Case Report

Monophasic (spindle cell) synovial sarcoma of mandible

Jagannath Dev Sharma, Argha Baruah, Lopa Mudra Kakoti, Anupam Sarma, Shiraj Ahmed, Amal Chandra Kataki[1]

Departments of Pathology and [1]Gynaecological Oncology, Dr. B. Borooah Cancer Institute, Guwahati, Assam, India

Abstract Synovial sarcoma (SS) is a malignant soft-tissue tumor of uncertain histogenesis. It is a malignant mesenchymal tumor commonly affecting the extremities. Rarely, it is seen in areas without any relationship to synovial structures. Only 6%–7% of cases have been reported in the head-and-neck region. In head-and-neck region, it usually involves the hypopharynx, parapharyngeal space, and posterior pharyngeal region. A few cases in mandible have been described in literature. Hereby, we report a rare case of monophasic (spindle cell) SS of the mandible in a 17-year-old male with a previous history of tooth extraction along with an unhealed socket.

Keywords: Mandible, spindle cell, synovial sarcoma

Address for correspondence: Dr. Argha Baruah, Room No. 111, Department of Pathology, Dr. B. Borooah Cancer Institute, Guwahati - 781 028, Assam, India.
E-mail: argha20@gmail.com

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INTRODUCTION

Synovial sarcoma (SS) is a malignant mesenchymal tumor commonly affecting the extremities. Only 6%–7% of cases have been reported in the head-and-neck region.[1] The term "synovial sarcoma" was first coined in 1936 by Knox.[2] The first reported case of SS in head and neck involving the pharynx was described by Jernstrom in 1954.[3]

SS arises in association with tendon sheaths, bursae, and joint capsules. On rare occasions, it is seen in areas without any relationship to synovial structures. In head-and-neck region, it usually involves the hypopharynx, parapharyngeal space, and posterior pharyngeal region. A few cases in the tongue, buccal mucosa, floor of mouth, soft palate, and mandible have been described in literature.[4]

CASE REPORT

A 17-year-old male presented with swelling in the lower right side of the face for 2 months. The swelling was associated with pain and discharge. There was a previous history of extraction of the right molar tooth following which the socket did not heal. On extraoral examination, this swelling was 6 cm × 5 cm in size, was firm in consistency, and was tender on palpation near the angle of mandible. The right submandibular lymph node was palpable which was mobile and nontender measuring 2 cm × 1.5 cm. Intraoral examination revealed an unhealed socket along with purulent discharge. On computed tomography scan, a destructive

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lesion was seen in the right hemimandible involving the body, ramus, and condylar process along with a heterogeneously enhancing soft-tissue component infiltrating the masticator space and retromolar trigone [Figure 1].

Wide local excision along with right-sided hemimandibulectomy and supraomohyoid neck dissection was carried out for the patient, and histopathological examination revealed a moderately pleomorphic spindle cell tumor with scattered mitotic figures [Figure 2]. A diagnosis of spindle cell sarcoma was made, and immunohistochemistry (IHC) was done on the paraffin block of the tumor tissue. The tumor cells were immunoreactive to CD99, epithelial membrane antigen (EMA), BCL2, and focally vimentin [Figures 3-6] and showed negativity for cytokeratin (CK), smooth muscle actin (SMA), and S100.

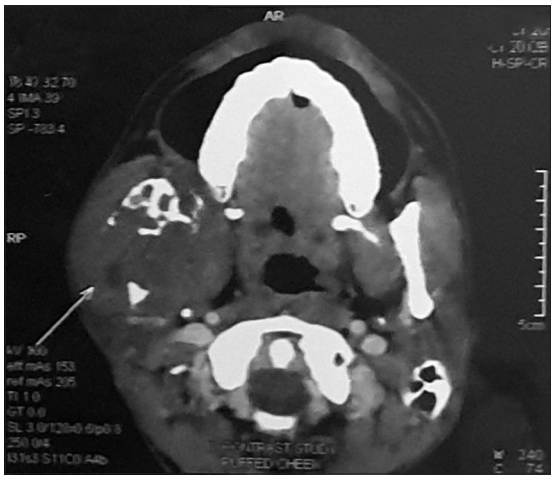

Figure 1: Computed tomography scan shows a destructive lesion in the right hemimandible involving the body, ramus, and condylar process along with heterogeneously enhancing soft-tissue component

