

Spindle cell carcinoma: A case report with review

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Abstract

Spindle cell carcinoma is a distinct entity with poor prognosis when compared to classical squamous cell carcinoma. Hence a careful histopathological analysis is warranted and the treatment modalities need to be modified. This tumor with biphasic proliferation assumes most often a sarcomatous appearance. A case report and a brief review is presented on this controversial lesion.

Key words: Spindle cell carcinoma, Sarcomatoid squamous cell carcinoma

Introduction

Spindle cell carcinoma is a malignant neoplasm with predilection for upper aerodigestive tract. The lesion is also known as Sarcomatoid squamous cell carcinoma or Polypoid squamous cell carcinoma. Though the lesions are rare in oral cavity, they can occur on the lip followed by tongue, alveolar ridge and the gingiva. The lesion is characterized by a dysplastic squamous epithelium in conjunction with an invasive spindle cell element¹.

Ocasional spicules of calcified material can be observed in between malignant cells which shows an osteosarcomatous appearance².

Case report

A 51 year old female patient reported to the Department of Oral Medicine & Maxillofacial Radiology, SPPGIDMS, Lucknow, India with a swelling in lower right side of the face and an intraoral growth. The swelling was observed by the patient nearly 5 months prior to the visit to the department. Patient reported with continuous dull pain and discomfort. The swelling appeared to have increased gradually over a period of time. Patient had poor oral hygiene and was a tobacco chewer since the past 30 years.

On extraoral examination a diffuse swelling was observed in relation to the right of angle of the mandible extending anteriorly to the corner of the mouth. Inferiorly the swelling extended 1cm below the lower border of the

mandible. The right submandibular lymph nodes were palpable and fixed to the underlying structures.

Intraorally, the swelling extended from right premolar region to the retromolar region (Fig. 1). It was found to be a fleshy, sessile growth approximately 3.5cm x 2.5cms in size with an irregular surface. On palpation the swelling was tender and firm in consistency. The second molar tooth was found to be encircled by the growth and it showed Grade II mobility.

Radiographic examination revealed a lytic lesion in the right molar area. The margins were ill defined and right buccal cortical plate was found to be breached. OPG revealed characteristic irregular destruction of bone in right lower premolar molar area. (Fig. 2)

Considering the above features, a differential diagnosis of Squamous cell carcinoma, Verrucous carcinoma and Metastatic carcinoma were arrived at.

An incisional biopsy was advised and the histopathological examination revealed dysplastic stratified squamous epithelium and underlying connective tissue. The epithelium was ulcerated and showed the dropping off effect and proliferation of neoplastic cells in a streaming manner. The neoplastic spindle cells characteristically showed pleomorphism of cells and hyperchromatism of the nuclei. The cells were arranged in large sheets

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and whorls. Occasional mitotic figures were observed in the sections. Chronic inflammatory cells were seen distributed diffusely through out the section (Fig. 3 & 4). The overall features were in favour for diagnosis of a Spindle cell carcinoma.

Patient was referred to Cancer Hospital for further management. Wide excision and radical neck dissection followed by radiation was performed by the oncology department. Patient is currently free of recurrence almost an year after instituting the treatment.

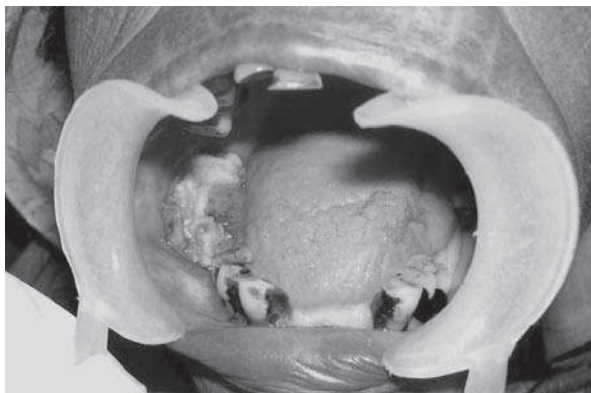


Fig 1: Intraoral photograph showing the exophytic lesional tissue in relation to lower right molar region.



