplanin is a mucin like glycoprotein essential for lymphangiogenesis and can be used to assess the ability of tumour to spread. Though it is not useful as a diagnostic marker, it can be used to determine the invasion potential of these neoplasms. In our case, tumour cells were positive for AE1/AE3, focally positive for EMA while negative for S-100, myogenin, desmin and p63.

The overall survival of spindle cell carcinoma is poorer than squamous cell carcinoma. In a study conducted by Su et al., the one year overall survival rate of spindle cell carcinoma was 36.7% [13]. Surgical resection of the tumour with neck dissection is accepted as the best treatment of choice in the oral cavity.

CONCLUSION

Spindle cell lesions of the oral cavity are diagnostically challenging group of lesions that may be either carcinomatous or sarcomatous in origin. Appropriate diagnosis is necessary to decide on the correct treatment modality to reduce post-operative morbidity and mortality. Further, it is imperative that in the absence of advanced diagnostic aids, a microscopically diagnosed spindle cell lesion should be considered to be of epithelial origin and treated likewise unless proved otherwise.

REFERENCES

[1] Thompson LDR. Squamous cell carcinoma variants of the head and neck. *Curr Diagn Pathol*. 2003;9:384–96.
[2] Wenig BM. Squamous cell carcinoma of the upper aerodigestive tract: precursors and problematic variants. *Modern Pathology*. 2002;15:229–54.
[3] Parikh N, Desai N. Spindle Cell Carcinoma of the oral cavity: A casereport of a rare entity and review of literature. *J. Academy Adv Dental Research*. 2011;2:31-36.
[4] Okhtay 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.
[5] 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.

[6] Kwon GY, Choi YJ, Song MS, Yun KI. Sarcomatoid carcinoma of the mandible-report of a case. *J Korean Assoc Oral Maxillofac Surg*. 2010;36:228-30.
[7] Choi HR, Sturgis EM, Rosenthal DI, Luna MA, Batsakis JG, El- Naggar AK. Sarcomatoid carcinoma of the head and neck. Molecular evidence for evolution and progression from conventional squamous cell carcinomas. *Am J SurgPathol.* 2003;27: 1216-20.
[8] Kashiwabara K, Sano T, Oyama T, Najahima T, Makita F, Hashimoto N, et al. A case of oesophageal sarcomatoid carcinoma with molecular evidence of a monoclonal origin. *Pathol Res Prac*t. 2001;197:41-46.
[9] Michaels L, Hellquist HB. Ear, Nose and Throat Histopathology. 2nd edition. Springer 2001; 401.
[10] Sarma A, Das R, Sharma J, Kataki A. Spindle Cell Carcinoma of the Head and Neck: A Clinicopathological and Immunohistochemical Study of 40 Cases. *J Cancer Ther. 2012;3:1055-59.*
[11] Prakash N, Kumar H, 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(1):55-58.
[12] 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.
[13] Su HH, Chu ST, Hou YY, Chang KP, Chen CJ. Spindle cell carcinoma of the oral cavity and oropharynx: factors affecting outcome. *J Chin Med Assoc.* 2006;69:478-83.

PARTICULARS OF CONTRIBUTORS:
1. Professor and HOD, Department of Oral Pathology and Microbiology, Y.M.T Dental College and Hospital, Institutional Area, Navi Mumbai, Maharashtra, India.
2. Post Graduate Student, Department of Oral Pathology and Microbiology, Y.M.T Dental College and Hospital, Institutional Area, Navi Mumbai, Maharashtra, India.
3. Post Graduate Student, Department of Oral Pathology and Microbiology, Y.M.T Dental College and Hospital, Institutional Area, Navi Mumbai, Maharashtra, India.
4. Post Graduate Student, Department of Oral Pathology and Microbiology, Y.M.T Dental College and Hospital, Institutional Area, Navi Mumbai, Maharashtra, India.
5. Reader, Department of Oral Pathology and Microbiology, Y.M.T Dental College and Hospital, Institutional Area, Navi Mumbai, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr Pratyusha P. Gaonkar,
Department of Oral Pathology and Microbiology, Y.M.T Dental College and Hospital, Institutional Area,
Sector-4, Kharghar, Navi Mumbai-410210, Maharashtra, India
E-mail: pratyushagaonkar1@gmail.com

FINANCIAL OR OTHER COMPETING INTERESTS: None.

Date of Submission: **Oct 26, 2015**
Date of Peer Review: **Dec 02, 2015**
Date of Acceptance: **Dec 16, 2015**
Date of Publishing: **Feb 01, 2016**