

Case Report

Spindle cell carcinoma in the maxilla: A rare case and literature review

M. Varshini¹, Varsha Salian¹, Pushparaja Shetty¹, Shalini Krishnan²

Departments of ¹Oral Pathology and Microbiology and ²Oral and Maxillofacial Surgery, A.B. Shetty Memorial Institute of Dental Sciences, Nitte (Deemed to be University), Mangalore, Karnataka, India

Received: September 2017

Accepted: March 2018

Address for correspondence:

Dr. Varsha Salian,
Department of Oral
Pathology and Microbiology,
A.B. Shetty Memorial
Institute of Dental Sciences,
Nitte (Deemed to be
University), Deralakatte,
Mangalore - 575 018,
Karnataka, India.
E-mail: varu_2002@yahoo.
com

ABSTRACT

In India, oral squamous cell carcinoma accounts for 90%–95% of oral malignancies. The WHO classifies spindle cell carcinoma (SpCC) under malignant epithelial tumors of squamous cell carcinoma (SCC) and is a rare entity accounting for only 1% of SCCs. It is an aggressive biphasic neoplasm exhibiting high mortality rate owing to increased metastasis and recurrence which signifies the need for recognition and treatment of this perplexed tumor. We present a case of maxillary SpCC where histopathological evaluation alone was indecisive, requiring immunohistochemistry for confirmation of the diagnosis.

Key Words: Maxillary bone, spindle cell carcinoma, squamous cell carcinoma

INTRODUCTION

Spindle cell carcinoma (SpCC) is an uncommon aggressive biphasic malignancy that has the propensity to manifest itself in the upper aerodigestive tract, including the oral mucosa. The WHO defines this tumor as “carcinoma within which there are some elements resembling a squamous cell carcinoma that are associated with a spindle cell component.”^[1] A handful of terminologies such as carcinosarcoma, collision tumor, pseudocarcinoma, sarcomatoid squamous cell carcinoma (SCC), and pleomorphic carcinoma have been used in the literature to portray its histopathological presentation,^[2] but in most cases, the tumor appears monophasic, with the spindle cell element dominating the histology.^[3] This makes the diagnosis of SpCC an enigma without electron

microscopy and immunohistochemistry. Reports of SpCC cases in the maxilla are rare but have been reported in the past.^[4] We present a case of SCC recurring as SpCC in the maxilla which was further confirmed with immunohistochemistry.

CASE REPORT

A 62-year-old male presented with a complaint of swelling and discharge in the right upper jaw which was progressively increasing in size. The patient gave a history of tobacco consumption for the past 20 years. Medical history revealed the presence of SCC in the same region that was treated by hemimaxillectomy and neck dissection followed by radiotherapy. Extraoral examination revealed gross asymmetry of the face

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

How to cite this article: Varshini M, Salian V, Shetty P, Krishnan S. Spindle cell carcinoma in the maxilla: A rare case and literature review. Dent Res J 2019;16:60-3.

Access this article online



Website: www.drj.ir
www.drjjournal.net
www.ncbi.nlm.nih.gov/pmc/journals/1480

corresponding to the previous site of surgery. Lymph nodes were nonpalpable. Intraoral examination showed a soft-tissue mass on the right posterior alveolus measuring 4 cm × 3 cm [Figure 1]. Incisional biopsy of the lesion was done [Figure 2], and immunohistochemistry was performed on paraffin-embedded tissues as per the manufacturer's protocol.

Histopathological examination of the incisional specimen showed ulcerated mucosa with areas of focal keratinization and extensive granulation tissue. Connective tissue stroma showed spindle-shaped and polygonal cells arranged in haphazard sheets showing pleomorphism, high mitotic activity, and atypia suggestive of malignancy [Figure 3a and b]. Immunohistochemistry showed positivity for pan-cytokeratin in the spindle-shaped cells [Figure 4a-c], and thus, considering the clinical history and immunohistochemical finding, a final

diagnosis of spindle cell variant of SCC was made. In the present case, the patient could not be operated due to practical difficulties, and hence, radiotherapy was advised as palliative treatment. The patient was lost to follow-up.

