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CASE REPORT

Sarcomatoid Carcinoma: "A Surgeons Nightmare"

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Abstract Sarcomatoid carcinoma (SC) is a rare and unique disease of the oral cavity. It is a biphasic squamous cell carcinoma with sarcoma-like characteristics. It is a strikingly aggressive lesion with a rapid rate of growth and a high rate of metastasis. Diagnosing a sarcomatoid carcinoma may present a challenge; most can only be ascertained by immunohistochemical study. An aggressive treatment plan should be devised for this uncompromising disease with a readiness to accept the dire outcome. We hereby present a case of a 50-year-old male patient, with no history of deleterious habit, diagnosed with SC of the buccal mucosa that proved to be fatal.

Keywords Spindle cell carcinoma · Rare tumour · Collision tumour · Biphasic · Pseudocarcinoma · Reconstruction local flaps

Introduction

Spindle cell carcinoma (SC) is a rare, unusual variant of squamous cell carcinoma that has a biphasic characteristic. It accounts for 3% of all head and neck tumours of epithelial origin. It is a poorly differentiated tumour, with cells proliferating as a squamous carcinoma along with spindle-like cell growth, which are of epithelial origin. The exact origin of the tumour is of a certain controversy amongst the authors. This is exhibited in the numerous names given to the lesion viz, sarcomatoid carcinoma, fusiform cell carcinoma, carcinosarcoma, pleomorphic carcinoma, pseudosarcoma, collision tumour, Lane tumour [1, 3]. SC affects mostly the larynx, the nasal cavity, hypopharynx, oesophagus, trachea, breast, oral mucosa and lungs. In lungs, a spindle cell carcinoma is rare, mostly associated with smokers and has a poor prognosis [4]. WHO coined the term as spindle cell carcinoma. It has a male predilection in a ratio of 7:1, seen mostly in the 6th to 8th decade of life [2]. Deleterious habits like smoking, alcohol consumption and tobacco chewing or any history of previous irradiation multiplies the chance of occurrence of the disease [1].

There are no definite clinical signs that can direct towards the diagnosis of SC although some telltale signs may be present. Diagnosis is mainly histopathological which is confirmed by immunohistochemistry. Surgery is the primary treatment option; adjuvant chemotherapy and radiotherapy may be required.

Owing to its aggressive nature and high chances of distant metastasis, the prognosis is not favourable [3]. We hereby present a case of spindle cell carcinoma of the right buccal mucosa.

Case Report

A 50-year-old male patient reported with a chief complaint of swelling on the right buccal mucosa since a month. Initially swelling was of the size of a peanut which

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gradually progressed to the size of a tennis ball within a span of 15 days. No significant medical, dental, family, habit history was reported. Extra oral examination revealed a solitary well circumscribed, globular swelling measuring 4 cm × 3 cm, in the right side middle third of the face extending supero-inferiorly from the line joining from right ala of the nose till the tragus of right ear; antero-posteriorly from the right angle of the mouth till the posterior border of ramus (Fig. 1). Skin over the swelling appeared tensed. On palpation, the swelling was indurated, firm, non-tender, non-fluctuant, non-pulsatile, non-reducible, and moved with the surrounding soft tissue but was not fixed to the bone. Right submandubular lymph nodes were palpable. Intra-oral examination revealed a well-defined, exophytic lesion in the right buccal mucosa extending antero-posteriorly from premolar till the last molar and supero-inferiorly from the upper dentition extending slightly into the mandibular vestibule, measuring about 3 cm × 2 cm in size. Surface appeared mildly keratinised with sloughing, was firm in consistency and non-tender. Oral hygiene was poor; teeth were periodontally compromised. Mouth opening was normal; swelling did not interfere in occlusion (Fig. 1).

