

A Case Report on Rare Entity: Spindle Cell Carcinoma of Maxillary Alveolus

Dr. Anju Mathew, Senior Lecturer, Department of Oral Medicine and Radiology, Pushpagiri College of Dental Sciences, Kerala, India.

Corresponding Author: Dr. Anju Mathew, Senior Lecturer, Department of Oral Medicine and Radiology, Pushpagiri College of Dental Sciences, Kerala, India.

Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Background: Spindle Cell Carcinoma also known as sarcomatoid carcinoma is a rare biphasic malignant neoplasm and an unusual form of poorly differentiated squamous cell carcinoma that occurs mainly in the upper aerodigestive tract. This tumour was labelled by World Health Organisation and they classified this tumour under a highly malignant variant of squamous cell carcinoma. This article deals with reporting an unusual case of exophytic spindle cell carcinoma of maxilla.

Methods: A 60 year old male patient presented with growth in the left maxillary alveolus region for past 3 months. Clinical examination revealed well defined exophytic growth present on the left side of maxillary alveolus extending to hard palate measuring with absence of bleeding or discharge. The patient was advised to get panoramic radiograph and Computed tomography done. With proper consent, an incisional biopsy was obtained from the growth.

Results: The diagnosis of spindle cell carcinoma was made and he patient was referred for treatment consisting of surgical excision of the tumour. Patient was reviewed periodically and healing was satisfactory with no signs of recurrence.

Conclusion: Spindle cell carcinoma involving head and neck region is a rare and unique variant of squamous cell

carcinoma which potentially aggressive than the classical squamous cell carcinoma. It has high recurrence rate and greater tendency to metastasize. Patients diagnosed at an early stage have excellent prognosis while those whose tumors are deeply invasive have a poor prognosis.

Keywords: Spindle cell carcinoma, squamous cell carcinoma, sarcomatoid carcinoma Computed tomography, maxillary alveolus, biphasic neoplasm.

Introduction

Sarcomatoid Carcinoma (SC) or Spindle cell carcinoma is a rare morphological and aggressive variant of squamous cells carcinoma that occurs principally in the upper digestive tract. This tumour is designated by various terms spindle cell squamous carcinoma, car-cinosarcoma, pleomorphic carci-nomal, and pseudosarcoma.^[1] Spindle cell carcinoma is considered to be a biphasic tumor as it is composed of a squamous cell carcinoma (in situ or invasive) and spindle cell carcinoma with sarcomatous appearance.^[2] WHO has classified this tumour as highly malignant variant of squamous cell carcinoma and has titled it as spindle cell carcinoma.^[3] Spindle cell carcinoma is very infrequent with a reported incidence of less than 1% of all tumours of oral regions and 3% of Squamous cell carcinoma^[4,5].

This article aims to report a case of 60 year old male patient diagnosed with spindle cell carcinoma of the left maxillary alveolus.

Case Report

A 60 year old male patient presented with exophytic growth in the upper left back tooth region for past 3 months. Patient noticed loosening of teeth in the left upper back tooth region which got self-exfoliated followed by a small growth in the same region which gradually increased to attain present size. No history of bleeding or discharge from the growth. No history of headache or nasal blockage. Patient had no habit of smoking or consuming alcohol. Lymph node examination revealed single left submandibular left node palpable 0.5cm in size, firm in consistency, non- tender and fixed to the underlying structures.

On intra oral examination, well defined exophytic growth present on the left side of maxillary alveolus extending to hard palate measuring about 3x4cm extending anteroposterioly from 25 to 28 region and mediolateraly from the maxillary alveolar mucosa to 2cm away from midpalatine raphe. Surface of the growth is irregular with erythematous patches. Colour of the growth same as that to adjacent mucosa. No bleeding or discharge present. [Figure 1]

The patient was advised to get Panoramic radiograph [Figure 2] and Computed tomography [Figure 3] done. With proper consent, an incisional biopsy was obtained from the growth. The histopathological diagnosis of spindle cell carcinoma was made and he patient was referred for treatment consisting of surgical excision of the tumour [Figure 4]. Final histopathology report was the same as incisional biopsy [Figure 5]. Patient was reviewed periodically [Figure 6] and post-operative panoramic radiograph was taken [Figure 7]. Healing was satisfactory with no signs of recurrence.

Figure 1: Intraoral view



Figure 2: Panoramic radiograph



Panoramic radiograph reveals root remnant in left upper back teeth region with soft tissue shadow surrounding it. The alveolar bone level is reduced on left side when compared to right side.

