

The Happenstance of Monophasic Fibrous Synovial Sarcoma in the Oral Cavity – A Rare Case Report Emphasising the Diagnostic Importance with Literature Review.

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ABSTRACT

Background: Synovial sarcoma is a rare malignant mesenchymal neoplasm of unknown aetiology accounting for 1.9%-3.7% in the oral cavity. More commonly it is seen in the soft tissues of lower extremities. It is a rare phenomenon of occurrence in the oral cavity.

Case presentation: A 31-year-old female patient presented with the painful swelling in the left lower back tooth region for two months. The histopathological examination revealed a monophasic fibrous synovial sarcoma. Immunohistochemical findings showed strong positivity for CD99, Bcl2 and negative for CD34 confirmed the diagnosis of synovial sarcoma. Surgical excision with adjuvant radiotherapy is found to improve the overall survival rate of the patient.

Conclusion: Occurrence of synovial sarcoma in the oral cavity is extremely rare. Advanced molecular diagnostic techniques are needed for rapid and definitive diagnosis.

Key words: Synovial sarcoma, Mesenchymal, Monophasic, Molecular.

BACKGROUND

Synovial sarcoma (SS) is a malignant mesenchymal tumor of rare entity¹. Synovial Sarcoma is a misnomer as it is not related to synovium or synovial membrane^{2,3}. They are named as SS for resembling the histological appearance of the synovium^{3,4}. SS is a rare tumor of erratic histogenesis but it is believed to arise from the pluripotential mesenchymal cells⁴. The occurrence of SS in the head and neck region range from 5.6% -10%⁵.

The most common primary sites of synovial sarcoma are the lower extremities, especially the joint capsules, knees, tendon sheaths and bursae⁴. Oral SS accounts for 1.9%-3.7% with high incidences in the hypopharynx, parapharyngeal region and parapharyngeal space⁵. Literature reviews showed that the SS is seen in the third decades with slight male predilection. Occurrence of SS in the oral cavity is an extremely rare condition with only 31 intraoral cases have been reported so far^{1,5}. It is often misdiagnosed because of the unusual location⁵. Here with reporting a case of SS of the retromolar region in a 31-year-old female which was previously diagnosed as solitary fibrous tumors.

CASE REPORT

A 31-year-old female patient presented with the chief complaint of painful swelling in the lower left back tooth region for two months. The pain was of sudden onset, continuous in nature and increased on mastication. The

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patient was moderately built and poorly nourished. All the vital signs are within the physiological limits. Extra orally, facial asymmetry with a swelling on left side of size 2×2 cm. Antero posteriorly, it extends from 3cm behind the corner of the lip to the angle of the mouth. Superiorinferiorly, from the line joining the corner of the lip to the tragus of the ear to the lower border of the mandible. The nasolabial fold is stretched and the patient felt pain while opening and closing the jaw. The overlying skin was smooth with no discharge. The mass was hard, tender and warm on palpation. Intraorally, the swelling extends from 37 to the retromolar region. Surface was smooth with no ulcerations. Complete blood investigations revealed an elevated lymphocytic count and elevated ESR.

OPG examination showed an ill-defined radiolucency in the retromolar region of 3x3 cm in size, involving the inferior alveolar canal. Cone beam computed tomography [CBCT] scan revealed a large osteolytic defect of size 3x3 cm in the 38-region extending till the anterior border of the ramus with perforation of lingual & buccal plates. An incisional biopsy was performed under local anesthesia and submitted for histopathological examination. Haematoxylin and eosin-stained soft tissue section revealed patternless arrangement of spindle cells in a collagenous background with a prominent vascularity that results in a hemangiopericytoma-like pattern. Therefore, an interim diagnosis of solitary fibrous tumor was made based on the characteristic microscopic appearance. Subsequently, an excisional biopsy was done as the pain persists. Histopathological examination revealed sheets of spindle cells with focal areas of interspersed epithelioid cells. The spindle cells were plump shaped with indistinct cytoplasmic borders. Dense cellular component alternates with zones of acellular hyalinization. Cellular atypia was minimal. Mitotic figures were absent. Vascularity was moderate exhibiting hemangiopericytoma like areas and perivascular hyalinization of blood vessels. Differential diagnosis of hemangiopericytoma, fibrosarcoma and malignant fibrous histiocytoma was made.