Ultrasonographic examination of swelling revealed vascularity in the deeper planes but was essentially inconclusive (Fig. 2). Hence MRI of head and neck was done (Fig. 3), which revealed a well-defined, lobulated mass, iso–hyperintense to muscle on T1 and hyperintense on T2, with gross enhancement post-contrast, involving the right gingivo-buccal sulcus and myo-fascial plane involving masseter, buccinator and subcutaneous area, abutting the right posterior maxillary alveolus and minimal scalloping of the mandibular alveolus. Also right submandibular lymph node enlargement was noted. CT thorax and chest radiographs appeared non-pathological, and blood investigations including prostate-specific antigen were within normal limit. Patient's attenders were not willing for PET–CT due to financial constraints. Incisional biopsy of the swelling was done, but report was inconclusive. Following the biopsy, swelling started to shrink in size, but the lesion appeared essentially normal. Rebiopsy was done after 1 week, by acquiring deeper tissues. IHC

Fig. 1 One and a half week postop rebiopsy showing exponential increase in size

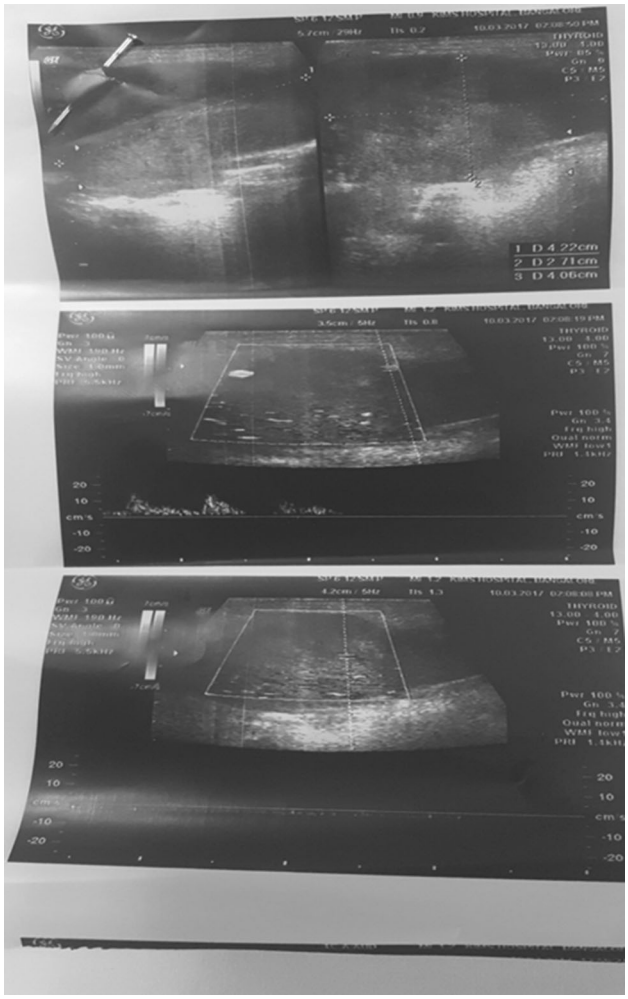


Fig. 2 USG scans

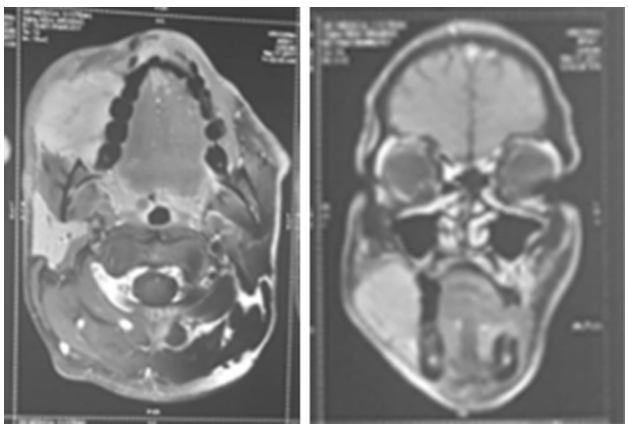


Fig. 3 MRI showing hyperintense lesion in right Buccal mucosa

reports showed the lesion to be strongly expressive for cytokeratin and vimentin suggestive of spindle cell carcinoma (Fig. 4). One and a half week following the rebiopsy, the swelling increased exponentially (Fig. 1), and hence a call was taken to operate at the earliest.

