



## Case Report

### Primary Synovial Sarcoma of the Mandible: Report of a Case

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#### Abstract

Synovial sarcomas of the head and neck region are uncommon, accounting less than 10% of all head and neck soft tissue sarcomas. The most frequent sites are hypopharynx and parapharyngeal space. Synovial sarcomas are extremely rare in the oral cavity and may increase the potential for misdiagnosis.

Only a few intraoral synovial sarcomas have been well documented. In this report we present a case of a primary synovial sarcoma of the mandible of a 15-year-old male. The histological features are described and diagnostic, prognostic and therapeutic problems are discussed. Special immunohistochemical stains and cytogenetic studies can help in confirming the diagnosis. Therapeutic standards are lacking due to the rarity of synovial sarcomas in the oral cavity. Combined modality therapy of this aggressive tumor better results; however the 5 year survival rate of these patients is poor.

**Keywords:** Malignant soft tissue tumor; Mandible; Oral synovial sarcoma.

#### Introduction

Synovial Sarcoma (SS) is a rare and highly malignant soft tissue tumor with a predilection to the lower extremities of adolescents and young adults. More usually the tumor is "Parasynovial" or adjacent to joint capsules, tendon sheaths or bursae, although an increasing number of SSs are found to arise in areas remote from any articular or periarticular structure [1]. Although almost any site can be affected, a distinct region of involvement is the head and neck, especially the hypopharynx, the postpharyngeal region and the parapharyngeal space [1,2]. It was eventually realized that SS did not arise from synovial cells, but was presumed to differentiate from an undefined mesenchymal cell.

SS has been reported only rarely in the oral cavity. There have been reported only few cases of synovial sarcomas of the

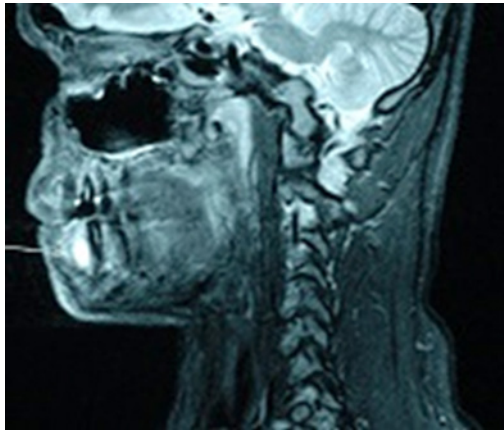
mandible. The aim of this article is to report one additional case in a 15-year-old male and discuss the related diagnostic, prognostic and therapeutic challenges.

#### Case Report

A 15-year old male was referred to our department for evaluation and treatment of recurrence of an intraoral biphasic synovial sarcoma of the mandible. There was a painless mass of soft constitution, without ulceration at the right side of the mandible, extending from the canine to the premolar region. There was no paresthesia of the right inferior alveolar nerve. The patient's medical history was unremarkable. On examination, his general health appeared well.

The patient underwent 7 months earlier in another hospital, a surgery for an intraoral lesion that revealed glandular biphasic synovial sarcoma. He did not receive any postoperative adjuvant therapy. At presentation, MRI control showed a mass of 1.27 X 0.75 cm in the right side of the mandible. In T1 gadolinium contrast

-enhanced images the mass produced homogenous density signal (Figure 1).

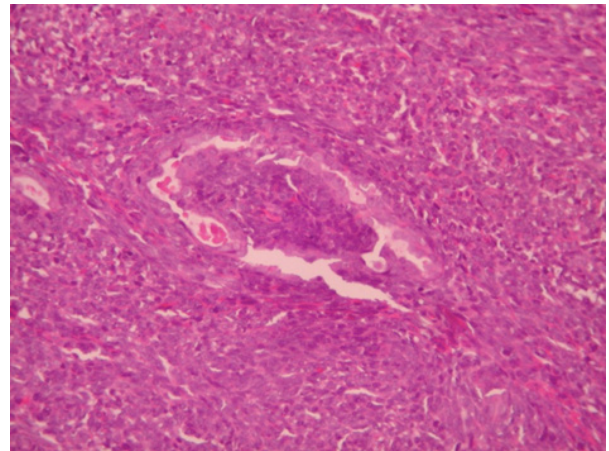


**Figure 1:** MRI showing the homogeneous signal of the mass and its relation to the mandible. Computer tomography showed the relation of the lesion with the body of the mandible (Figure 2).



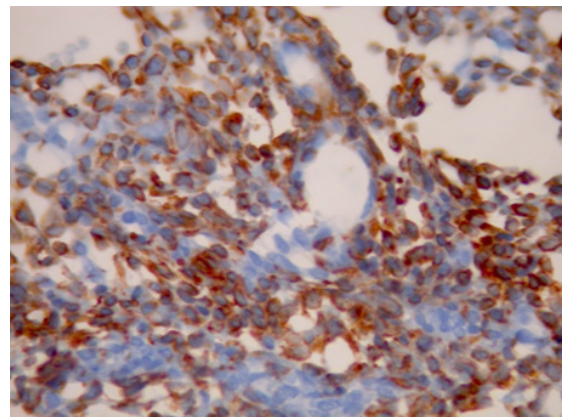
**Figure 2:** CT showing the mass in direct relation with the right mandible. Submandibular, submental lymph nodes and lymph nodes from the posterior cervical triangle were found enlarged.

The tumor was widely excised through a marginal mandibulectomy and a simultaneously performed selective neck dissection. Grossly, the specimen contained a 2.0 cm in greatest dimension, fleshy and red-gray mass. The surgical margins of the specimen were negative and in a safe distance of more than 1 cm. There were no cervical metastases. Histologically the tumor contained spindle cells. Some epithelioid cell and no glandular formations were found (Figure 3).



