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Recurrent Synovial Sarcoma of Floor of Mouth: A Rare Case Report

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Abstract
Background: *Synovial sarcoma is typically an aggressive malignant tumor of soft tissues, usually in the extremities but rare in head and neck region. They are called Synovial sarcoma because of their histologic resemblance to the synovium, but they do not involve a synovial structure and are believed to originate from pluripotent mesenchymal cells.*
Case Presentation: *We report a case of a 23-Year old male who had a complaint of swelling over left chin region previously operated in 2015 and diagnosed as spindle cell sarcoma of floor of mouth which was low-grade, now presented with recurrent swelling over floor of mouth so the patient underwent surgical intervention for the same in September 2021 at our institute and diagnosed by immunohistochemistry as recurrent synovial sarcoma of floor of mouth, then the patient was referred to our radiation oncology department for adjuvant treatment.*
Conclusion: *Synovial Sarcoma is rare in the oral cavity, which is slow growing and aggressive in nature. Misdiagnosis results in the delay of diagnosis and treatment of Synovial Sarcoma. Immunohistochemistry analysis might be the most important tool to confirm the diagnosis of Synovial Sarcoma. Treatment should involve wide local excision followed by adjuvant Radiotherapy depending on patient-specific factors. Lung is the most common site of metastasis.*
Keywords: *Recurrent Synovial Sarcoma, Immunohistochemistry, Radiation, Lung metastasis.*

Introduction

Synovial sarcoma (SS) is a rare type and accounts for 5 to 10% of all soft tissue sarcomas, only 3% of which are presented in the head and neck region also known as malignant synovioma.[1,2] It is generally accepted that SS originates from primitive undifferentiated cells and not the synovial structures and is the fourth most common malignancy after malignant fibrous histiocytoma, liposarcoma, and rhabdomyosarcoma.[2,3,4]

The median age of presentation is between 15 to 40 years old with slight male predominance.[3,4] Head and neck synovial sarcoma are having high incidences in hypopharynx, post pharyngeal region, and parapharyngeal space, rare incidences of SS in the tongue, soft palate, mandible, buccal mucosa, floor of mouth were also noted.[1,2,3,4]

Diagnosis of oral synovial sarcoma is difficult because of atypical clinical features and uncertain location.[4] Owing to its rarity, synovial sarcoma of

head and neck is often misdiagnosed until late in the course of the disease process.[2] A diagnosis of synovial sarcoma is dependent on a histopathological and immunohistochemistry study.[1]

The treatment of choice for this type of tumor is surgical excision with a wide safety margin whenever possible followed by adjuvant radiotherapy can be given. Chemotherapy's role has not been well established.[1]

We report a rare case of recurrent SS of floor of mouth in a 23-year-old male patient.

Case Presentation

A 23-year-old male patient reported a swelling over left chin region and neck mass in 2015, radiological diagnostic workup followed by Fine needle aspiration cytology (FNAC) was done from left submandibular swelling showing acute sialadenitis. He had undergone surgery, it had been histopathologically diagnosed as spindle cell sarcoma of low grade. The patient was not given any adjuvant treatment and was kept on observation.

After a disease-free interval of 5 years, he developed a recurrent swelling over floor of mouth in 2021. Magnetic resonance imaging (MRI) Diagnostic Scan showed a 46×31×44 mm lesion in the anterior two third of floor of mouth, infiltrated left side of tongue, and with another skip lesions or metastasis of 16×10×13 mm in posterior one-third of the left side of tongue. (fig.1 A & B) FNAC from left submandibular region showed malignant recurrent spindle cell sarcoma. Following the diagnosis, Wide local excision of floor of mouth with submandibular and sublingual gland excision and platysma flap reconstruction surgery was performed. The histopathology was spindle cell proliferation with palisading nuclear arrangement, fascicles and hyalinization present. (fig. 2 A & B)

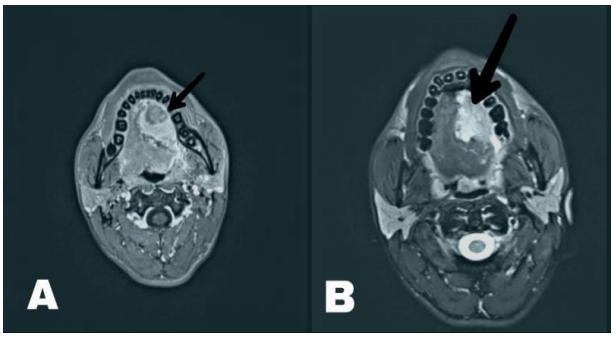

Figure 1: (A) Contrast-enhanced T1 weighted **(B)** T2 weighted axial section MRI showing hyperintense lesion in anterior 2/3rd of floor of mouth and going to lateral border of tongue on left side.

