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Monophasic Synovial Sarcoma of Mandible: A Rare Entity

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Abstract: Synovial sarcoma [SS] is a very rare soft tissue tumor accounting for 5.6 to 10% of all soft tissue sarcomas. Approximately 9% synovial sarcomas occur in head and neck region. SS is highly aggressive in nature and has tendency for distant metastasis. Most common site for head and neck synovial sarcoma is hypo pharynx, post pharyngeal region and parapharyngeal space. Very few cases of SS involving tongue, soft palate, mandible, buccal mucosa, and floor of the mouth have been described in the literature. Based on extensive literature review, very few cases of primary synovial sarcoma of mandible were noted in the literature .Here, we report a rare case of primary synovial sarcoma arising from mandible in a 40 year old lady and treated with definitive surgery.
Keywords: Synovial sarcoma, Mandible, Primary, Monophasic.

INTRODUCTION

Synovial sarcoma is a malignant soft tissue tumor that represents 5.6-10% of all soft tissue sarcomas [1]. Majority of tumors have been reported to occur in tendon sheaths, bursae and joint capsules, primarily in the para-articular regions of the extremities with approximately 9% occurring in the head and neck region. But its relationship to the synovium is not always obvious [2]. Exact origin of synovial sarcoma remains unknown, but the neoplasm may arise from pluripotential mesenchymal cells unrelated to synovial tissue [2, 3].

Jernstrom P in 1954 reported the first case of head and neck synovial sarcoma in a case involving the pharynx [4]. The most common sites in head and neck region include the hypo pharynx, post pharyngeal region, and the parapharyngeal space. Literature of few cases of SS that involves tongue, mandible, soft palate, buccal mucosa and floor of the mouth are also available [5-9]. Here, we report a rare case of primary synovial sarcoma arising from mandible and effectively treated with definitive surgical resection.

CASE REPORT

A 40 year old female patient, chronic tobacco chewer presented with growth over right lower alveolus since 3 months, associated with pain and occasional bleedng. No other significant history noted.

Oral examination revealed ulcero- proliferative growth over right gingivobuccal sulcus and lower alveolus around 3x 4 cm, extending right first premolar to left central incisor region and bleeds on touch (Fig. 1). No obvious cervical adenopathy.

Orthopantogram (OPG) revealed osteolytic lesion in the body of mandible (Fig. 2). Computed tomography (CECT) of face and neck showed ill defined heterogeneously enhancing soft tissue attenuation is seen involving body and alveolar process of mandible on right side causing ill defined expansile lytic lesion measuring 3.5 x 2x 2cm (Fig. 3). Incision biopsy of the lesion showed malignant spindle cell neoplasm and immunohistochemistry confirmed diagnosis of monophasic synovial sarcoma. Chest radiography was within normal limit.

Patient underwent right segmental mandibulectomy with supraomohyoid dissection (level 1-3). On gross examination of specimen showed 2.4 x 1.5 x 1.1 cm ulcero-infiltrative growth involving gingivobuccal sulcus and alveolus (Fig. 4 & 5). Bone appears involved on gross.

Microscopic examination showed spindle shaped cells, have pleomorphic ovoid to elongated nuclei with granular chromatin moderate cytoplasm with indistinct cell margins. Few mitotic figures are seen (Fig. 6). Margins were clear and regional nodes were negative for metastasis. Immunohistochemical

studies revealed tumour cells were strongly positive for vimentin (Fig. 7), focal positivity for TLE-1 , bcl-2 positivity and negativity for desmin, S-100, p63, pan CK, SMA, and CD 34. Postoperative course was uneventful and is disease free on 8 months of follow up.

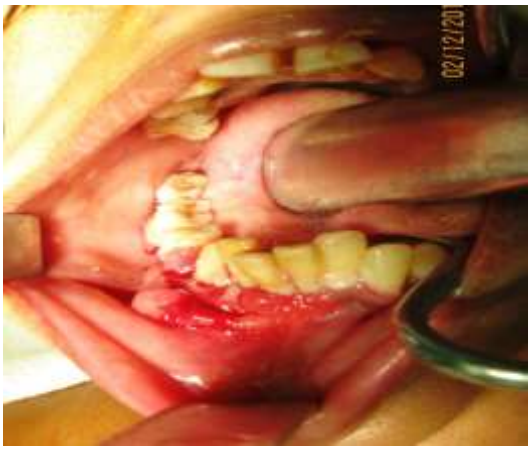

Fig. 1a & 1b: Clinical photograph showing ulceroproliferative lesion in the right alveolus