Fig 2: OPG showing radiolucent area around the lower right and left molar tooth

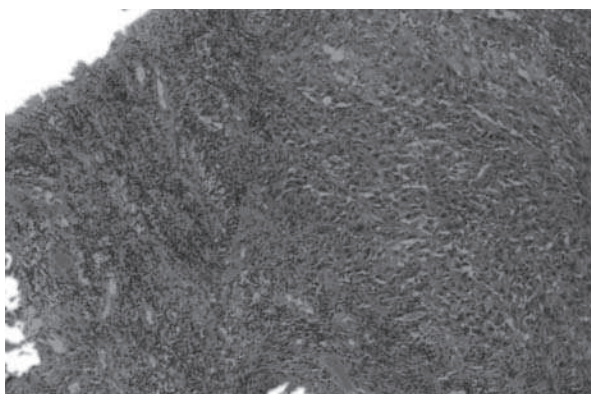


Fig 3: Photomicrograph (H&E) showing dysplastic stratified squamous epithelium with infiltration into underlying connective tissue. (X20)

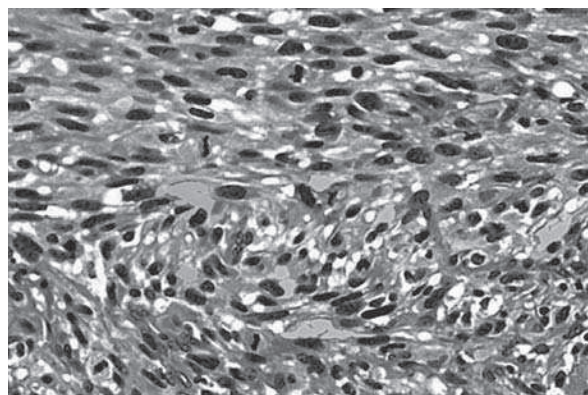


Fig 4: Photomicrograph (H&E) showing the spindle shaped tumour cells which characteristically reveal marked pleomorphism of cells and hyperchromatism of the nuclei. (X40)

Discussion

The mean age of diagnosis of Spindle cell carcinoma is 57 yrs. However it can be diagnosed in younger age group and very old age group (range 29-93yrs). As far the gender predilection is concerned most authors reported that there is equal frequency in both males and females. Some studies have reported a male predilection³.

Upon reviewing the precipitating or risk factors associated with this lesion, it was observed that the risk factors were similar to those that are involved in occurrence of Squamous cell carcinoma, chronic tobacco usage being the prominent factor.

The neoplasm can occur anywhere in upper aero digestive tract which includes larynx and esophagus. In the mouth, lower lip, lateral posterior tongue and alveolar ridges are the most commonly involved sites, but any area in oral cavity can be involved⁴.

Clinical appearance of Spindle cell carcinoma of the oral cavity varies and most lesions appears as pedunculated and polypoid masses Ellis and Corio have reviewed many cases and mentioned that the appearance of Spindle cell carcinoma predominantly occurred as exophytic large mass that increases in size over a short period of time¹.

However other investigators reported that Spindle cell carcinoma sometimes appear as a nodular and fungating mass, and may be associated with a non healing ulcer⁶.

Many cases eventually end up with pronounced parasthesia as the lesion increases in size involving the nerves in the affected region. It was also commented that lower lip lesions have a special propensity to travel along the nerves through the mental foramen and into the mandibular canal⁴.

Pain and rapid growth are the typical features of this carcinoma. The metastasis is also at a faster rate when compared to classic Squamous cell carcinoma. The cervical lymphnodes are invariably involved and may appear as indurated and immobile⁵.

Radiographic appearance of Spindle cell carcinoma is not pathognomic. Like any invasive malignant tumour of the bone it shows destruction of bone according to the severity of involvement. OPG may reveal characteristic irregular destruction of bone causing sometimes thinning of the lower border of the mandible, when mandible is involved^{1,6}.

In the past Spindle cell carcinoma has been thought to be a collision tumour between carcinoma and sarcoma. Virchow was the first person to introduce the term carcinosarcoma for these groups of lesions⁷.

In the beginning of 20th century, Kettle E H in his article cited that many authors were of the opinion that the spindle cell component is actually epithelial cells being transformed into mesenchymal cells⁸.

Previous studies using electron microscope had presented conflicting reports as to the histogenesis and pathogenesis of these lesions. According to some authors the spindle cells consisted of non-neoplastic histiocytic and fibroblastic cells⁹. Others were of the opinion that spindle cells were malignant fibroblasts¹⁰. However a new school of thought became more acceptable and it suggested that the spindle cells though appeared to be originating from the mesenchyme, had nothing to do with mesenchymal elements, rather it was purely epithelial in origin¹¹.

