CASE REPORT

Primary biphasic synovial sarcoma of gingiva: Report of a rare case

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ABSTRACT

Synovial sarcoma is a mesenchymal spindle cell tumor with variable epithelial differentiation. It is unrelated to the synovium as the name might suggest but arises in the soft tissues of the extremities around the knee joints and tendon sheaths. The tumor cells are thought to resemble normal synovial tissue histopathologically, hence named "synovial sarcoma" (SS). Head and neck lesions are less common and oral cavity involvement is extremely rare. Few cases in tongue, soft palate, mandible, buccal mucosa and floor of mouth have been described in the literature. Here, we probably report the first case of primary biphasic SS (BSS) involving gingiva in the retromolar area of the mandible in a 21-year-old male patient.

Key words: Gingiva, spindle cell tumor, synovial sarcoma

INTRODUCTION

Synovial sarcoma (SS) is a mesenchymal spindle cell tumor with variable epithelial differentiation.[1] It accounts for 8-10% of all soft tissue malignancies.[2] It is the fourth most common type of sarcoma following malignant fibrous histiocytoma, liposarcoma and rhabdomyosarcoma. It was first documented by Simon in 1865.[3] The term synovioma was coined by Smith in 1927 but Knox in 1936 defined it histologically.[4] The first description of SS in the head and neck (H and N) region was by Pack and Ariel in 1950. According to Amble *et al*., approximately 9% tumors occur in this region.[5]

SS is most commonly seen between 15-40 years of age. Males are more susceptible than females with ratios of 1.2:1.[3] The most common sites in the H and N region include hypopharynx, post-pharyngeal region and parapharyngeal space. Few cases in tongue, soft palate, mandible, buccal mucosa and floor of mouth have been described in the literature.[2] To our knowledge, this is probably the first case of SS involving gingiva being reported in the English literature.

CASE REPORT

A 21-year-old male patient reported with a swelling in lower left back tooth region since 6 months. The growth started as a small painless nodule that slowly increased in size. Intraorally there was a pedunculated and ulcerated growth covered with slough, extending from the distal aspect of 37 to uvula, measuring approximately 5 × 3 cm [Figure 1]. On palpation the swelling was firm in consistency, non-tender and freely movable. Grossly decayed 36 with a draining sinus was noticed. A provisional diagnosis of peripheral giant cell granuloma was made.

Panoramic radiograph showed no lesion-related radiological findings. Other findings include radiolucency in the periapical region of 36. The tumor was surgically excised and surgical margin was clear of any tumor tissue.

The histopathology revealed cellular areas with a biphasic pattern and less cellular myxoid areas. The cellular areas with cuboidal cells consisting of eosinophilic cytoplasm and hyperchromatic nuclei arranged in nests, along cleft-like and glandular spaces were seen. These were surrounded by sheets of spindle cells with marked atypical nuclei [Figure 2]. Many mitotic figures and areas of necrosis were noticed.

Immunohistochemistry revealed strong nuclear positivity in both the components for transducer-like enhancer of split 1 (TLE1) [Figure 3], epithelial membrane antigen (EMA) positivity [Figure 4] in the cuboidal epitheloid cells, vimentin [Figure 5] and Bcl-2 positivity [Figure 6] in the spindle cells, which confirmed the diagnosis of biphasic SS (BSS). The patient is on regular follow up since one year and no recurrence has been noticed.

DISCUSSION

SS is a clinically, morphologically, histologically and genetically well-defined entity that may arise from primitive

Access this article online
Quick Response Code:
Website: www.jomfp.in
DOI: 10.4103/0973-029X.131916

undifferentiated pleuripotential mesenchymal cells unrelated to synovial tissue.[5] Mittinen and Virtanen in 1984 suggested SS is a carcinosarcoma-like tumor with true epithelial differentiation and the term SS is a misnomer.[2] According to Leader *et al*., SS can be more appropriately classified as carcinosarcomas based on frequent coexpression of epithelial and mesenchymal markers such as vimentin and cytokeratin.[6,7]

In head and neck (H and N) region, parapharyngeal region is the most frequently affected site.[5] Notably, intraoral cases are