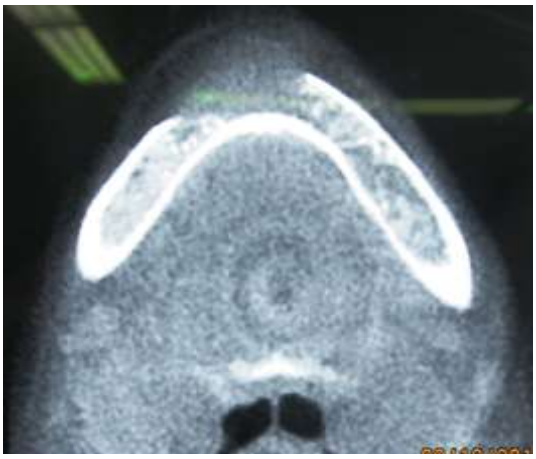

Fig. 2: Orthopantogram showing cortical erosion of the mandible

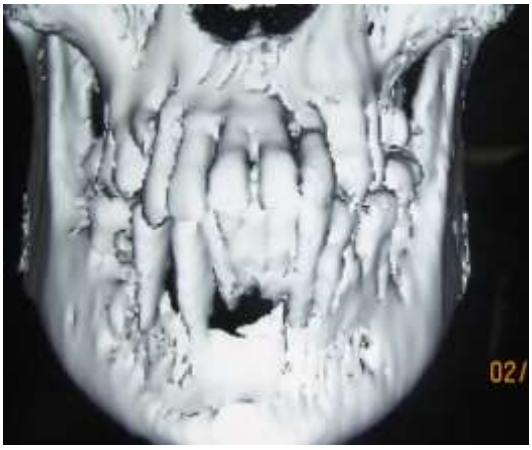

Fig. 3: CECT scan showed osteolytic lesion in the mandible

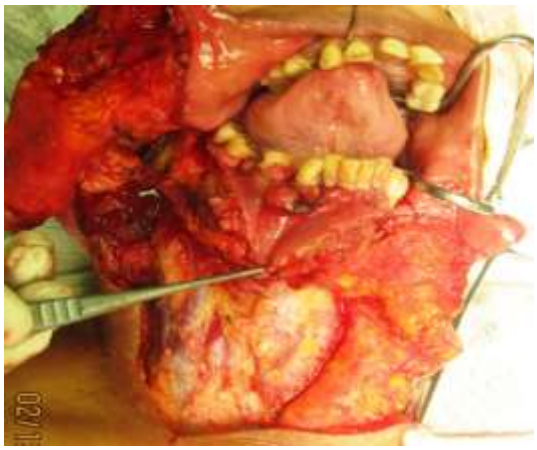

Fig. 4: Intraopertive photograph showing tumor arising from lower gingivobuccal sulcus

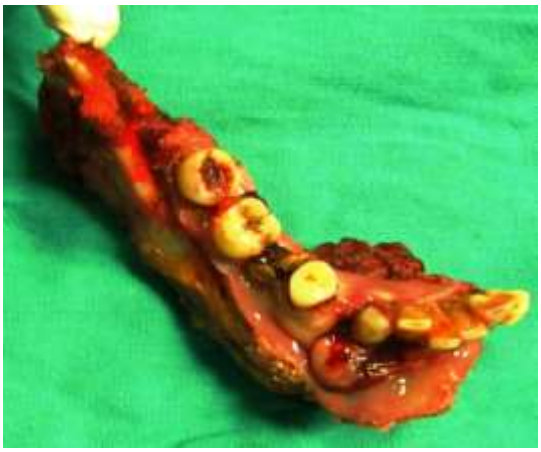

Fig. 5: Surgical resection specimen showing a 2.4 x 1.5 x 1.1 cm ulcero-infiltrative growth involving gingivobuccal sulcus and alveolus

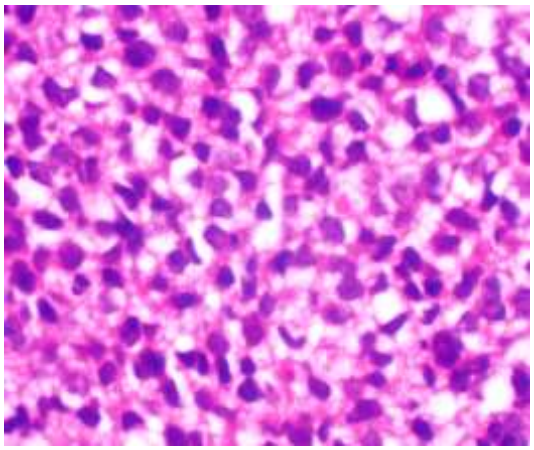

Fig. 6: Photomicrograph showing a hypercellular tumor with spindle shaped cells with granular chromatin moderate cytoplasm with indistinct cell margins

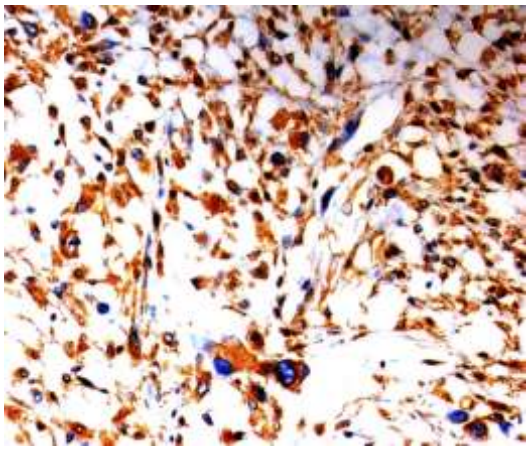

Fig. 7: Tumor cells are positive for Vimentin (DAB, x200)

DISCUSSION

Synovial sarcoma (SS) is an unusual malignant mesenchymal neoplasm that arises most commonly in joint capsules and articular tendons [1] and is the fourth most common soft tissue sacroma [10]. Its exact association with synovium is not always obvious [1]. Synovial sarcoma is a well defined clinical entity, described by Simon in 1865 [1]. The term "Synovioma" was first coined by Smith in 1927 and Knox described the term Synovial sarcoma in 1936 [1, 11].

