

Biphasic Synovial Sarcoma of Oral Cavity

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ABSTRACT

Synovial sarcoma is a mesenchymal spindle cell tumour, which is unrelated to synovium and shows variable epithelial differentiation. Typically, synovial sarcoma arises in the soft tissues of the extremities but cases in the head and neck region are less common and oral cavity involvement is extremely rare. A 17-year-old girl presented with a gradually increasing swelling on the right cheek for 2 years, which on biopsy, revealed a biphasic tumour comprising fascicles of spindle shaped cells with gland formation by epithelial cells and scattered masts cells. Histological diagnosis of biphasic synovial sarcoma was confirmed on immunohistochemistry by strong positivity for EMA, S-100 and CD-99 in both epithelial as well as spindle cell areas.

Key words: *Synovial sarcoma. Oral cavity. Biphasic tumour.*

INTRODUCTION

Synovial Sarcoma (SS) is a mesenchymal spindle cell tumour with variable epithelial differentiation including glandular formation. It is unrelated to synovium as the name suggests but arises in the soft tissues of the extremities around knee, joints and tendon sheaths.¹ Presentation in the head and neck is less common and oral cavity is extremely rare.² The reported case is the first of its kind at our institute.

CASE REPORT

A 17-year-old female patient presented with a gradually increasing swelling on the right cheek of 2 years duration. Initially, it was painless but started to increase in size rapidly for the last one month. On physical examination and CT scan, a 2 x 1.8 x 3 cm swelling was noted in the buccal side protruding into the oral cavity on the lateral aspect of the maxilla and extending to the ramus of the mandible. No ulceration of the overlying skin or mucosa was present (Figure 1). Initially, a small per-oral incisional biopsy was performed, which revealed a spindle cell tumour on histopathology. Subsequent excision was done as the tumour started to increase in size rapidly after initial incisional biopsy. The specimen submitted for histopathology was fragmented, therefore, its margin clearance could not be assessed. Microscopic examination of the tissue revealed a biphasic tumour comprising fascicles of spindle shaped cells with mild pleomorphism. Solid nests of epithelial cells forming glands were seen with focal areas of

keratinisation. Scattered masts cells were also seen in between the spindle cells (Figure 2).

A differential diagnosis of squamous odontogenic tumour, ameloblastoma, spindle cell, squamous cell carcinoma and biphasic synovial sarcoma was made. Since it was a soft tissue lesion without the involvement of bone or overlying mucosa, a diagnosis of biphasic synovial sarcoma was favoured. Immunohistochemistry, using antigen-antibody immunoperoxidase method, showed strong positivity for EMA, S-100 and CD-99 (Figure 3). Focal positivity for CKAE1/AE3 was also present in both epithelial as well as spindle cell areas. Postoperative MRI scan showed residual tumour with enlarged lymph nodes in the submandibular and upper cervical lymph nodes. Neo-adjuvant chemotherapy followed by modified radical neck dissection was planned for subsequent management.

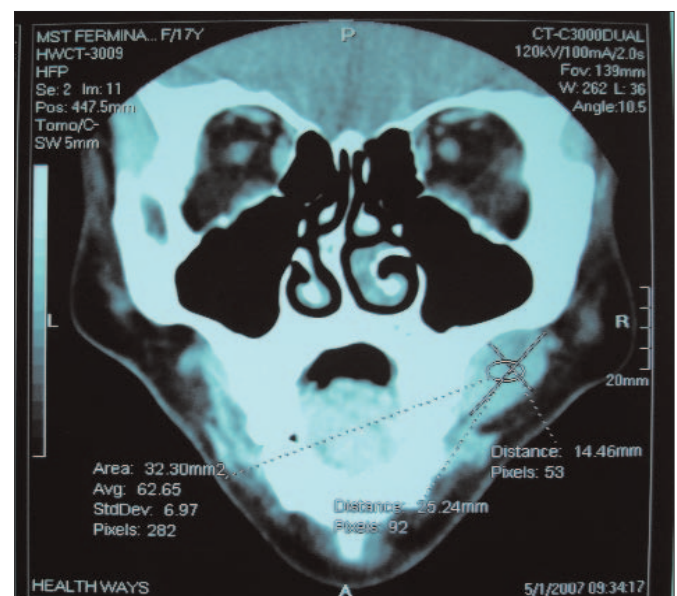


Figure 1: Photograph showing a soft tissue density mass in the right buccal space on the lateral aspect of maxilla extending posterior to the ramus of mandible.

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Received April 15, 2008; accepted October 08, 2008.