Wide excision of the lesion on the right side with hemimandibulectomy, posterior maxillary alveolectomy, and modified neck dissection-I were performed (Fig. 5). Sample was sent for frozen section biopsy, and margins were revised accordingly and were found to be tumour free. For intra-oral reconstruction Pectoralis Major Myocutaneous (PMMC) flap in combination with deltopectoral (DP) tube flap to cover the extra oral skin defect was used (Fig. 6). Partial thickness skin graft from the left lateral thigh was raised and meshed to cover the defect created by the DP flap. Complete closure was achieved, and patient was extubated uneventfully (Fig. 7). DP flap started to progressively discolour within 5 days of the surgery, and wound dehiscence was noted on the superior aspect of the flap. Margins of the flap were revised, and reclosure was done. Overall muscle wasting and weight loss were noted. Patient was discharged after 2 weeks. General condition of patient was apparently fine. The next day following discharge, patient reported to the emergency department with complaint of 1 episode of vomiting, giddiness and weakness. Patient was admitted, and intravenous fluids were started. Patient was cachexic, tachypnoeic, delirious and was unable to maintain oxygen saturation on his own. Chest X-ray was normal. Patient died within 5 days owing to respiratory failure and cardiac arrest.

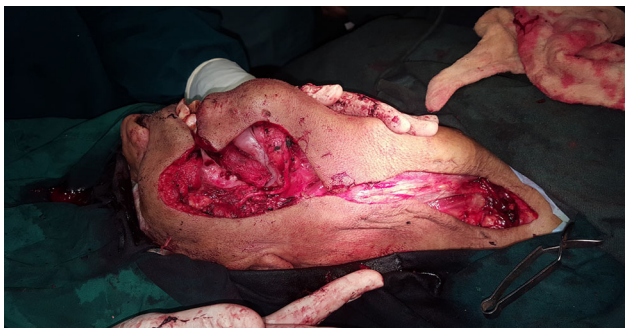


Fig. 5 Wide excision with right hemimandibulectomy and right maxillary alveolectomy with MRND type II