DISCUSSION

Virchow first described SpCC in 1864^[5] as a biphasic tumor characterized by areas of SCC in conjugation with sarcomatoid proliferation of spindle cells. The term SpCC was proposed by Sherwin *et al.* and accepted by the WHO under the malignant epithelial tumors of SCC.^[6,7] There is a high male predominance (male: female = 11: 1) and commonly occurs in the 6–7th decade of life.^[2] Risk factors include tobacco usage, especially cigarette smoking, alcohol, and radiation exposure.^[3,7] This case was seen in a 62-year-old male patient with a history of tobacco and alcohol consumption for the past 20 years as seen in previous literature.^[2,3,7]



Figure 1: Ulceroproliferative mass on the right posterior alveolus measuring 4 cm × 3 cm.

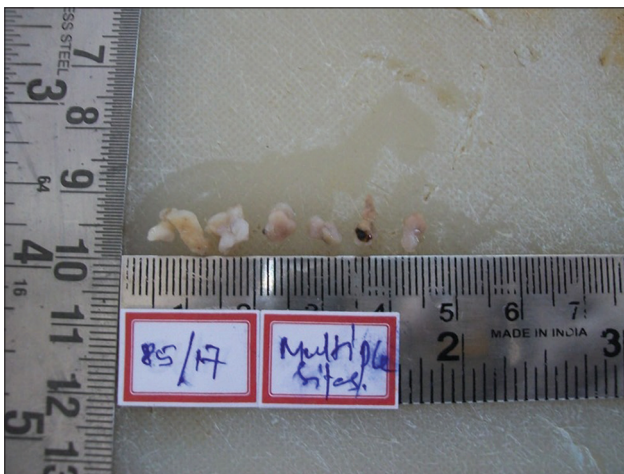


Figure 2: Incisional biopsy specimen from multiple sites of the lesion.

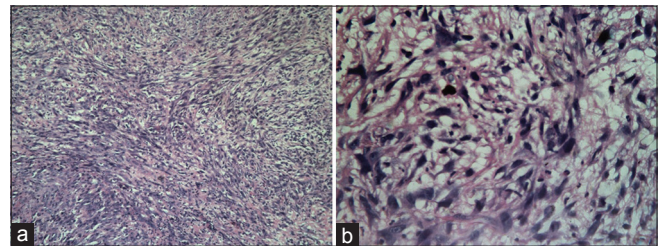


Figure 3: (a) Sheets of spindle-shaped cells admixed with haphazardly arranged polygonal cells (H and E, ×100), (b) Polygonal and spindle cells showing pleomorphism, high mitotic activity, and atypia (H and E, ×400).

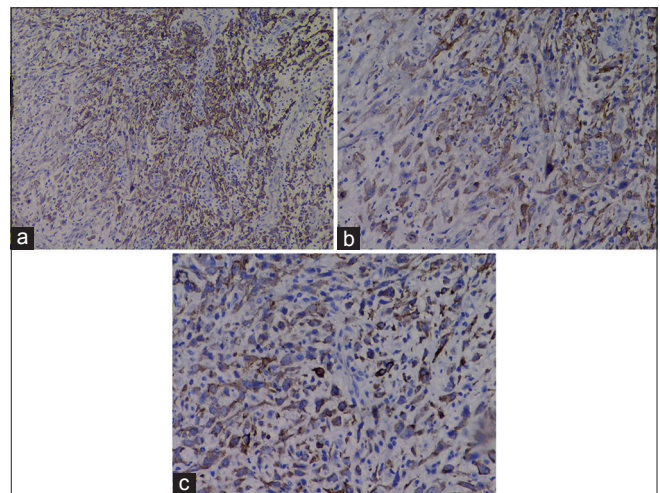


Figure 4: (a) Neoplastic spindle-shaped cells showing positivity for pan-cytokeratin (×100), (b and c) neoplastic spindle-shaped cells showing positivity for pan-cytokeratin (×400).