Special stain like periodic acid Schiff stain was found to be negative indicating that these hyaline deposits are mesenchymal mucins secreted by the malignant spindle cells. Immunohistochemistry showed strong positivity for CD99, Bcl-2 and negative CD34, pan cytokeratin confirmed the diagnosis of monophasic fibrous synovial sarcoma. These results were consistent to the histopathological diagnosis.

The patient was referred to higher medical centre for further

management. The patient expired within three months as she neglected the treatment.

DISCUSSION

Synovial sarcoma was first defined by Simon in 1865⁶ and was labelled as SS by Sabrazes et al. in 1934³. The etiopathogenesis and the histogenesis of SS is undefined but thought to be originated from the mesenchymal pluripotential cells. They are labelled as SS as it resembles to the histological features of synovium or synovial membrane⁷. About 90-95% of SS revealed consistent, specific reciprocal translocation, t(X:18) (p11.2-q11.2) that forms the fusion of SYT with either SSX1 or SSX2 gene. The most common cytogenetic findings in monophasic fibrous tumour SYT: SSX2 whereas biphasic tumour demonstrates the fusion of SYT: SSX1^{8,9}. The SS is mainly mediated by the Wnt/ β -catenin, ERK and PcG signalling pathways¹⁰. In addition, EMT signalling pathway is also implicated in the molecular pathogenesis of SS¹¹.

Head and neck synovial sarcoma accounts for 5-10% of total sarcomas whereas occurrence of oral SS is 1.9%-3.7%⁵. SS is more commonly seen in the adolescents and young age group with a mean age of 15-40 years with a slight male predilection^{12,13}. Palpable deep-seated swelling with mild pain or tenderness is the common clinical manifestation of SS⁸. Early stages of SS present with an indolent swelling, upsurges over months to years, varying in sizes from 2-3 cms to 10-14 cms. Initial stages may mimic as benign lesion¹⁴. About 85-95% SS is predominantly seen in the lower extremities intimately associated with the tendon and tendon sheaths, knees, large joints and bursae¹². Para vertebral space is the second most common site of SS and manifest as solitary masses in the

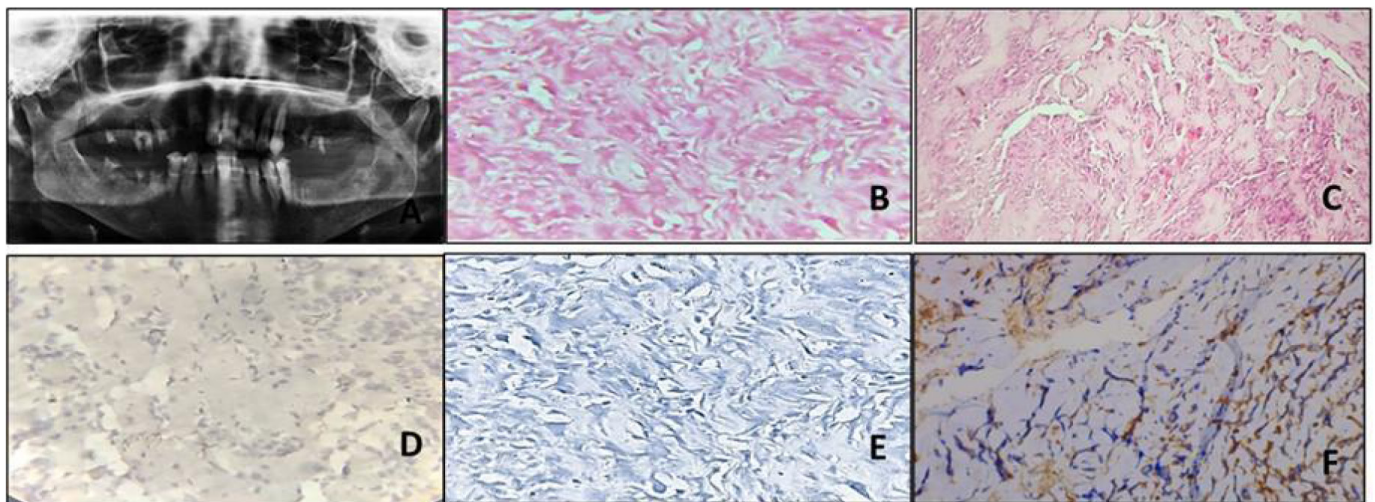


Fig. 1: OPG showing ill-defined radiolucency of size 3x3 cm retromolar region with limitation of movement of jaw [A]; Photomicrograph showing sheets of spindle cells with indistinct cytoplasmic borders \times 40X [B]; Dense cellular component alternates with zones of acellular hyalinization with hemangiopericytoma-like areas \times 40X [C]. Photomicrograph showing immunohistochemistry staining Pan cytokeratin [D]; CD 34 [E] negative in malignant spindle cells and CD99 positive in malignant spindle cells [F].