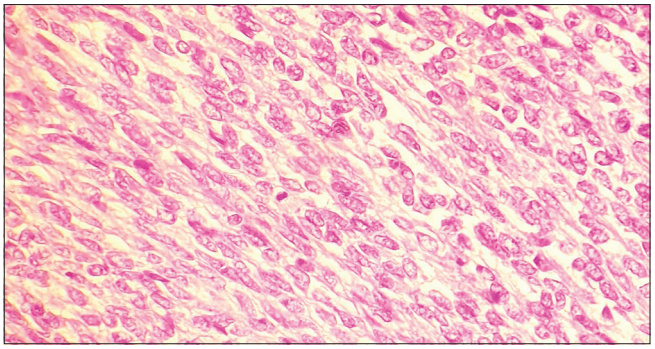

Figure 2: Spindle cells with moderate pleomorphism and scattered mitotic figures (H and E, 40X)

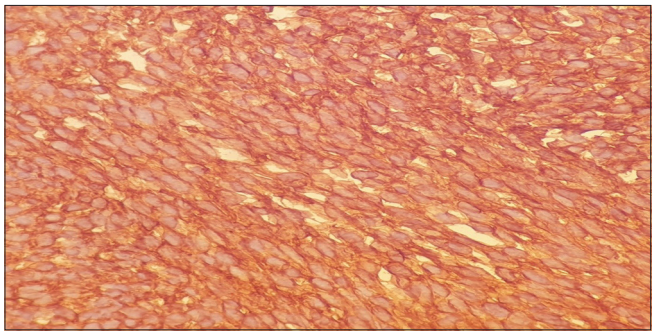

Figure 3: Spindle cells showing positivity for CD99 (×40)

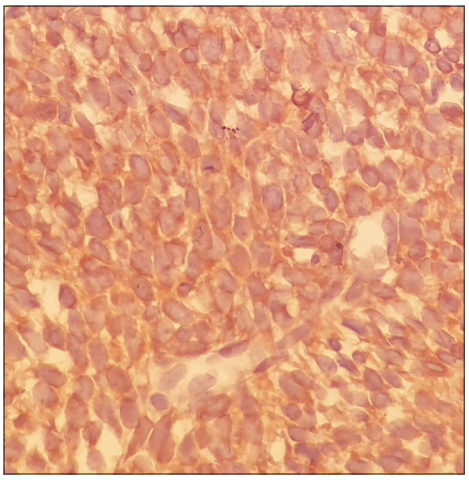

Figure 4: Spindle cells showing positivity for epithelial membrane antigen (×40)

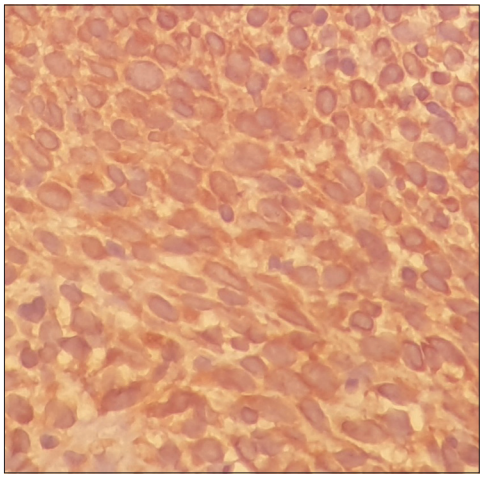

Figure 5: Spindle cells showing positivity for BCL2 (×40)