Research using electron microscope and immunohistochemistry have put forward an amicable solution to the debate and currently most investigators conclude that lesional cells are epithelial in origin with ability to produce mesenchymal intermediate filaments^{5,6}.

Spindle cell carcinoma can demonstrate varied histopathological appearance from case to case or within different areas of the same tumorous tissue. The cells may appear like epithelial cells or may appear as atypical mesenchymal cells to add to the confusion. A histopathologist using a light microscope may find it extremely difficult to assess the case as it may appear as a carcinoma as well as a sarcoma. The tumour generally consists of fascicles of anaplastic spindle cells with considerable number of mitotic figures⁴. The histopathologist often has an impression of an anaplastic fibrosarcoma and is more likely to report it as the same unless he or she probes into the details of the sections

where inconspicuous squamous elements are detected. Often the cells assume whorl, sheet or rosette pattern which may be observed concurrently in sections.

The squamous component can be analysed by looking into the overlying surface epithelium where the dysplastic squamous cells that may retain the original shape can be detected along with the spindle cells. In some cases direct transition between the two cell types are evident. At light microscopic level the best way to come to a conclusion is to examine serial sections.

It has been reported that to add to the confusion about this controversial lesion, on rare occasions, bone & cartilage or muscle differentiation could be evident. What is more perplexing is the fact that overall histopathologic picture is similar to that of an anaplastic fibrosarcoma⁴.

The lesions from metastatic sites may reveal only spindle cells, only squamous cells or a combination of the above two. Invariably the malignant cells show prominent dysplastic features. This characteristically includes prominent pleomorphism of the cells and the nuclei, characteristic elongation and hyperchromatism of the nuclei and abnormal number of mitotic figures. Occasional giant cells may be also seen.

Immunohistochemical techniques may reveal the difference between Spindle cell carcinomas and mesenchymal malignant neoplasms. The lesional cells of most mesenchymal tumours produce vimentin and not cytokeratin. It is noted that more than two thirds of cases of Spindle cell carcinoma react with antibodies directed against cytokeratin. Yasusei Kudo et al in their study reported that spindle cells expressed wnt-5a and vimentin mRNA at high levels but did not express E-cadherin mRNA. Their study proved that the spindle cell may behave similar to mesenchymal cells. However positivity for cytokeratins showed epithelial nature of the spindle cells¹². According to Ulrich Volver H et al improved survival rate in patients with Squamous cell carcinoma is associated with a low level of cytokeratin expression¹³. In the present case the inconspicuous squamous element detected in serial sections were sufficient to diagnose the lesion.

The treatment in case of Spindle cell carcinoma has been much debated. Many authors are of the opinion that wide excision which includes a radical dissection to get rid of the metastatic residues to be the best mode of treatment^{1,3,5}.

Some authors are of the opinion that surgery alone is insufficient in that a radiotherapeutic intervention following surgery is mandatory to avoid possible recurrences¹. Many authors are of the opinion that radiotherapy and

chemotherapy are ineffective in Spindle cell carcinoma³. Unfortunately the five year disease free survival rate is approximately 30% for all oral cases with most cases ending up fatally within one year¹. This is similar to the prognosis of high grade Squamous cell carcinoma. Superficial tumours are said to demonstrate better prognosis irrespective of the tumour size. The presence of metastasis signals a poor prognosis since 81% of patients recorded with metastasis in studies conducted by Ellis and Corio died of the disease¹.

Summary and conclusion

Spindle cell carcinoma is a neoplasm of epithelial origin and has been called also as polypoid squamous tumour or carcinosarcoma. It is considered to be a variant of Squamous cell carcinoma¹⁰.

The interesting feature of this dimorphic neoplasm lies in the fact that it mimics other connective tissue sarcomas and spindle cell malignancies at light microscopic level. In the past the nature of spindle cells and their histogenesis has been strongly debated. Immunohistochemical and electron microscopic studies have contributed drastically to solve this issue^{4,7}.

Spindle cell carcinoma has poor prognosis with metastasis to the distant sites being considered as a reliable prognostic indicator. There is some evidence that microscopic depth of invasion is a strong prognostic indicator in oral lesions. It must be emphasized that Oral Spindle cell carcinoma most often is indistinguishable from connective tissue sarcomas or other spindle cell malignancies at the level of light microscopy.

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