**Figure 3:** Synovial sarcoma: spindle and some epithelioid cells (H+E X200)

Immunohistochemical staining was performed according to standard protocols and antibodies directed against EMA, keratins, S-100, Bcl-2, SMA, Vimentin, CD99, CD34, CD57, Desmin were used (Figure 4).



**Figure 4:** The tumor cells are positive for Vimentin immunostain (X400)

The new histological diagnosis mentioned monophasic synovial sarcoma. The recovery was uneventful. The patient was referred to the oncological council with the question of the need of adjuvant radiotherapy. A wait-and-see decision was made, based on the wide excision and the free of metastasis neck. Two years later the patient is healthy and free of disease.

## Discussion

Synovial Sarcoma (SS) is a rare, malignant soft tissue tumor of children and adults representing 5.6% to 10% of all soft tissue sarcomas [3]. Almost 60% of the patients are between 15 and 40 years of age. It can occur in almost any part of the body [1,4], with approximately 9% occurring in the head and neck region. The first description of SS was by Pack and Ariel in 1950 [5]. There are only 7 cases affected the mandible reported in the literature (Table 1).

Case	Author	Age	Sex
1	[1]	28	Male
2	[2]	10	Male
3	[6]	42	Male
4	[4]	29	Female
5	[7]	11	Male
6	[8]	20	Female
7	[9]	76	Male
8	Present case	15	Male

**Table 1:** Primary Synovial Sarcoma of the Mandible: Review of cases in the literature

The cell of origin for this tumor is believed to be an uncommitted mesenchymal cell that undergoes synovial differentiation [3,10]. Histologically, the following subtypes can be distinguished: a) monophasic SS containing spindle cells, b) biphasic SS, containing both epithelial and spindle cell components, c) a poorly differentiated type of SS (20% of SS) and d) a rare form of monophasic SS, containing only epithelial-like cells [11]. Nearly 90 to 95% of SS demonstrates specific t (x; 18) (p11.2-q11.2) chromosomal translocation that forms the SYT-SSX fusion gene [8]. In some cases, the cytogenetic analysis of the tumor cells can be of great help in clarifying the histogenesis. Histological differential diagnosis varies with the sub-type of synovial sarcoma. It can include low-grade fibrosarcoma, undifferentiated spindle cell carcinoma, malignant schwannomas and carcinosarcomas.

Wide surgical resection and local control should be the primary surgical goal [11]. SS was noted to have increased sensitivity to high-dose ifosfamide-based chemotherapy with encouraging results [12]. SS has been long recognized as a high grade soft tissue sarcoma with a high metastatic propensity. Late distant metastases particularly to the lungs may occur up to 10 years after initial treatment [13]. Tumors greater than 5 cm seem to have a significantly poorer 10-year patient survival (49%) than

tumors of 5 cm or less (77%). The 5-cm criterion has been verified as a useful prognostic factor in this disease by several investigators [12]. In conclusion, radical surgical intervention and long-time follow-up are necessary for patients with synovial sarcoma.

## Acknowledgement

No conflict of interest.

## References

1. Torsiglieri AJ Jr, Hendrix RA, Quinn PS (1991) Synovial sarcoma of the jaw. *Ear Nose Throat J* 70: 396-398.
2. Captier G, Montoya P, Duché R, Le Barazer M, Bigorre M, et al. (1999) Synovial sarcoma of the mandible in children. Apropos of a case. *Rev Stomatol Chir Maxillofac* 100: 187-191.
3. Meer S, Coleman H, Altini M (2013) Oral synovial sarcoma: A report of 2 cases and a review of the literature. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 96: 306-315.
4. Tilakaratne WM (2006) Synovial sarcoma of the mandible. *J Oral Pathol Med* 35: 61-63.
5. Rao GV, Sravya T, Sivaranjani Y, Bhat VR (2014) Primary biphasic synovial sarcoma of gingiva: Report of a rare case. *J Oral Maxillofac Pathol* 18: 77-80.
6. Koga C, Harada H, Kusukawa J, Kameyama T (2005) Synovial sarcoma arising in the mandibular bone. *Oral Oncology extra* 41: 45-48.
7. Granowetter L, Ladas E, Taromina K, Rooney D, Kelly KM (2006) Integrative tumor board: Pediatric synovial sarcoma. *Integr Cancer Ther* 5: 48-55.
8. Tao Q, Qiao B, Wang Y, Hu F (2011) Diagnosis and treatment of primary synovial cell sarcoma that occurred in the left mandible body: a case report and literature review. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 111: e12-e20.
9. Khalili M, Eshghyar N, Ensani F, Shakib PA (2012) Synovial sarcoma of the mandible. *J Res Med Sci* 17: 1082-1085.
10. Mahesh KT, Ponnuswamy IA, David MP, Shivhare P, Puttaranganayak MI, et al. (2013) Synovial sarcoma of the buccal mucosa: a rare case report. *Case Rep Dent* 2013: 938291.
11. Wang Y, Zhu F, Wang K (2020) Synovial sarcoma of the floor of the mouth: a rare case report. *BMC Oral Health* 20: 5.
12. Spillane AJ, A'Hern R, Judson IR, Fisher C, Thomas JM (2000) Synovial Sarcoma - a Clinicopathologic Staging and Prognostic Assessment. *J Clin Oncol* 18: 3794-3803.
13. Alberty J, Dockhorn-Dworniczak B (2020) Monophasic synovial sarcoma of the neck in an 8-year-old girl resembling a thyroglossal duct cyst. *Int J Pediatr Otorhinolaryngol* 2002: 61-65.