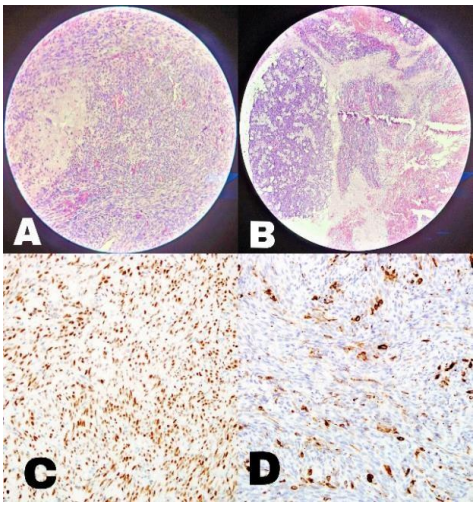

Figure 2: A & B shows a tumor composed of spindle cells arranged into fascicles, ill-defined nuclear palisading arrangement, hyperchromatic with hyalinization in between spindle cells. [Hematoxylineosin, A- High power view (40x), B- Low power view (10x)]. **C**: TLE1 positive shows spindle cells, 10x view, **D**: AE1 positive in tumor, 10x view.

Immunohistochemistry examination was done to confirm the diagnosis. (Figure 02: C & D)

Marker	Status
AE1	Positive
TLE1	Positive
CK7	Negative
S100	Negative
DESMIN	Negative
ACTIN	Negative
SOX10	Negative
CD34	Negative

A postoperative clinical examination was done, and on basis of pathological and clinical findings, the patient was planned for post-operative radiation therapy by conformal technique. He underwent adjuvant radiation therapy with the dose prescription of 66Gy in 30 fractions with conventional fractionations, 5 fractions a week, by Intensity modulated radiation therapy (IMRT) on Linear Accelerator at our institute (fig. 03).

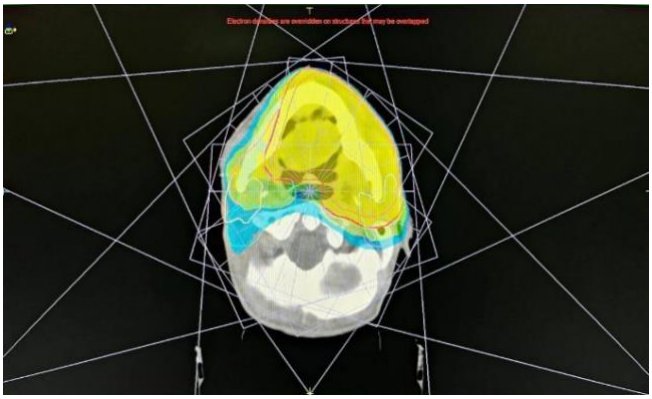

Figure 3: Conformal IMRT Planning showing CTV (orange color) and Isodose of PTV 66, 60 and 54Gy dose in yellow, green, and blue in color respectively.

After adjuvant radiation on 2nd month follow up Computed tomography (CT) diagnostic scan showed residual lesion over the same site as previously involved with reduction of size of tumor and showed lung metastasis in bilateral lung fields, largest mass present 20×20 mm in left lower lobe. The patient was then referred to the medical oncology department. The Patient was given palliative chemotherapy Ifosfamide and Doxorubicin for 3 cycles. The patient was re-evaluated with CT scan which showed an increase in the size of lung metastasis. The patient started on palliative chemotherapy with Gemcitabine. (fig.04)

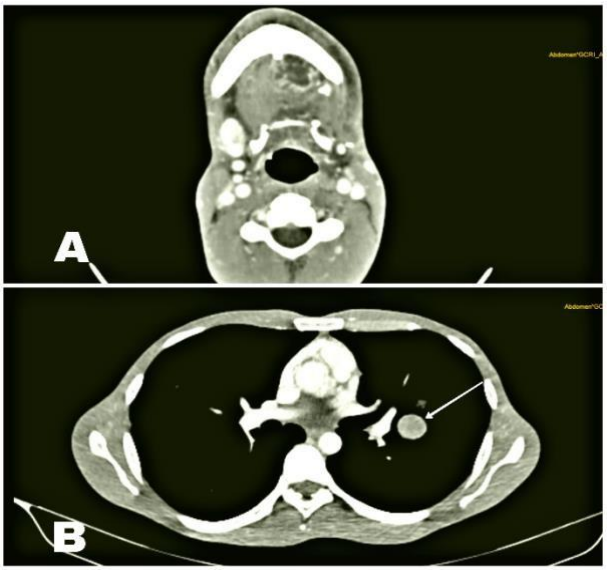

Figure 4: Axial view of contrast-enhanced CT scan images shows **A)** heterogeneously enhancing residual lesion over left anterior 2/3rd of floor of mouth and tongue. **B)** lung metastasis in left lower lobe.

Discussion

SS was first documented by Simon in 1865 and was so named in 1934 by Sab razes et al. Jernstrom was the first to report the occurrence of SS in head and neck region in 1954.[3,4,5,6,7] Despite its name SS it doesn't originate from synovial tissue, but generally, it is accepted that SS is derived from primitive undifferentiated or pluripotent mesenchymal cells with multiple differentiation potential.[1,3,4] Adolescents and young adults of 15 to 40 years are mainly affected with slight male predominance.[3,4]

SS typically shows high histologic grade and poor prognosis. SS usually occurs in the lower limbs and trunk and appears as an asymptomatic mass until it acquires sufficient volume to cause compressive effects on neighboring structures. It can be associated with symptoms such as dysphagia, hoarseness, earache, sore throat, limited mouth opening, lower lip numbness, and bleeding depending on the site of origin of tumor.[1,4,5,9] It is usually a slow-growing tumor and easily be confused with benign tumors in the early stage, as the gradual increase of tumor size, shows same symptoms as oral squamous cell carcinoma.[4]

CT, MRI, and Positron emission tomography (PET) scan can be used as diagnostic tools.[3,7] Despite lack of specific findings, CT and MRI are still useful for determining the location of the primary tumor, adjacent tissue infiltration, and metastasis. A chest x-ray is useful in looking for any pulmonary metastasis.[2]

The gold standard for the diagnosis of SS is a tissue biopsy and pathological examination.[2] The specimen may be obtained by either FNAC or an open biopsy.[2] IHC reveals tumor is usually positive with AE1/AE3, EMA, CD99, vimentin, CEA, BCL2 and negative with CD34, CD31, actin, or myoglobin, but still there is no specific immunological marker specific to SS has been found.[1,8]

Cytogenetics is extremely helpful in confirming the diagnosis. A characteristic translocation (t (X; 18) (p11, q 11): SYT-SSX1 or SYT-SSX2 genes) is seen in 90% of cases.[3,6] SS can be classified into three histopathological subtypes: 1) monophasic SS-containing Uniform spindle cells or epithelial cells, 2) biphasic SS-composed of epithelial cells arranged into glandular structures with spindle cells arrange into fascicles, 3) poorly differentiated SS characterized by the presence of spindle and /or round blue cells.[8] Regardless of the subtype, the conventional clues and IHC evidence of the epithelial element are the characteristic of SS.

For treatment, there is still no standard protocol available. Surgical interventions, for example, wide local excision and adjuvant radiation with or without chemotherapy is recommended.[4,5] Lymphatic spread is not a feature of the natural course of this tumor, and for that prophylactic neck dissection is not done. However, when palpable neck disease is present, it should be treated with a node dissection.[2] At present, for SS in head and neck, simple surgical treatment can be recommended for smaller and superficial lesions; but for the larger and deeper SS, surgery followed by adjuvant radiation should be considered, since it is difficult to perform complete resection as many nerves and vessels are involved in head and neck region. The defects after resection can be repaired with flap transfer.[4] Likewise, there is no evidence that chemotherapy alters the course of the disease. In fact, it is well known that chemotherapy does not affect sarcomas to a great extent at any site.[2] Metastasis may develop in 30-60% of patients and like other sarcomas, lung is the most common site of metastasis and also can metastasize to lymph nodes and bone marrow. Tumor recurrence typically manifested in the first 2 years after the initial therapy.[5,7] Prognosis is generally poor (5-year survival rate- 55%). Prognosis is adversely affected by a tumor size of more than 5 cm, tumor site, age over 60 years, high-grade malignancy, and the presence of metastatic disease.[5,9]

Conclusion

We present this report to add to the existing literature about SS of the head and neck. SS of oral cavity is slow growing and aggressive in nature with a poor prognosis. In view of rarity of this malignancy, the diagnosis can be difficult, IHC is required for the confirmation of diagnosis. Treatment includes wide local excision followed by adjuvant radiotherapy depending on subjective histopathological findings, followed by chemotherapy if metastasis developed. In our case patient is having reduce the tumor size post-radiation but had a residual lesion and developed lung metastasis, as the lung is the most common site of metastasis.

Conflict of interest: There are no statements with conflicts of interest.

Ethical Standard: Ethics approval and consent have been taken by The GCRI (Gujarat Cancer and Research Institute) Institutional review committee.

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Abbreviations

SS - Synovial Sarcoma
FNAC - Fine Needle Aspiration Cytology
CT - Computed Tomography
MRI - Magnetic Resonance Imaging
PET - Positron Emission Tomography

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