Figure 3: CT 3D Reconstructed



Figure 4: Intra operative View

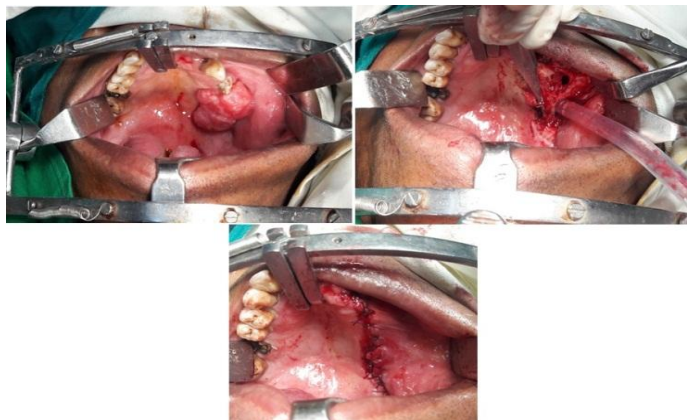


Figure 5: Photomicrograph



Discussion

Virchow in 1864 initially described this malignancy and titled as carcinosarcoma. Further research was carried out in 153 cases of carcinosarcoma by Saphir and Vass that occurred at various sites and noted sarcomatous component represented a variation in the squamous portion of the carcinoma.^[6]

Spindle cell carcinoma was proposed to be a collision tumor, composition tumor or combination tumors^[7]. There was a great misperception on whether this tumor is carcinoma or sarcoma, and benign or malignant. When studies were carried out on basis of their morphologic, immune histochemical, ultra structural and molecular features of spindle cell carcinoma, they concluded that this tumour is recognized as a carcinoma that has surface epithelial changes and an underlying spindle-shaped neoplastic proliferation.^[5]

Spindle cell carcinoma most frequently affects elderly men, with a peak occurrence in the 6th to 7th decade^[8]. Approximately 50% to 70% of head and neck spindle cell carcinoma presents as exophytic or polypoid lesions, occasionally with a stalk.^[9]

The tumour has a site predilection for the lower lip, tongue and alveolar ridge or gingiva. Previous history of radiation, trauma, poor oral hygiene, tobacco use or alcohol abuse are the potential risk factors.^[10] In our case,

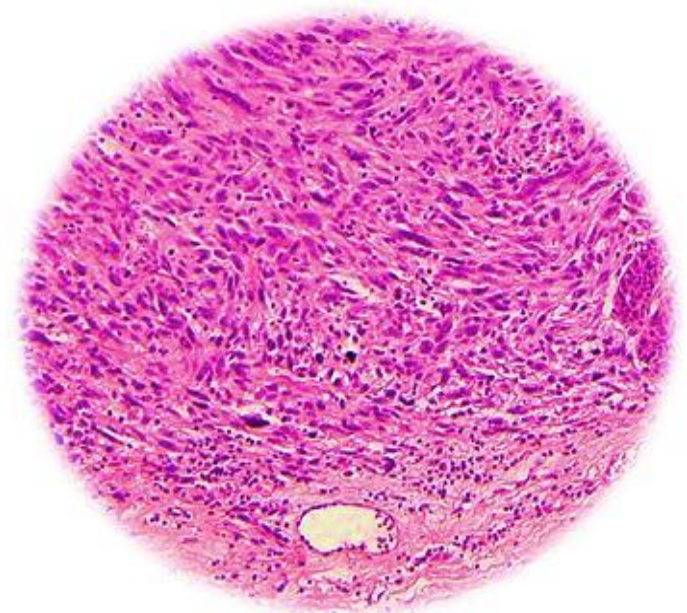


Figure 6: Post-operative View - Review after 1 month.



Figure 7: Post-operative radiograph

it was an exophytic growth on the alveolar ridge with no symptoms of pain. In our case, there were no predisposing factors.

The differential diagnosis includes squamous cell carcinoma, fibrosarcoma, malignant fibrous histiocytoma, Kaposi's sarcoma, leiomyosarcoma, rhabdomyosarcoma, angiosarcoma, synovial sarcoma, malignant melanoma, fibromatosis, leiomyoma, nodular fasciitis, malignant peripheral nerve sheath tumour, osteosarcoma, mesenchymal chondrosarcoma and reactive epithelial proliferations.^[11]

On histopathological examinations, bulk of the tumor is made up of sarcomatous component and consists of plump spindle cells, which can be rounded and epithelioid in some areas. It usually represents a fasciculated pattern which is composed of highly cellular groups of elongated bipolar cells in a parallel, interwoven alignment. A peculiar feature of this tumor is the relative inadequacy of the carcinomatous component which creates a dilemma as the histopathologic diagnosis becomes dependent on the site of the biopsy.^[1]

Routine light microscopy cannot predict the morphology of the spindle cells in Spindle cell carcinoma but it requires the use of immunohistochemistry (IHC). Cytokeratin (CK) is considered the most dependable epithelial marker but epithelial membrane antigen (EMA) and carcinoembryonic antigen (CEA) can also be useful.^[12]

Management of SC is as complicated and debateable as its diagnosis. The most acceptable and successful treatment modality is wide surgical excision, with or without radical neck dissection. Many authors consider radiotherapy to be ineffective, but it is an acceptable alternative for inoperable patients as well as for those in which the surgical margins are positive or in patients with nodal metastasis.^[13]

