CASE REPORT

Spindle cell carcinoma in maxilla

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SUMMARY

Spindle cell carcinomas (sarcomatoid carcinomas) are rare tumours. It is a variant of squamous cell carcinoma which has spindled tumour cells, which simulate a true sarcoma, but are epithelial in origin. They are extremely uncommon in the head and neck region. Only five cases with maxillary origin have been discussed in the literature. As compared to squamous cell carcinoma of maxilla, this variant is associated with poor diagnosis and advanced disease at presentation, as is demonstrated in the case presented. There are no standard recommendations for management owing to the rarity of this histology. Surgery and radiotherapy form the mainstays of treatment. We report a rare case of spindle cell carcinoma involving the maxilla.

BACKGROUND

Spindle cell carcinoma is an unusual form of poorly differentiated squamous cell carcinoma consisting of elongated epithelial cells that resemble a sarcoma.¹ WHO named it 'Spindle Cell Carcinoma' under the malignant epithelial tumours of squamous cell carcinoma (SCC).

Literature describes that a variety of terms are used to describe spindle cell carcinoma which include carcinosarcoma, pseudosarcoma, sarcomatoid squamous cell carcinoma, pleomorphic carcinoma and polypoid carcinoma.²

Spindle cell carcinoma is a biphasic tumour composed of a SCC, either in situ and/or invasive and a malignant spindle cell component with a mesenchymal appearance, but of epithelial origin.³ Spindle cell carcinoma is an epithelial neoplasm, with a divergent differentiation, and not a collision tumour, biphasic derivation or pseudosarcoma.⁴

This tumour has specific clinical features and behaviour, and the establishment of the correct diagnosis has important implications for management. Only a few cases have been reported in the medical literature. This case report describes its immunohistochemical features and a brief review of the literature.

CASE PRESENTATION

A 52-year-old woman presented to the department of the Oral and Maxillofacial Surgery with a 1 month history symptom of mobility of upper anterior teeth. Interestingly, she gave a history 2 years earlier of buccal space infection for which she underwent incision and drainage and extraction of the posterior teeth on that side due to mobility.

Her medical history was unremarkable and there was no history of sinus-related disease or trauma to the maxillofacial complex. A history of mobility of tooth was significant as it started from the posterior

region of left maxilla and gradually affected the anterior teeth. Also, a history of numbness of the upper lip since 2 months with no history of extra-oral swelling was significant. An intraoral examination revealed a proliferative growth of about 3.5×1 cm in the left upper molar region with no signs of ulceration (figures 1 and 2). On palpation, the palate was soft in consistency with no underlying bone from left molar to anterior region crossing the midline. Nasal examination showed no growth. No neck nodes were palpable and the cranial examination was normal.

INVESTIGATIONS

Orthopantomogram revealed an osteolytic lesion extending from the left maxillary tuberosity area to the right lateral incisor (figure 3).

A CT scan revealed soft tissue density lesion in the left upper alveolar region with erosion of the floor and medial wall of the left maxillary sinus, left side of hard palate and left alveolar process. The lesion also shows erosion of the medial and lateral pterygoid plates. Multiple left-sided cervical lymphadenopathies were also seen (figure 4).

An incisional biopsy revealed a hyperkeratinised, stratified squamous-type epithelium with long rete-ridges. The underlying connective tissue consisting of tumour mass was in close proximity to the

Figure 1 Facial profile of patient showing no obvious changes.

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Figure 2 Intraoral photograph showing loss of posterior teeth and proliferative growth on the right maxillary alveolar region.

surface epithelium. The tumour mass appears to be fibrocellular with large amount of spindle-shaped cells in a fascicular pattern and storiform pattern in few areas, also noted areas with large oval cells. These spindle-shaped cells show plump nucleus as well as thin wavy nucleus with increased mitotic activity as well as pleomorphism, also consist of plasmacytoid and inflammatory cells suggestive of a spindle cell lesion (figures 5 and 6).

Immunohistochemistry study revealed creatine kinase and vimentin: strongly positive for tumour cells, S-100: focally positive in tumour cells, epithelial membrane protein (EMA), human melanoma black 45 (HMB45) and smooth muscle actin (SMA): negative for tumour cells. Impression was of features suggestive of spindle cell carcinoma with coexpression of cytokinin and vimentin (figures 7–9).

DIFFERENTIAL DIAGNOSIS

- ▶ Intraosseus carcinoma
- ▶ Chondrosarcoma
- ▶ Myofibroblastic sarcoma
- ▶ Squamous cell carcinoma
- ▶ Fibrosarcoma
- ▶ Malignant fibrous histiocytoma
- ▶ Leiomyosarcoma
- ▶ Rhabdomyosarcoma
- ▶ Malignant peripheral nerve sheath tumour
- ▶ Osteosarcoma
- ▶ Mesenchymal chondrosarcoma
- ▶ Kaposi's sarcoma
- ▶ Angiosarcoma
- ▶ Synovial sarcoma

Figure 3 Orthopantomogram showing loss of bony architecture on the right maxillary region and floating teeth appearance of the right maxillary anterior teeth.