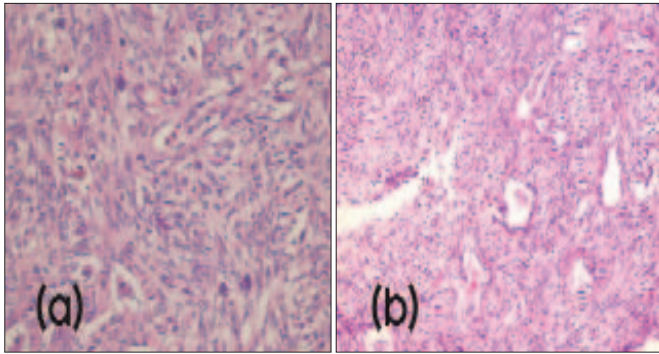


Figure 2: Photomicrographs showing (a) spindle cell component with scattered mast cells. (b) There are interspersed epithelial cells forming glands embedded in the sarcomatous component. (Hematoxylin and Eosin stain at x 400 and x 200 magnification).

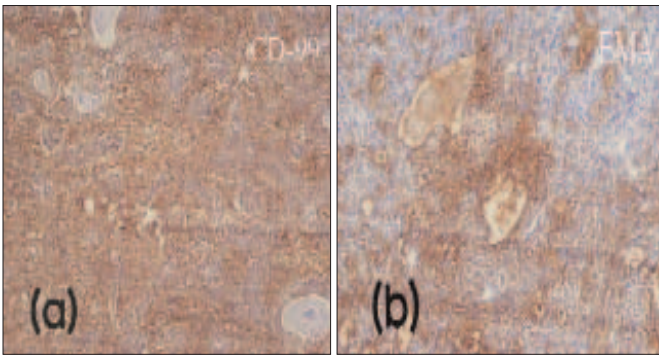


Figure 3: Photomicrographs showing (a) CD-99 positivity in the spindle and epithelial cells and (b) epithelial and a few spindle cells show positivity for EMA. (antigen-antibody immunoperoxidase method at x 200 magnification).

DISCUSSION

Synovial sarcoma is a clinically, morphologically and genetically distinct entity unrelated to synovium. Because of its epithelial features, it has been proposed that it may be renamed as spindle cell carcinoma of the soft tissues.³ It accounts for 5-10% of the soft tissue sarcomas with a wide age at presentation. However, upto 90% cases occur in young adults between the age of 15-35 years with a male predominance.¹ Apart from extremities cases in the retropharyngeal area, anterior abdominal wall, retroperitoneum and mediastinum have also been reported. Even rare are the cases in the oral cavity, lung and prostate, as was seen in this case.⁴

The patients present clinically as a mass with or without pain and local symptoms are present relative to the site. It is a slow growing tumour increasing in size over 2-4 years. Radiologically massive irregular calcifications are seen. Gross examination of the resected specimens show tumours varying in size from 3-10 cm. They are either well-circumscribed or infiltrative and grayish pink on cut surface with multicystic areas.⁵

Histologically, the classical form of SS is a biphasic tumour composed of sharply segregated epithelial and sarcomatous components. Epithelial areas are present in the form of glands lined by cuboidal or columnar cells or as solid nests as was seen in this case. Squamous

differentiation is exceptional and occurs in <1% cases. Sarcomatous component is made up of hypercellular spindle cells with fibroblast-like cells or hemangiopericytoma-like areas. Extensive sampling is required to display epithelial features. Synovial sarcoma with only spindle cells or purely glandular component is called monophasic synovial sarcoma. Calcifying SS is another subtype, which has extensive calcification and carries a much better prognosis.⁶

Histologically, no tumour in the body resembles biphasic SS. However, monophasic SS simulates fibrosarcoma, malignant peripheral nerve sheath tumour and hemangiopericytoma. Reticulin stain highlights the imperceptible clusters of epithelial cells. Immunohistochemically S-100, CD-99, bcl-2 and vimentin positivity is seen in the spindle cells of SS. Cytokeratin expression is seen in 90% of all SS both in the epithelial and a few cells of the sarcomatous component. Specific expression of cytokeratin subtypes 7 and 19 along with EMA is observed, and expression of epithelial markers in the epithelial as well as sarcomatous element excludes the possibility of Ewing's sarcoma and MPNST, which may also show positivity for CD-99 and S-100 on immunohistochemistry in the spindle cell areas.⁷ Ultrastructurally, true glandular epithelial features are seen in epithelial areas. Spindle cell areas also show subtle epithelial features. The t (x; 13) (p11; q11) is the cytogenetic hallmark of synovial sarcoma. FISH and RT-PCR have been employed for the rapid diagnosis of SS.⁸

The prognosis in SS is related to the size and margin status. The tumour recurs locally with metastasis to the lung and lymph nodes. An incidence of 10-15% nodal metastasis is higher than seen in other soft tissue sarcomas. The preferred treatment is local excision with wide margins of normal tissue supplemented by radiation therapy.¹ The good prognostic markers include young patient, distal location of tumour and a size < 5cm. Clear surgical margin, mitosis <15/10 HPF and calcification are also favourable indicators. However, necrosis, poorly differentiating areas and aneuploidy are worse prognostic indicators. The overall 5-year survival is 36-76% and reaches upto 80% in the calcifying SS.⁹

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