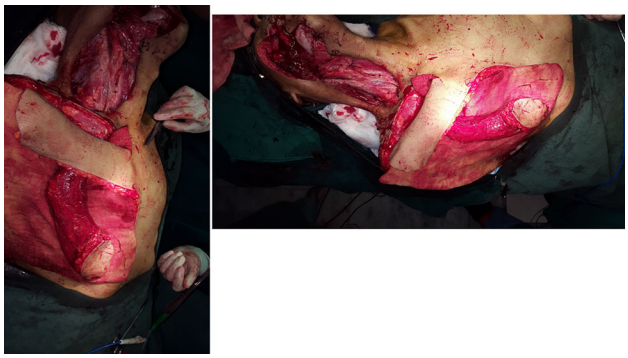


Fig. 6 PMMC flap and DP dissected and raised for reconstruction

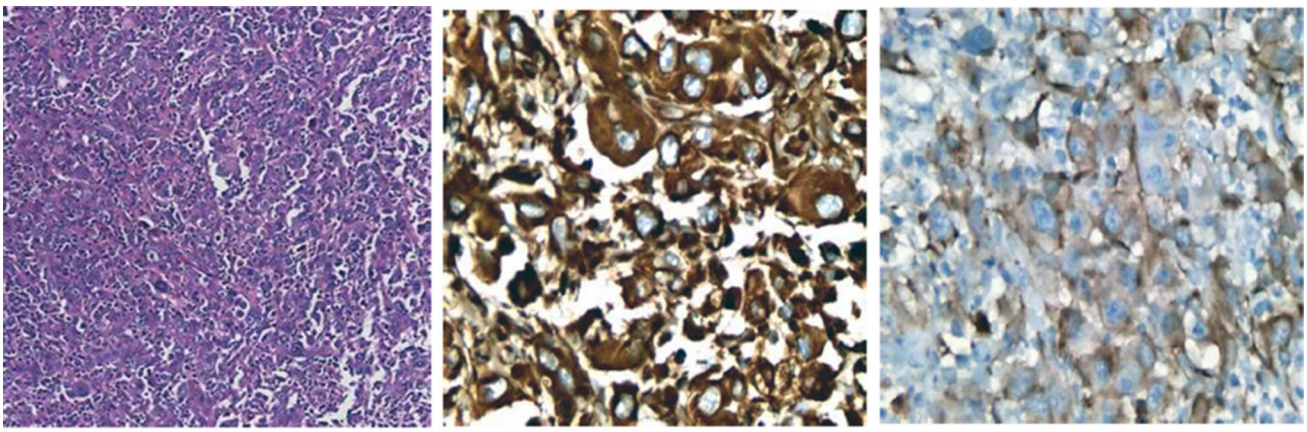


Fig. 4 Histopathology and immunohistochemistry report

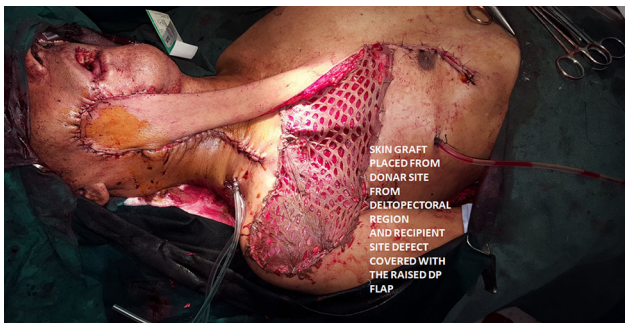


Fig. 7 Immediate postop

Discussion

Spindle cell carcinoma (SC) is an atypical, aggressive variant of squamous cell carcinoma (SCC). It is a relatively primitive form of SCC that is derived from both epithelial and mesenchymal origin [1]. About 1 per cent of oropharyngeal carcinomas are SC [2]. It affects predominantly in 5th to 7th decade of life and has a male predilection [1, 3]. Although Kesseler et al. has reported SC in a 4-year-old male, albeit it is very rare in childhood. SC is seen mostly in the larynx and is infrequently seen in the oral cavity and the oro-pharynx. Gingiva, tongue, buccal mucosa SC has also been reported. Pharynx, nose, upper gastrointestinal tract, lungs, breast are the other reported sites [3]. Vishwanathan et al. studied 103 cases of SC retrospectively found that oral cavity was the most affected part (63.1%) [7]. Aetiology is largely unknown; possible risk factors include radiation exposure, tobacco, alcohol abuse, poor oral hygiene [3].

SC typically presents with an outward growing, expansile, irregular or polyp-like mass. Surface of the lesion may show ulceration, emitting foul odour due to necrotic changes. Difficulty in mouth opening and change in voice may be seen in oral cavity and laryngeal tumours, respectively [5]. Histologically, the mesenchymal component may be overpowered by the squamous element, but one cannot diagnose based upon this ratio as even complete vacancy of spindle component may turn out to be a SC. Hence SC presents a major diagnostic doldrums to the pathologist and the surgeon as well. Due to absence of expression of adhesion molecules (e.g. cadherins) and change in keratin microfilaments, spindle-shaped tumour cells are noted [3]. Generally histopathological reports are non-conclusive and may resemble conventional SCC with atypical cells, and therefore, confirmation is done using immunohistochemical studies [1]. The lesion is strongly positivity for vimentin and cytokeratin [3].

Pathogenesis of the disease is classified in three categories. Firstly, the spindle and epithelial elements occurs concurrently albeit being derived from different stem cells and were termed "collision tumour." Secondly, spindle component with bizarre proliferation of the stroma, hence called "pseudocarcinoma" [3]. Lastly, the same monoclonal antibodies were thought to be the predecessor of both the components [3, 5]. Differential diagnosis of SC includes the likes of squamous cell carcinoma, fibrosarcoma, fibrous histiocytoma, rhabdomyosarcoma, synovial myosarcoma and peripheral malignant nerve sheath tumour [3].