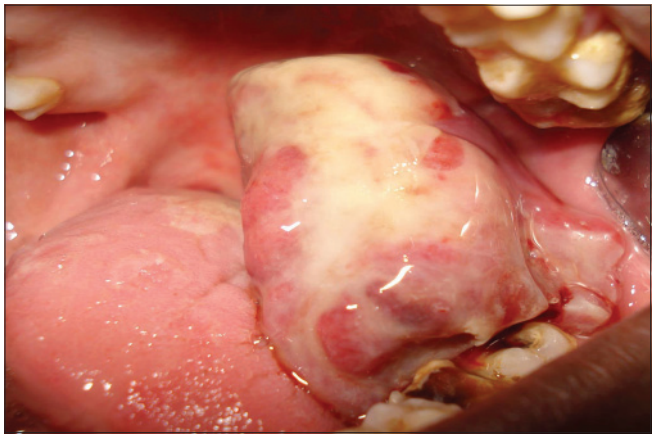

Figure 1: Tumor extending from distal aspect of 37 to uvula

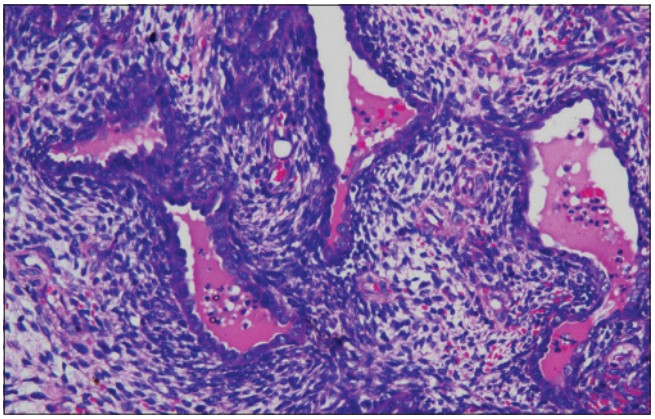

Figure 2: Photomicrograph showing epithelial component around the glandular spaces and arranged in solid nests surrounded by spindle component. (H&E stain, x400)

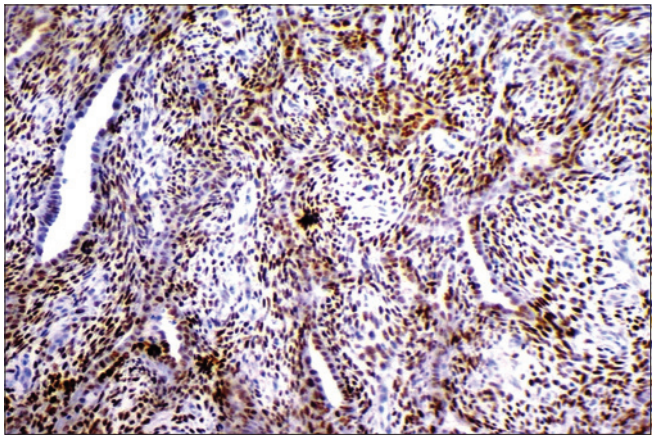

Figure 3: Immunohistochemistry revealed strong nuclear positivity of tumor cells for Transducer-like enhancer of split 1 (TLE1) in both spindle and epithelial component (IHC stain, ×100)

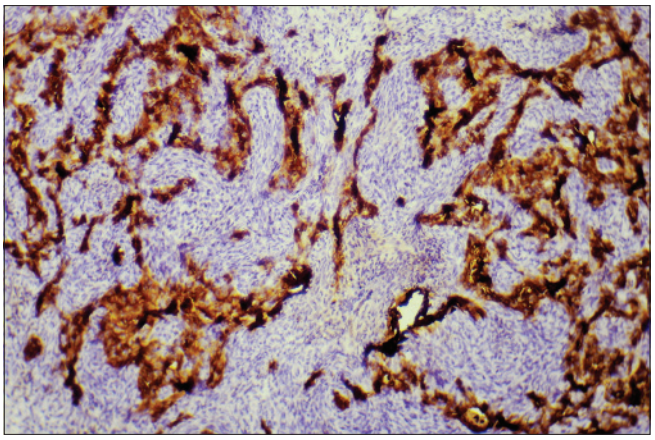

Figure 4: Immunohistochemistry revealing Epithelial Membrane Antigen (EMA) positivity in the cuboidal epitheloid cells and negativity in the spindle cells (IHC stain, ×100)

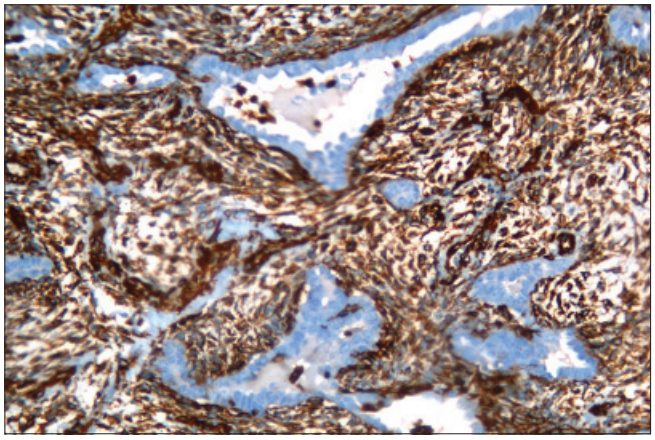

Figure 5: Immunohistochemistry revealing vimentin positivity in the spindle cells and negativity in the cuboidal cells (IHC stain, ×400)