SpCCs are most common in the oral cavity and larynx, sinonasal areas, and pharynx. In the oral cavity, they are frequently encountered in the lower lip, tongue, buccal mucosa, alveolar ridge, and gingiva.^[3] SpCCs in the oral cavity have been published by many authors previously. Rizzardi *et al.* reported a case of SpCC in the tongue and floor of mouth.^[8] Su *et al.* in their study quoted 15 cases of SpCC arising in different locations in the oral cavity, of which tongue was the most common site.^[9] Reports of three cases of SpCC in the mandibular alveolus have been published previously.^[1,2,10] Nineteen cases of SpCC occurring in the maxilla have been documented from 1957 to 2015 where majority of cases complied with previous literature with regard to age and gender.^[4] Dedifferentiated cases with previous history of conventional SCC with recurrent sarcomatoid carcinoma have also been published in literature. Tse *et al.* reported a recurrent case of SpCC following radiotherapy.^[11] Minami *et al.* published a case of SpCC of the palatine tonsil, with a history of SCC previously treated with radiotherapy.^[12] Chang *et al.* in their 30-year review mentioned 29 cases of sarcomatoid carcinoma with a previous history of SCC.^[13] Pittman *et al.* in their analysis of three cases mentioned the recurrence of SCC as sarcomatoid carcinoma in two cases located in the bronchial and esophageal regions.^[14] Okuyama *et al.* also published a case of recurrent SpCC in the tongue after glossectomy for well-differentiated SCC [Table 1].^[15] Ours may be the next case following, which shows that although the lesion is uncommon in the jaws, the trend may be changing recently.

The classical picture of SpCC comprises ulceration of the overlying epithelium with areas of SCC admixed with areas of pleomorphic spindle cells. Sharp borders between SCC areas and spindle cell component and/or gradual transition with the SCC cells dropping off from epithelial nests into spindle cell areas may be seen.^[3] Theories on origin of spindle cells suggest that they are either variant growth pattern of SCC, nonneoplastic reactive phenomena, or admixture of malignant

epithelial and mesenchymal cells.^[9] Bizarre granulation tissue formed by exposure to ionizing radiation and chemotherapy present with pleomorphic spindle cells with atypical mitotic figures poses great difficulty in diagnosis. This case showed the presence of ulcerated surface epithelium and granulation tissue with mixture of dysplastic polygonal and spindle cells. Such appearance coupled with the fact that there has been a previous history of SCC at the same site could be considered as reliable diagnostic parameters for SpCC in the absence of immunohistochemistry. Both epithelial and mesenchymal markers have been used for confirmation of SpCC. Recent theory supports the monoclonal hypothesis that states that they arise from the same stem cells and have undergone “dedifferentiation.”^[16] This phenomenon of “dedifferentiation” can occur after radiotherapy, leading to a more differentiated SCC.^[17] We further confirmed the diagnosis with cytokeratin positivity in the spindle cells.

Differential diagnoses for SpCCs are fibrosarcoma, angiosarcoma, malignant fibrous histiocytoma, leiomyosarcoma, malignant melanoma, malignant peripheral nerve sheath tumor, osteosarcoma, mesenchymal chondrosarcoma, Kaposi sarcoma, synovial sarcoma, leiomyoma, and reactive epithelial proliferations.^[7] Negativity for CD31 and CD34 differentiates it from angiosarcoma. Fibrosarcoma and leiomyosarcoma show an intervening fibrous layer often separating the epithelial and stromal components in sarcomas. Smooth Muscle Actin (SMA) and Desmin positivity differentiates leiomyosarcoma and myoepithelial tumors from SpCC. Mucosal melanomas can be isolated from SpCC by their positivity for Melan-A and HMB-45.^[2] Surgery and radiotherapy form the mainstay of treatment. In inoperable cases, radiotherapy has been considered as an acceptable alternative.^[9]

CONCLUSION

SpCC of the maxilla is no more a rare entity, with more cases being reported recently. The aggressive