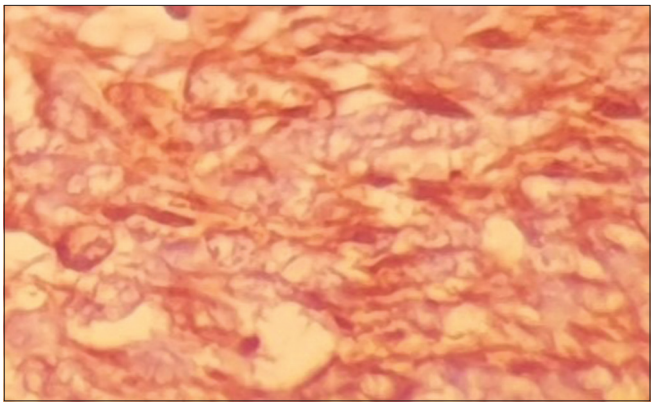

Figure 6: Spindle cells showing positivity for vimentin (×40)

A final diagnosis of "monophasic (spindle cell) synovial sarcoma" was made.

DISCUSSION

SS is a malignant soft-tissue tumor of uncertain histogenesis, which has the potential to differentiate into either mesenchymal or epithelial component. In spite of the name, the cells of origin are not synovial cells. Histogenesis is still debated.

SS can occur at any age but most commonly seen in the early years of life (second to third decade). SS is most commonly located in the extremities (60%–70%). SS has been detected in unusual sites, such as the head and neck, retroperitoneum, bones, nerves, lungs, pleura, heart, and kidney. Only 6%–7% of cases have been reported in the head-and-neck region.[5] In previous studies, the most common site in the head and neck was involvement of the parapharyngeal space followed by the hypopharynx.[6] In a study of 167 cases of head-and-neck SS (HNSS), oral cavity and parotid gland were found to be the most common sites of involvement.[7] In a study by Salcedo-Hernández *et al*., HNSS occurred commonly in males in their third decade of life.[8] In our case report, the patient was male and 17 years of age and SS affected the mandible. Few cases of SS have been reported in the mandibular region. Bansal and Sipayya reported a case of monophasic SS of mandible in a 16-year-old female patient.[9] Wadhwan *et al*. reported a case of biphasic SS of mandible in a 28-year-old male patient.[10]

Histologically, SS is of two main subtypes: biphasic and monophasic spindle cell. There are other rarer subtypes such as monophasic epithelial, poorly differentiated, calcifying/ossifying, and myxoid. Monophasic spindle cell variant is more common. The spindle cells are usually arranged in fascicles along with uniform tapering nuclei with pale cytoplasm. They can have a hemangiopericytoma-like vascular pattern. Epithelial component consists of numerous glandular structures lined by cuboidal or columnar epithelium. Poorly differentiated SS has an undifferentiated round cell morphology.

A myriad of histologic presentation causes discrepancy in diagnosis and also initial misdiagnosis. The closest differential diagnosis in monophasic spindle cell type includes spindle cell squamous cell carcinoma, malignant peripheral nerve sheath tumor, solitary fibrous tumor, spindle cell rhabdomyosarcoma, and leiomyosarcoma. As SS is located in the mandible, odontogenic fibroma, ameloblastic fibroma, and ameloblastic fibrosarcoma should be included in the differential diagnosis.[11] Poorly differentiated type is commonly misdiagnosed as Ewing's sarcoma because it consists of sheets of atypical small and large cells.[12]

Immunohistochemically, around two-thirds of SS are positive for CD99. Transducer-like enhancer of split 1 is overexpressed in SS. It is helpful to distinguish SS from histologic mimic in difficult cases and is a sensitive and specific marker.[13] Other markers that are frequently positive are BCL2, EMA, and beta-catenin. S100 and SMA should be done to rule out neurogenic or smooth muscle origin of the cells. In our case, the tumor cells showed positivity for vimentin, CD99, BCL2, and EMA and showed negativity for CK, SMA, and S100.

SS is characterized by the presence of the *t* (X; 18) (p11.2; q11.2) translocation. Ninety percent of SS shows SS18-SSX fusion oncogenes.[14]

The primary treatment option for SS is wide surgical resection. Prognosis is poor, and many patients develop lung metastases after prolonged follow-up.[15] Radiotherapy and adjuvant chemotherapy is helpful in high-risk situations (tumors >5 cm or difficult to resect). The present case is on regular follow-up without adjuvant treatment.

