Spindle Cell Carcinoma of Oral Cavity: A Rare Entity
Ng CS¹, Mohd Razif M Y², Chew MX³, Suria HMP⁴

Abstract
Introduction: Spindle cell carcinomas (SpCC) are a rare variant of squamous cell carcinoma which is rarely encountered in the head and neck. It is also known as “pseudocarcinoma”, “sarcomatoidcarcinoma,” and “carcinosarcoma”. It has an aggressive nature and has poor prognosis despite aggressive treatment. Case report: A 48-year-old lady presented to us with a right painless upper gum swelling which was progressively enlarging for 3 months after tooth extraction. Examination of the oral a fungating mass over the right upper alveolus extending to the right upper gingivolabial sulcus. She was subjected to a debulking palliative to reduce the tumour bulk. The intraoperative samples sent for histopathological examination was reported as SpCC, AJCC (8th edition) stage pT2pN3b. She recovered well after the surgery with quality of life after the surgery.

Keywords: Spindle Cell Carcinoma; pseudocarcinoma; sarcomatoid carcinoma; carcinosarcoma

Introduction
Spindle cell carcinomas (SpCC) of the head and neck is a rare variant of squamous cell carcinoma that only represents less than 3% of all head and neck malignancies of epithelial origin. It is an aggressive tumour with poor prognosis and has a predilection formales. It is often associated with smoking, alcohol consumption, and previous radiotherapy to the head and neck.

Case report
A 48-year-old lady with no previous known medical problems presented to us with a right painless upper gum swelling for 3 months after tooth extraction. The swelling progressively increased in size and is associated with bleeding. Clinical examination of the oral cavity revealed a fungating mass over the right upper alveolus, measured 2 x 4cm in size. The mass extended to the right upper gingivolabial sulcus, crossed the midline into the left gingivolabial sulcus. There were no other oral lesions noted and examination of the neck showed no evidence of neck metastasis. Endoscopic assessment of the nose and throat were also normal. Biopsy taken from the mass showed atypical spindle cell lesion. Differential diagnosis was spindle cell variant of squamous cell carcinoma, SpCC and spindle cell sarcoma. A computed tomography (CT) scan from the base of skull to neck done after the biopsy showed an enhancing mass at midline of maxilla measured at 3.3x1.7x2.8cm, with erosions of the underlying bony maxilla. There was no extension into the bilateral maxillary sinuses, nasal or oral cavities. There were multiple enlarged upper cervical lymph node and submandibular lymph node with the largest diameter measured at 1.8cm. There was also an enlarged right supraclavicular lymph node measured at 3cm and a lytic lesion seen at C6 body. The impression given after the CT scan was carcinoma of themaxilla with secondaries to bilateral cervical, submandibular and right supraclavicular lymph node and C6 vertebrae. A positron emission tomography (PET) scan done later showed a hypermetabolic right supralabial lesion

1. Ng CS
2. Mohd Razif M Y
   Department of Otorhinolaryngology, National University of Malaysia, Kuala Lumpur, Malaysia.
3. Chew MX
4. Suria HMP
   Department of Pathology, National University of Malaysia, Kuala Lumpur, Malaysia

Correspondence to: Professor Dr. Mohd Razif Mohamad Yunus, Professor in Department of Otorhinolaryngology, Head and Neck Surgery in National University of Malaysia. Email: rafiz72@gmail.com
with invasion into right alveolar process with multiple enlarged hypermetabolic cervical lymph node, lung nodules and lytic skeletal lesion. The report stated that it was a stage IV disease, with regional nodal, lung and skeletal metastases. A diagnosis of right alveolar carcinoma TNM Stage T3N2bM1 with lung and skeletal metastasis was made.

Debulking palliative surgery was done with the consent of the patient and aims to reduce the tumour bulk for better palliative care. Right anterior inferior maxillectomy was subsequently performed with removal of the fungating mass at the right maxillary alveolus which involved the gingivobulbary sulcus and philtrum (*Figure 1*), measured at 2x4cm (*Figure 2*), followed by right modified radical neck dissection Type I, left selective neck dissection (Level I-IV) and right submandibulectomy. The intraoperative samples sent for histopathological examination was consistent with SpCC, AJCC (8th edition) stage pT2pN3b. The tumour is cellular, composed of predominantly spindle cells arranged in haphazard, storiform growth pattern and exhibits mild to moderate pleomorphic vesicular nucleus with conspicuous nucleolus (*Figure 3a*).

**Figure 1.** Intraoperative figure showing a fungating mass (arrow) at the right maxillary alveolus involving the gingivobulbary sulcus and philtrum.

**Figure 2.** Resected tumour measured at 2x4cm.

**Figure 3a.** Microscopic image (X40) of spindle cells arranged in halphazard, storiform growth pattern and exhibits mild to moderate pleomorph vesicular nucleus with conspicuous nucleolus.

There is perineural invasion (*Figure 3b*) and both the left medial and anterior margins of the specimen was involved by the tumour. The lymph nodes from left level Ia, right level Ib, II, III, IV are also involved by the tumour. She recovered well after the surgery where a temporary obturator was inserted by the oral maxillofacial surgeon 6 days later. She claimed that she had a drastic improvement in quality of life after the surgery as she can eat orally. She was last seen 3 months after the surgery. During all the subsequent follow ups she exhibited no signs of locoregional recurrence. However, she developed jaundice and a repeated PET scan showed distant metastasis to her liver. Her care has since been taken over by palliative care.
Figure 3b. Microscopic image showing perineural invasion of tumour cells.

**Discussion**

SpCC is a rare form of malignancy in the head and neck region and it is rarely encountered. The challenges in managing a patient with SpCC are mainly due to rarity, difficulty of histological confirmation, and a shortage of literature about disease pathogenesis, outcome survey, and management guidelines. However, surgery seems to be the first option for primary SpCC, where complete resection of the tumour with larger safety border is suggested followed by adjuvant radiotherapy or chemotherapy. Histologically, SpCC is characterized by spindled tumour cells that simulate a sarcoma but are epithelial in nature. Therefore, it is always misdiagnosed in the olden times because of unfamiliarity. Within the oral cavity, SpCC frequently involve sites such as the tongue, gingiva/alveolar mucosa, buccal mucosa, floor of the mouth, and lip. The diagnosis of SpCC requires the identification of a component of squamous neoplasia or epithelial differentiation of the spindle cells. Immunohistochemical stains may help to differentiate SpCC from another lesion but may not be specific or sensitive.

Our patient who presented to us with SpCC of the dentoalveolar region with extensive distant metastasis possessed a challenge to us in terms of our aim of management, which is to relieve the patient from further discomfort due to the aggressive nature of the tumour. In our case, a palliative resection of the tumour with nodal clearance from the neck was done followed by chemotherapy later to reduce the tumour load for improved quality of life. This is to ensure a better palliative care in future which the patient has been thoroughly explained and has agreed to. The outcome of the surgery is good, and the patient can enjoy an improvement in her quality of life after tumour load reduction.

**References**