Figure 4 CT scan showing loss of bony architecture on the right side of the maxilla.

- ▶ Malignant melanoma
- ▶ Fibromatosis
- ▶ Leiomyoma
- ▶ Nodular fasciitis
- ▶ Reactive epithelial proliferations

TREATMENT

No treatment was performed as the patient failed to report for admission and further treatment.

DISCUSSION

Sarcomatoid carcinoma is a type of malignant mixed tumours and is extremely rare in the maxilla. The initial description of this type of malignancy was reported in 1864 by Virchow, who labelled it as carcinosarcoma.⁵ Later on, Saphir and Vass analysed 153 cases of carcinosarcomas of various sites. They concluded that the sarcomatous component represented a variation

Figure 5 Photomicrograph showing tumour mass appearing fibrocellular with large amount of spindle-shaped cells in a fascicular pattern and storiform pattern in few areas, with areas showing large oval cells.

Figure 6 Photomicrograph showing tumour mass appearing to be fibrocellular with large amount of spindle-shaped cells in a fascicular pattern and storiform pattern in few areas, with areas showing large oval cells.

in the squamous portion of the carcinoma or an inflammatory reaction of the underlying stroma.⁵ Numerous hypotheses regarding the histogenesis of this type of tumour have been proposed. Three dominant pathogenetic theories have been proposed: the tumour (1) represents a 'collision tumour' (carcinosarcoma), (2) is a squamous cell carcinoma with an atypical reactive stroma (pseudosarcoma) or (3) is of epithelial origin, with 'de-differentiation' or transformation to a spindle cell morphology (sarcomatoid carcinoma).⁶ Recently, the third hypothesis has been supported by the following evidences: their occurrence in the exact sites that normally have squamous epithelium and a preponderance of carcinomas rather than sarcomas; a superficial location; a polypoid appearance; the direct continuity and smooth transition of the spindle cells with areas of squamous epithelium; immunoreactivity with epithelial antigens; a dual expression of epithelial and mesenchymal differentiation with double labelling techniques in some neoplastic spindle cells; and the presence of epithelial only, sarcomatous only or a duality of expression in metastatic deposits from laryngeal sarcomatoid carcinoma.⁷ Furthermore, recent molecular studies have shown evidence of a monoclonal origin from a stem cell capable of divergent differentiation.⁸⁻⁹ When the malignant surface epithelium is histologically evident, the

Figure 8 Immunohistochemistry pictures confirming diagnosis of spindle cell carcinoma.

diagnosis of a sarcomatoid carcinoma is made with confidence. However, when the surface epithelium is ulcerated or denuded, the correct diagnosis is more difficult. Furthermore, the biopsies from the squamous cell component tend to be misdiagnosed as squamous cell carcinoma and biopsies from spindle cell component tend to be diagnosed as sarcoma. In our case, the lesion looked like proliferative lesion on initial clinical examination.

Sarcomatoid carcinoma of the oral cavity presents a male predominance at a mean age of 57 years and site predilection for the lower lip, tongue and alveolar ridge or gingiva although most tumours in head and neck region occur in the larynx.²⁻⁴ Growth configuration is often exophytic polypoid, but sessile, nodular or endophytic configuration has also been described. The lesion usually has an extensive surface ulceration with friable, fibrinoid necrosis of variable thickness or shaggy exudates.⁷⁻¹⁰ Radiation, trauma, tobacco use or alcohol consumption seemed to play a role in aetiological factors.¹⁰ The differential diagnosis includes a number of benign and malignant tumours, such as squamous cell carcinoma, fibrosarcoma, malignant fibrous histiocytoma, leiomyosarcoma, rhabdomyosarcoma, malignant peripheral nerve sheath tumour, osteosarcoma, mesenchymal chondrosarcoma, Kaposi's sarcoma, angiosarcoma, synovial sarcoma, malignant melanoma, fibromatosis, leiomyoma, nodular fasciitis and reactive epithelial proliferations.⁹ Wide surgical excision is the treatment of choice. Most authors agree that irradiation is ineffective.

Figure 7 Immunohistochemistry (IHC) pictures confirming diagnosis of spindle cell carcinoma.

Figure 9 Immunohistochemistry pictures confirming diagnosis of spindle cell carcinoma.

Learning points

- ▶ Adjuvant irradiation might be of benefit in cases where the surgical margins are positive or in patients with nodal metastasis at the time of diagnosis.
- ▶ Prognosis is related to location, tumour size, depth of invasion, stage of disease and with the presence of any keratin staining in the spindle cells.¹⁰
- ▶ A tumour of the oral cavity and oropharynx is potentially aggressive and seems to recur and metastasize easily.
- ▶ The incidence of metastases was 36% and the 2-year survival rate was 55% in tumours involving the oral cavity.
- ▶ Radiation therapy is considered an acceptable alternative for inoperable patients.

Contributors SS, SSV diagnosed the patient and performed the various procedures on the patient. NH compiled the manuscript and prepared the initial draft. PPN edited and supervised the submission.

Competing interests None.

Patient consent Obtained.

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