Poor prognostic factors include poorly differentiated subtypes, older age at diagnosis, size ≥5 cm, mitotic activity ≥10/10 high-power field, higher Ki67 activity, and positive surgical margins.[16] In a study, it was found that HNSS had similar survival rates compared to SS of limbs.[8] However, several reports indicate that the prognosis of HNSS is better than that of SS arising in the extremities.[7]

CONCLUSION

Because of rarity of this neoplasm in the mandibular region, it is more likely to be misdiagnosed and may pose a diagnostic challenge to pathologists. Therefore, correct diagnosis with the aid of histology and IHC is required to prevent misdiagnosis and improve treatment outcome.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Park JK, Ham SY, Hwang JC, Jeong YK, Lee JH, Yang SO, *et al*. Synovial sarcoma of the head and neck: A case of predominantly cystic mass. AJNR Am J Neuroradiol 2004;25:1103-5.
2. Knox LC. Synovial sarcoma: Report of three cases. Am J Cancer 1936;28:461.
3. Jernstrom P. Synovial sarcoma of the pharynx; report of a case. Am J Clin Pathol 1954;24:957-61.
4. Wang H, Zhang J, He X, Niu Y. Synovial sarcoma in the oral and maxillofacial region: Report of 4 cases and review of the literature. J Oral Maxillofac Surg 2008;66:161-7.
5. Salcedo-Hernández RA, Lino-Silva LS, Luna-Ortiz K. Synovial sarcomas of the head and neck: Comparative analysis with synovial sarcoma of the extremities. Auris Nasus Larynx 2013;40:476-80.
6. Thway K, Fisher C. Synovial sarcoma: Defining features and diagnostic evolution. Ann Diagn Pathol 2014;18:369-80.
7. Bukachevsky RP, Pincus RL, Shechtman FG, Sarti E, Chodosh P. Synovial sarcoma of the head and neck. Head Neck 1992;14:44-8.
8. Mallen-St Clair J, Arshi A, Abemayor E, St John M. Factors associated with survival in patients with synovial cell sarcoma of the head and neck: An analysis of 167 cases using the SEER (surveillance, epidemiology, and end results) database. JAMA Otolaryngol Head Neck Surg 2016;142:576-83.
9. Bansal A, Sipayya V. Monophasic synovial sarcoma presenting as a primary mandibular mass. Indian J Oral Sci 2015;6:78-80.
10. Wadhwan V, Malik S, Bhola N, Chaudhary M. Biphasic synovial sarcoma in mandibular region. J Oral Maxillofac Pathol 2011;15:239-43.
11. Bertolini F, Bianchi B, Pizzigallo A, Tullio A, Sesenna E. Synovial cell sarcoma of the neck. Case report and review of the literature. Acta Otorhinolaryngol Ital 2003;23:391-5.
12. Kashyap B, Reddy PS, Kumar RN. Poorly differentiated synovial sarcoma of the mandible. Oncobiol Targets 2016;3:1-3.
13. Terry J, Saito T, Subramanian S, Ruttan C, Antonescu CR, Goldblum JR, *et al*. TLE1 as a diagnostic immunohistochemical marker for synovial sarcoma emerging from gene expression profiling studies. Am J Surg Pathol 2007;31:240-6.
14. Barnes L, Eveson JW, Reichart P, Sidransky D. World Health Organization Classification of Tumors. Pathology and Genetics, Malignant Soft Tissue Tumors. Lyon: IARC Press; 2005. p. 149.
15. Tilakaratne WM. Synovial sarcoma of the mandible. J Oral Pathol Med 2006;35:61-3.
16. Harb WJ, Luna MA, Patel SR, Ballo MT, Roberts DB, Sturgis EM. Survival in patients with synovial sarcoma of the head and neck: Association with tumor location, size, and extension. Head Neck 2007;29:731-40.