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
Biphasic variant of synovial sarcoma

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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

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**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, ×400)

**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.

Cytogetenically 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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