INTRODUCTION

Solitary fibrous tumor (SFT) is a rare mesenchymal cell neoplasm that was initially described in the pleura by Lietaud in 1767, followed by Wagner in 1870.\(^1\) The occurrence in the head and neck region are uncommon, accounting for only 6% of all cases.\(^2\) To the best of our knowledge, there are 38 case reports of SFTs originating from the cheek, buccal space or buccal mucosa of the oral cavity in English literature.\(^3\) However, presentation in maxilla is rare, with only a few previously reported cases. SFT and hemangiopericytoma (HPC) are soft tissue tumors with known histologic and immunohistochemical overlap.\(^4\) SFT is preferred by most pathologists as a better term than “HPC” that gathers numerous unrelated entities and is presently employed only by neuropathologists.\(^5\) The fourth edition of the World Health Organization classification of tumors of soft tissue and bone “blue book” published in February 2013 has abandoned the term “HPC”\(^6\) and has grouped SFT, HPC, lipomatous HPC and giant cell angiofibroma under the “extrapleural SFT” category.

CASE REPORT

A 30-year-old female patient was referred to the Department of Oral Pathology, with a history of growth in the upper right molar region for the past 2 months. The mass slowly increased in size with occasional pain and bleeding. Extraorally there was no swelling or trismus. Clinical examination revealed a 5 cm × 5 cm × 3 cm well circumscribed, nodular, nontender, nonfluctuant mass, in relation to the distal aspect of the upper right third molar region extending backward to the pharynx [Figure 1]. The right submaxillary lymph nodes were palpable and tender.

Magnetic resonance imaging showed a 3.7 cm × 1.4 cm × 2.2 cm well-defined dumbbell-shaped lesion, occupying the region posterior to right upper 3rd molar tooth. The lesion was...
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Hyperintense in T2W1 with widening of the pterygomaxillary fissure and was seen extending laterally. Provisional diagnosis was given by the surgeon as vascular malformation, as the lesion tend to bleed during surgery.

Macroscopic examination revealed a firmly circumscribed mass, the cut section of which was gray-white which excluded the clinical diagnosis. Histopathology showed tumor cells arranged around dilated vascular spaces in a storiform pattern [Figure 2]. Alternating zones of hypercellular and hypocellular area were also seen [Figure 3]. Individual tumor cells were round to ovoid and spindle shaped with a moderate amount of eosinophilic cytoplasm and elongated vesicular nucleus with bland chromatin. On the basis of the histopathology, the initial diagnosis was given as soft tissue tumor. However, immunohistochemical studies were done and tumor was diffusely positive for CD34 [Figure 4] and was negative for CD99 [Figure 5], Bcl-2 [Figure 6], vimentin, desmin, S-100 and CD68. These features were consistent with that of SFT, and the final diagnosis was given as SFT.

DISCUSSION

Klemperer and Rabin in 1931 classified pleural tumors into two types: Diffuse mesotheliomas and localized mesotheliomas or SFT. SFT is one generally associated with serosal surfaces, but in recent years, it has been reported in extrapleural sites such as liver, adrenal glands, skin and less commonly in the head and neck region. Experts suggest that the majority of lesions previously termed HPCs are in fact, SFTs that do not show pericytic differentiation.[7]

Ultrastructural and IHC studies of HPC as defined by Enzinger and Smith have suggested that it is a tumor of undifferentiated mesenchymal cells rather than true microvascular pericytes. It has been pointed out that the cells comprising these tumors are
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SFT occurs mostly in middle-aged adults (between 20 and 60 years) and affects both sex equally. The tumor presents as a slow growing, painless mass. In the present case, histopathology showed ovoid to spindle-shaped tumor cells arranged around dilated vascular spaces in a storiform pattern amidst alternating zones of hypercellular and hypocellular areas. In some zones, the cells are arranged in short, ill-defined fascicles, whereas in others they appear randomly in a “patternless pattern.” Although the striking feature of SFT, the characteristic hyalinization, was absent in our case, all other features favored the final diagnosis as SFT. SFT also has a malignant counterpart that shows features such as hypercellularity, high mitotic figures (>4/10 HPF), cytologic atypia, tumor necrosis and infiltrative margins. These features were absent in the present case.

SFT of pleural and extrapleural origin typically express CD34 (80–90%), CD99 (70%), Bcl-2 (30%) and EMA (30%). Desmin, cytokeratin and S-100 protein are usually negative. In the present case, microscopy showed spindle cells with a focal pericytic vascular pattern with diffuse CD34 antigen positivity. This excludes the diagnosis of HPC which typically shows staghorn pattern of the vasculature with strong CD34 positivity. Moreover, spindle-shaped cells are absent in HPC.

The histopathological diagnosis of this vascularized tumor is challenging, in particular, because of the difficulty in differentiating SFT from other tumors types that have prominent vascularization such as schwannoma, myofibroblastoma, metastasis, spindle-cell carcinoma, low-grade fibromyxoid sarcoma, synovial sarcoma and malignant peripheral nerve sheath tumor.

CONCLUSION

SFT is a rare soft tissue tumor in the head and neck region and rarer in the oral cavity. Many soft tissue lesions in the oral cavity can mimic SFT clinically. SFT resembles many malignant tumors in the head and neck region and should be properly assessed using histopathology. The final diagnosis depends solely on histopathology and immunohistochemistry (IHC). In our case although the clinical diagnosis was vascular malformation, histopathology and IHC favored the diagnosis of SFT. