Case Report

# Aggressive monophasic synovial sarcoma involving sublingual gland: First case

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## ABSTRACT

Synovial sarcoma (SS) of the arising head and neck region is rare. The most common site being the hypopharynx. No case of SS from the sublingual of 35-year old salivary gland has been reported. We report a case of monophasic SS in sublingual gland a female making it the first documented case of English Language literature.

**Key words:** Aggressive monophasic synovial sarcoma, head and neck, sublingual gland

## Introduction

Synovial sarcoma (SS) is a malignant mesenchymal tumor mainly involving extremities and comprise up to 10% of all soft-tissue sarcomas. These tumors occur more frequently in lower extremities at para articular locations, although it may be found in areas unrelated to synovial tissues.[1] Head and neck SSs account for less than 0.01% of all malignancies.[2] Head and neck SS was first described in 1954 by Jernstrom, who reported a case of SS of the pharynx.[3] Since that time, numerous cases of head and neck SS have been reported. The majority of these lesions are located in the pre-vertebral, parapharyngeal and retropharyngeal spaces from the skull base to the hypopharynx. Occasionally, such tumors are located near articulations, such as the cricoarytenoid, sternoclavicular and temporomandibular joints. To the best of our knowledge, no case of monophasic SS occurring in the sublingual gland has been reported in the literature until date. This makes our report first documented case in the English Language literature.

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## Case Report

A previously healthy 35-year-old female patient presented with 1½ year history of gradually progressive swelling in the floor of mouth on the left side. She had no difficulty in speech or swallowing. Intraoral examination revealed a smooth swelling of floor of mouth on the left side [Figure 1]. There was no other lymph node palpable in the neck. Results of routine laboratory tests were within the normal limits. Cytological analyses fine-needle aspiration (FNA) of the mass pointed to a mixed salivary tumor-pleomorphic adenoma. Contrast enhanced computed tomography scan showed 3.2 cm × 1.8 cm well-defined homogenously enhancing lesion in the floor of mouth on the left side with no abnormal regional lymph nodes [Figure 2].

At surgery, tumor was well encapsulated with lobulations and enbloc excision along with the capsule was done [Figure 3]. On histopathology, the tumor contained monotonous proliferation of small spindle shaped or ovoid neoplastic cells with pale, eosinophilic cytoplasm [Figure 4]. On immunochemical staining, tumor cells were strongly positive for Mic 2, calponin and focally for cytokeratin (CK), Vimentin and bcl2. They were negative for epithelial membrane antigen (EMA), S100 protein and HMB45. Final diagnosis of monophasic synovial sarcoma from sublingual gland was made. Patient was advised adjuvant radiotherapy. With only 10# of radiotherapy she developed mucositis and refused any further treatment. On follow-up she developed

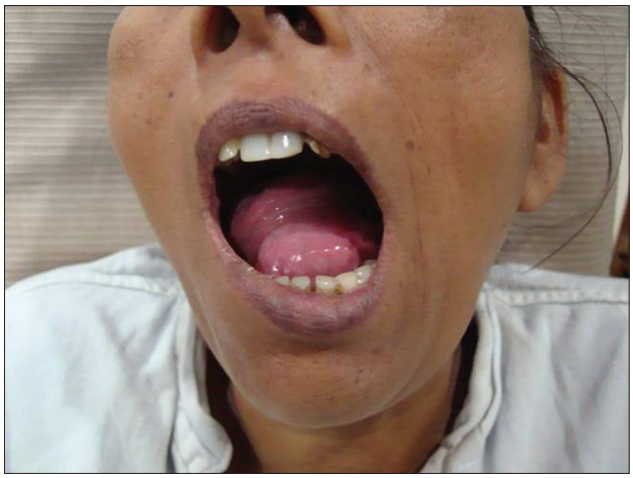

**Figure 1:** Intra oral picture showing smooth mass in the left floor of mouth anteriorly

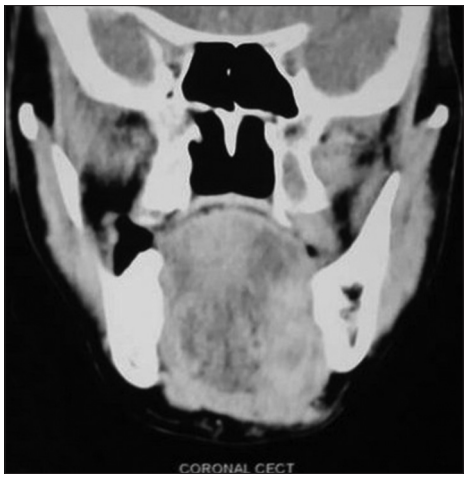


**Figure 2:** Contrast enhanced computed tomography scan showed well defined homogenously enhancing lesion in the floor of mouth on the left side