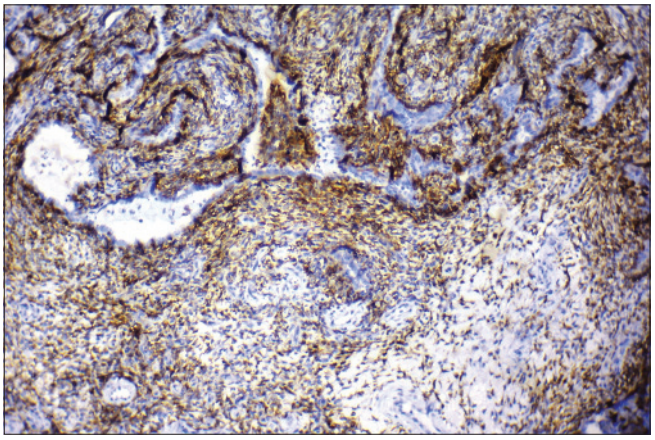

Figure 6: Immunohistochemistry revealing Bcl-2 positivity in the spindle cells (IHC stain, ×100)

extremely rare. Only 37 cases were reported by 2009 with the tongue being the common intraoral site.[6] This is probably the first case of primary biphasic variant of SS involving gingiva of the mandible.

Clinically SS presents as a mass with or without pain and is a slow growing tumor increasing in size over 1-2 years. The size is varying from 3-10 cm, which is either well circumscribed or infiltrative.[1] In our case, the swelling is painless, slow growing and attained a size of 5 × 3 cm approximately.

Depending on prominence of the cells, SS are subclassified into three groups: i) monophasic epithelial cell, ii) monophasic spindle cell and iii) biphasic type with distinct epithelial and spindle cell components. In addition to the three subtypes, Enzinger and Weiss have described a "poorly differentiated" type of SS.[8]

BSS contains distinct but intermingled epithelial and spindle cell components. The epithelial component consists of cuboidal to tall columnar cells around the glandular spaces and arranged in solid nests. These cells are characterized by large, round or oval nuclei and abundant pale cytoplasm with distinct cellular border. The glandular lumen often contains epithelial mucin. The spindle cell component consists of well-oriented, rather plump, small and uniform spindle cells with a high nuclear-cytoplasmic ratio, growing in solid sheets or fascicles. Mitotic figures may be found in both the cell components.[5]

Immunohistochemically TLE1 is a sensitive and specific marker for SS and can be helpful to distinguish SS from other histological mimics.[9] The spindle cells of SS show strong and uniform expression of vimentin with occasional positivity of cytokeratin particularly in biphasic variant. In all, 90% of SS demonstrates strong cytokeratin positivity. Other epithelial markers like CK7, CK19 and EMA are also positive.[7] Intense Bcl-2 cytoplasmic positivity is seen only in SS besides lymphoma. However, hemangiopericytoma, fibrosarcoma, leiomyosarcomas, malignant peripheral nerve sheath tumor and malignant mesothelioma are Bcl-2 negative[10] and thus can be differentiated. Our case showed immunoreactivity for EMA in epitheloid cells and vimentin and Bcl-2 positivity in the spindle cells.

Cytogenetically more than 90-95% SS demonstrates specific t(x; 18) (p11.2;q11.2) chromosomal translocation.[5-7] The detection of SYT-SSX fusion transcript is a diagnostic marker of SS. It can be used to confirm tumor-free margins during surgical resection and to verify the presence of metastatic disease. SYT-SSX1 fusion is present in both monophasic or biphasic type, whereas SYT-SSX2 fusion is seen only in monophasic variant.[5,7]

Prevention of local recurrence and distant metastasis are pivotal functions of treatment.[6] The standard treatment in H and N SS is surgery with adequate wide excision. Radiotherapy has an established role in improving local control after inadequate surgical resection. In our case, wide surgical excision was done with a regular follow up since one year.[8] Posttreatment recurrence rate for SS arising from all body sites is about 80%. A 5-year survival rate is about 36-51%.[7]

CONCLUSION

To the best of our knowledge, this is the first case of primary BSS occurring in the gingiva. These tumors should be maintained in the working histopathologic differential diagnosis of both malignant primary and metastatic spindle cell tumors of the oral cavity as there is a chance for misdiagnosis. The final diagnoses of these tumors were confirmed by histological and immunohistochemical means.

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How to cite this article: Rao GV, Sravya T, Sivaranjani Y, Bhat VR. Primary biphasic synovial sarcoma of gingiva: Report of a rare case. J Oral Maxillofac Pathol 2014;18:77-9.

Source of Support: Nil. **Conflict of Interest:** None declared.