retropharyngeal or parapharyngeal region⁸. Oral SS manifest as minor limitation of movement, hoarseness, numbness of the lip, dysphagia¹⁵. Because of the unusual location, it is often misdiagnosed. In the present case, initially it was misdiagnosed as solitary fibrous tumour due to insufficient tissue specimen and of the rarity of oral SS.

Histologically, SS is classified into four sub types: biphasic, monophasic fibrous type predominantly consists of spindle cells, monophasic epithelioid cells and poorly differentiated tumours⁸. The monophasic fibrous type exhibits sheets of well oriented spindle cells having indistinct cytoplasm alternating with less cellular areas of hyalinization. The perivascular hyalinization and thick collagen bands separating the malignant spindle cells is the striking feature of monophasic fibrous type⁸. The reported case exhibits sheets of spindles with unclear cytoplasmic borders alternating with dense areas of hyalinization and perivascular hyalinization. Hemangiopericytoma, fibrosarcoma, malignant fibrous histiocytoma and leiomyosarcoma are considered as the differential diagnosis of this lesion. Periodic acid Schiff (PAS) stain was found to be negative as these hyalinized areas are stromal or mesenchymal mucins secreted by the malignant spindle cells. Immunohistochemical evaluation showed that tumour cells expressed CD 99 and 70% positivity of Bcl-2. CD34 and pancytokeratin were found to be negative in this case. A positive pancytokeratin antibody aids in separating sarcomas from carcinomas and epithelial tissues¹⁵. Because mucosal melanoma and malignant spindle cell carcinoma are epithelial in origin, these tumours were ruled out in this case by the negative pancytokeratin test. In some cases, uncertain histopathological features or mystifying immunohistochemical results, RT-PCR or FISH helps to diagnose oral SS.^{16,17} The specificity and sensitivity of RT-PCR test was approximately 83.8% and Fluorescent in situ hybridization (FISH) was 80% and the collective detection rate of both was 92.9%¹⁸.

Most reviewers pointed out that SS is a chemo sensitive sarcoma but radical local excision with adjunctive radiotherapy is recommended to be the successful treatment for Oral SS⁸. Oral SS most commonly metastasis to lung, bone marrow and lymph nodes⁵. High grade malignant SS has a recurrence rate of 80% whereas the recurrence rate of oral SS is 20.8%⁴.

The prognosis (5-year survival rate) for high grade SS is found to be 64-76% and in oral SS it is about 55%¹⁹. In addition, the prognosis is poor if the margins are inadequately cleared, without any adjunctive therapy, age, size and location of the tumour, high grade staging and metastasis¹¹.

The favourable clinical outcome of the patient includes clear surgical margin minimal cellular atypia, less than 5/10 HPF and presence of calcifications². Calcifications is always preceded by hyalinization. Extensively calcified SS has a good long-term prognosis. The median survival rate in case of extensively calcified SS was about 80% and in monophasic fibrous synovial sarcoma was about 50%⁸. In the present case, patient had a poor prognosis as the patient neglected the treatment.

CONCLUSION:

The term "synovial sarcoma" is thought to be misleading because it develops in areas where there aren't any typical synovial structures. Because of its odd location and rarity, oral SS is frequently misdiagnosed. Diagnosing monophasic SS is challenging in unusual locations than the biphasic variant of SS. For a precise diagnosis of oral synovial sarcoma, advanced molecular techniques including fluorescent in situ hybridization (FISH) and reverse transcription-polymerase chain reaction (RT-PCR) are required. When conventional approaches are ineffective for diagnosing oral synovial sarcoma, the identification of SYT-SSX2 fusion transcript by RT-PCR is an important diagnostic tool. Because of the oddity occurrence of Oral SS and locoregional recurrences, ideal treatment has not been established but radical surgery adjuvant with radiotherapy is the recommended treatment of choice. Because of the high recurrence rate, aggressive management and 10 years follow-up are mandatory for better prognosis.

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