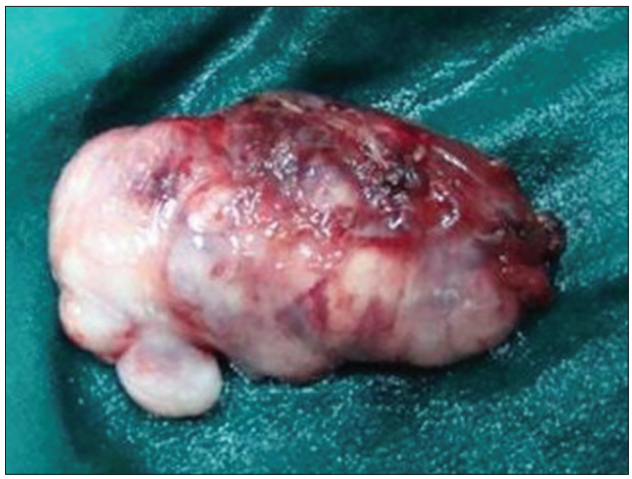

**Figure 3:** Surgical specimen showing lobulated well encapsulated mass

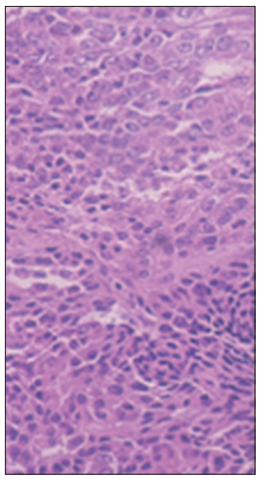

**Figure 4:** Photomicrograph displayed the epithelial pattern characterized by large, round or oval cells with focal nuclear palisading in a scanty spindle component

huge recurrence locally and died of local disease after 6 months.

## Discussion

SS of the salivary glands is rare and cases of monophasic pattern in the sublingual gland have not been reported in the literature previously. The case presented here is the first documented report of monophasic SS arising in the left sublingual gland. SSs are most commonly seen in adolescents and young adults between 15 and 40 years of age.[4] The patient under study was 35 years of age. The most common site involved by SS is lower limb (60-75%) followed by upper limb (15-20%).[4] The head and neck region accounts for 6-7% of all cases of SS. The most common site in the head and neck is hypo pharynx.[5]

SS cases with a biphasic pattern are easy to diagnose microscopically when located in any area of the body. The diagnosis at the rare sites like head and neck is problematic. They may sometimes be wrongly diagnosed as epithelial tumors such as poorly differentiated squamous cell carcinomas or myoepitheliomas.[5] Monophonic SS is even more likely to be misdiagnosed and have been mistaken for other soft tissue tumors such as hemangiopericytoma, fibrosarcoma and malignant peripheral nerve sheath tumors.[5] The diagnosis in our patient was difficult, the FNA was reported as mixed tumor of salivary gland origin (pleomorphic adenoma). Considering this diagnosis we did local excision of the sublingual gland. It was on histopathology that the tumor displayed the epithelial pattern characterized by large, round or oval cells and formed a glandular, cleft-like or cyst-like space. Nuclear palisading in spindle component was also noticed. Therefore diagnosis of malignant myoepithelioma was suggested and tissue subjected to immunohistochemistry. Immunohistochemistry confirmed the diagnosis of monophasic SS. In the present study, the tumor cells were positive for Mic 2, calponin and focally for CK,

vimentin and bcl2. They were negative for EMA, S100 protein and HMB45. There are no clear cut guidelines for the management of the SS of the head and neck region. The ideal treatment for head and neck SS is surgery with complete excision of the tumor.[6-8] If it is not possible to excise the tumor completely, then post-operative radiotherapy is recommended. Accurate diagnosis of SS helps to rule out sarcamatoid carcinoma and thus avoids unnecessary nodal clearance. It is also important to differentiate SS from the primitive neuroectodermal tumor as for the later chemotherapy is required. Most authorities believe that head and neck SS is as aggressive as its counterpart in the trunk and extremities.[6,9] Others believe head and neck tumors to be generally less aggressive, with a somewhat "better" prognosis.[8,10] This has been attributed to early detection when the size is small and easy accessibility in the head and neck region. Since these tumors in the head and neck occur in younger patients who are otherwise healthy. In our case, she developed huge local recurrence within 3 months of surgery. This reveals the aggressive nature of the pathology. She was advised adjuvant radiotherapy, which she could not tolerate and refused any further treatment. More research needs to be done to see the real behavior of SSs of head and neck.

## Conclusion

SS is an aggressive tumor that can occur nearly anywhere in the body including salivary glands. SS should be kept as differential diagnosis in the swellings of salivary glands. This is important since the diagnosis may not be apparent until after pathological examination of the surgical specimen, resulting in alterations to the optimal treatment strategy and negative impact on the patient.

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