Synovial sarcoma predominantly affects young adults and adolescents with slightly male dominance. Majority of head and neck SS occur in hypopharynx and parapharyngeal spaces. Less than 100 cases of head and neck SS have been reported in the literature. SS may also occur in tonsils, tongue, buccal mucosa, floor of the mouth and jaw bones. Tongue is the most common sub site for intraoral synovial sarcoma [5-9]. Mandible is an extremely rare site affected and very few cases of primary SS arising from mandible are reported in the literature. Exact aetiopathogenesis of SS is not known. No well defined predisposing factors associated with SS. Synovial sarcoma is thought to arise from primitive cells that can differentiate into either mesenchymal or epithelial elements [12]. Four subtypes of SS have been described in the literature, typically includes monophasic, biphasic (distinct epithelial and spindle elements), epithelial and poorly differentiated tumors (round cell) [13]. Poorly differentiated variety is highly aggressive in nature and have poor prognosis.

Immumohistochemistry (IHC) study reveals epithelial component stained positivity for cytokeratin (CK) and epithelial membrane antigen (EMA), while spindle element is strongly/focal positive for vimentin, Bcl-2, Mic 2, smooth muscle actin (SMA) and desmin. Bcl-2 protein is typically expressed in synovial sarcoma and CD 99, the product of MIC 2 gene is seen in 67% of all cases. Positivity for Vimentin, Bcl-2 and MIC-2 is characteristic of poor differentiated (small cell) variety of synovial sarcoma. In the present case, tumor cells were positive for vimentin, bcl-2 and negative for cytokeratin thus confirming the diagnosis of monophasic synovial sarcoma.

Based on molecular and cytogenetic studies, synovial sarcoma of other sites (extremities) and base of tongue s revealed a characteristic translocation involving chromosomes X and 18, t(X;18) as suggested by Bridge *et al*. in 1988 [14, 15].

SS should be best treated with definitive surgical resection wherever possible. Surgical treatment includes wide local excision with aim to achieve margin negative resection. Role of chemotherapy in SS is still debatable. It may have role in inoperable/metastatic setting with minimal to moderate efficacy. Various anecdotal reports describe use of several chemotherapeutic agents including cisplatin, doxorubicin, 5-fluorouracil, ifosfamide and etoposide with variable response rates. Role of radiotherapy is debatable. It is difficult to achieve complete excision of intraoral tumors with negative margins. Adjuvant radiotherapy is indicated in close or positive margin.

Head and neck synovial sarcoma have low metastatic potential and recurrence rate as compared to extremity sarcoma. Most common distant site for metastasis is lung, lymph nodes and the bone marrow. Recurrence rate following surgery is more common in the first 2 years following definitive treatment with a poor survival rate. Specific prognostic factors for SS comprise tumor size, site, and age over 60 years, high grade malignancy, margin of resection, mitotic index and the presence of distant metastasis.

In the present case, patient underwent curative surgical resection with negative margin and is disease free on 8 months of follow up. Synovial sarcoma is uncommon condition in the oral cavity and should be considered in histopathological differential diagnosis of both malignant primary and secondary spindle cell neoplasm's of oral cavity. Prognosis of intra oral SS remains poor despite curative resection as compared to squamous carcinoma of oral cavity.

CONCLUSION

Synovial sarcoma arising from mandible is very rare, highly aggressive condition with early tendency for distant metastasis. Wide surgical resection of the tumor and immediate reconstruction with bone graft gives normal contour of mandible with good cosmesis and functional outcome. Because of the rarity of SS in this location, a proper analysis is recommended for accurate diagnosis and improves the treatment outcome. Also, we would like to emphasize the critical role of special techniques such as immunohistochemistry and cytogenetic in the diagnosis

of monophasic SS. We present this case article to highlight the rarity of this entity, early diagnosis, proper treatment, disease outcome and utility of immunohistochemistry analysis.

The present case emphasizes the need for better detection, evaluation and analysis of such rare synovial sarcoma, to identify their natural course and effective treatment modalities.

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Abbreviations

SS- Synovial Sarcoma; OPG- Orthopantogram; CECT- Contrast Enhanced Computed Tomography; FNAC- Fine Needle Aspiration Cytology; IHC- Immunohistochemistry; CK- Cytokeratin; SMA- Smooth Muscle Antigen

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