Surgical excision is considered as the gold standard of treatment of SC [6]. Radiotherapy can be used as adjuvant, although contradictory reports are found in literature. Ballo et al. in their study, where laryngeal SC was treated by radiotherapy alone, found a recurrence rate of only 14.3% in a 10-year follow-up period [7]. Dubai et al. in their study on laryngeal SC reported a 5-year survival rate after treating SC with wide excision and radiotherapy to be 84.2% and that of only surgery being 84.1%. This difference is statistically insignificant. He also showed that treatment without any surgery resulted in a 5-year survival rate of 57.1%, a much poorer percentage. They also inferred that there is no significant impact on the survival rate by using radiotherapy. Radiotherapy as a single treatment modality also presented with a lower percentage of 5-year survival rate (60.5%) and was used in cases of inoperable stage of the disease [8]. Thompson et al. concluded that surgery as the single modality of treatment offers a better chance of survival in laryngeal SC than patients treated with combination of surgery and radiation [9].

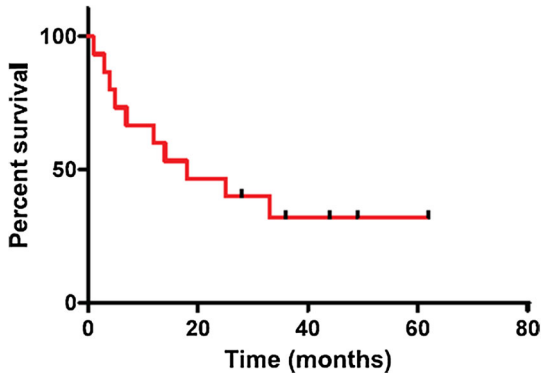
Benniger et al. and Su et al. in two retrospective studies concluded that oral cavity SC are more aggressive than others [10, 11]. Aggressively treated SC had a lesser 5-year survival rate as compared to conventional Squamous cell carcinoma (SCC) (SC—32%, SCC 45%). Due to this aggressive nature, combination of surgery and radiotherapy was employed in treating 63.6% of SC cases as compared to 40.4% in conventional SCC [12]. Although many SC are unresectable, palliative therapy becomes the treatment of choice.

As per Vishwanathan et al., metastasis to regional lymph node is seen in 21.5% of oral cavity lesions, 33.3% in oro/hypo pharyngeal lesion and in only 1% of laryngeal lesions [5], although the rate of metastasis is varied in the literature ranging from 7.5 to 26% for head and neck SC. Distant metastasis was seen in lungs and soft tissue in 5% cases [13]. It is prudent for the surgeon to perform a radical dissection of the involved lymphovascular tissues for better prognosis of the disease. Local recurrence can be seen in 16 to 32% of times [5, 14]. Newer trends in treatment of SCC includes anti-EGRF (epidermal growth factor receptor) therapy, but has limited applications in treatment of SC [15].

Conclusion

To summarise, successfully diagnosing of SC is a surgeon's dilemma but a pathologist's dream. Oral and maxillofacial surgeons must be aware of the mortality that comes hand in hand with this aggressive, unrelenting disease and should be treated with a well-planned, aggressive surgical excision. Radiotherapy should be used in cases that are advanced or in cases of unclear disease margins following surgery. Even after adequate surgical clearance and chemoradiotherapy, the death toll of the disease is much, and thus it is fortunate that spindle cell carcinoma of the head and neck is a rare occurrence. Thus a thorough clinical, histomorphological acumen of the disease can help the doctor treat it in a systematic approach.

Overall survival probabilities were estimated using Kaplan–Meier curves [2]



Compliance with Ethical Standards

Conflict of interest No conflict of interest declared between any authors. No financial support/grant obtained from any source. Patient's and relatives' consent for publication has been taken.

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