Table 1: Dedifferentiated cases of spindle cell carcinoma following radiotherapy

Author names and year	Number of case(s)	Diagnosis	Location	Previous history of SCC
Tse <i>et al.</i> , 1987 ^[11]	1	Sarcomatoid carcinoma	Oral mucosa: Floor of the mouth	Yes
Minami <i>et al.</i> , 2008 ^[12]	1	Sarcomatoid carcinoma	Palatine tonsil	Yes
Chang <i>et al.</i> , 2013 ^[13]	29	Sarcomatoid carcinoma	Tongue, buccal mucosa, gums	Yes
Pittman <i>et al.</i> , 2016 ^[14]	2	Sarcomatoid carcinoma	Endobronchial and esophageal mass	Yes
Okuyama <i>et al.</i> , 2017 ^[15]	1	Sarcomatoid carcinoma	Tongue	Yes

SCC: Squamous cell carcinoma

and recurrent nature of this tumor may require early diagnosis and prompt intervention.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

The authors of this manuscript declare that they have no conflicts of interest, real or perceived, and financial or nonfinancial in this article.

REFERENCES

1. Al-Bayaty H, Balkaran RL. Spindle cell carcinoma of the mandible: Clinicopathological and immunohistochemical characteristics. *J Oral Biol Craniofac Res* 2016;6:160-3.
2. Bavle RM, Govinda G, Venkataramanaiah PG, Muniswamappa S, Venugopal R. Fallacious carcinoma- spindle cell variant of squamous cell carcinoma. *J Clin Diagn Res* 2016;10:ZD05-8.
3. Slootweg PJ, Richardson M. Squamous cell carcinoma of the upper aerodigestive system. In: Gnep DR, editor. *Diagnostic Surgical Pathology of the Head and Neck*. Philadelphia: Saunders; 2001. p. 54-7.
4. Junaid M, Kazi M, Qadeer S. Spindle cell carcinoma of the maxilla: A case report of rare entity. *Surg Sci* 2017;8:220-7.
5. Virchow R. *Die Krankhaften Geschwulste*. Vol. 2. Berlin, Germany: Hirschwald; 1864-1865. p. 181-2.
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. Samuel S, Sreelatha SV, Hegde N, Nair PP. Spindle cell carcinoma in maxilla. *BMJ Case Rep* 2013;2013. pii: bcr2013009611.
8. Rizzardi C, Frezzini C, Maglione M, Tirelli G, Melato M. A look at the biology of spindle cell squamous carcinoma of the oral cavity: Report of a case. *J Oral Maxillofac Surg* 2003;61:264-8.
9. 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.
10. Patankar SR, Gaonkar PP, Bhandare PR, Tripathi N, Sridharan G. Spindle cell carcinoma of the mandibular gingiva – A case report. *J Clin Diagn Res* 2016;10:ZD08-10.
11. Tse JJ, Aughton W, Zirkin RM, Herman GE. Spindle cell carcinoma of oral mucosa: Case report with immunoperoxidase and electron microscopic studies. *J Oral Maxillofac Surg* 1987;45:267-70.
12. Minami SB, Shinden S, Yamashita T. Spindle cell carcinoma of the palatine tonsil: Report of a diagnostic pitfall and literature review. *Am J Otolaryngol* 2008;29:123-5.
13. Chang NJ, Kao DS, Lee LY, Chang JW, Hou MM, Lam WL, *et al.* Sarcomatoid carcinoma in head and neck: A review of 30 years of experience – Clinical outcomes and reconstructive results. *Ann Plast Surg* 2013;71 Suppl 1:S1-7.
14. Pittman PD, Guy CD, Cardona DM, McCall SJ, Zhang X. Sarcomatoid carcinoma of the esophagus. *JSM Gastroenterol Hepatol* 2016;4:1067.
15. Okuyama K, Fujita S, Yanamoto S, Naruse T, Sakamoto Y, Kawakita A, *et al.* Unusual recurrent tongue spindle cell carcinoma with marked anaplasia occurring at the site of glossectomy for a well-differentiated squamous cell carcinoma: A case report. *Mol Clin Oncol* 2017;7:341-6.
16. 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;4:265-75.
17. Neville BD, Damm DD, Allen CM, Chi A. *Oral and Maxillofacial Pathology: First South Asia Edition*. Gurgaon: EIH Limited- Unit Printing Press; 2016.