Prognosis of Spindle cell carcinoma is highly controversial and is dependent on location, size, and depth of invasion of tumor, stage of disease, and the presence of any keratin staining in the spindle cells. Spindle cell carcinoma of the oral cavity and oropharynx is potentially aggressive and tends to recur and metastasize easily.^[14] Some literatures stated that the prognosis is poorer than the conventional squamous cell carcinoma at a similar stage. About 30% of all oral cases ended with mortality within one year. The outcome is similar to the high grade squamous cell carcinoma.^[15]

Conclusion

The SC is an aggressive and rare variant of the squamous cell carcinoma creating a dilemma for diagnostician and oral pathologist whose histogenesis is controversial. Diagnosis should include biopsy of the lesion from different sites to possibly include both the epithelial and sarcomatous components. Keeping this in mind, IHC support is essential and understanding of its clinicopathologic characteristics, which is fundamental for the diagnosis and for an appropriate clinical management. The prognosis of disease is controversial. Patients diagnosed at an early stage have excellent prognosis while those whose tumors are deeply invasive have a poor prognosis.

References

1. C. Rizzardi, C. Frezzini, M. Maglione, G. Tirelli, and M. Melato, "A look at the biology of spindle cell squamous carcinoma of the oral cavity: report of a case," *Journal of Oral and Maxillofacial Surgery*, vol. 61, no. 2, pp. 264–268, 2003. DOI: 10.1053/joms.2003.50025
2. N. Katase, R. Tamamura, M. Gunduz et al., "A spindle cell carcinoma presenting with osseous metaplasia in the gingiva: a case report with immunohistochemical

- analysis,” *Head and Face Medicine*, vol. 4, no. 1, article 28, 2008. **DOI:** org/10.1186/1746-160X-4-28
3. Parikh N, Desai N (2011) Spindle Cell Carcinoma of the oral cavity: A case report of a rare entity and review of literature. *J Academy Adv Dental Research* 2: 2 **DOI:** org/10.1177%2F2229411220110206
4. Boamah H, Ballard B (2012) A case report of spindle cell (sarcomatoid) carcinoma of the larynx. *Case Rep Med* 2012: 370204. 2. Parikh N, Desai N (2011) Spindle Cell Carcinoma of the oral cavity: A case report of a rare entity and review of literature. *J Academy Adv Dental Research* 2: 2. PMID: 23319954 **DOI:** 10.1155/2012/370204
5. Ravindran R, Mohan V, Saji AM (2013) Spindle Cell Carcinoma of Maxilla: Case report of a rare entity and review of literature. *Oral & Maxillofacial Pathology Journal*: 4.
6. Berthelet E, Shenouda G, Black MJ, et al. Sarcomatoid carcinoma of the head and neck. *Am J Surg* 1994;68:455–8. PMID: 7977972
7. Thompson LD, Wieneke JA, Miettinen M, Heffner DK (2002) Spindle cell (sarcomatoid) carcinomas of the larynx: a clinicopathologic study of 187 cases. *Am J Surg Pathol* 26: 153-170. PMID: 11812937
8. Leventon GS, Evans HL. Sarcomatoid squamous cell carcinoma of the mucous membranes of the head and neck: a clinicopathologic study of 20 cases[J]. *Cancer*, 1981, 48(4):994–1003. PMID: 7272943
9. Barnes L. Spindle cell carcinoma, sarcomatoid carcinoma, carcinosarcoma. In: Barnes L, Editor. *Surgical pathology of the head and neck*, vol. 1, 3rd ed. New York (NY)7 Marcel Dekker, 2001, 174–181.
10. 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.
11. Samuel S, Sreelatha SV, Hegde N, et al. Spindle cell carcinoma in maxilla. *BMJ Case Rep* 2013 **DOI:** 10.1136/bcr-2013-009611
12. J. G. Batsakis and P. Suarez, “Sarcomatoid carcinomas of the upper aerodigestive tracts,” *Advances in Anatomic Pathology* , vol. 7, no. 5, pp. 282–293, 2000. PMID:10976907
13. Anshul Mahajan, Sujata Mohanty,Sujoy Ghosh, Aadithya B. Urs,Nita Khurana, and Sunita Gupta. Sarcomatoid Carcinoma of the Oral Cavity: A Diagnostic Dilemma. *Case Reports in Dentistry*. 2017 **DOI:** org/10.1155/2017/7495695
14. G. Y. Kwon, Y. J. Chol, M. S. Song, and K. I. Yun, “Sarcomatoid carcinoma of the mandible: report of a case,” *Journal of the Korean Association of Oral and Maxillofacial Surgeons*, vol. 36, no. 3, pp. 228–230, 2010
15. Anupam Sarma, Rajjyoti Das, Sharma JD, Kataki AC (2012) Spindle Cell Carcinoma of the Head and Neck: A Clinicopathological and Immunohistochemical Study of 40 Cases. *Journal of Cancer Therapy* 3: 1055-1059. **DOI:** 10.4236/jct.2012.36137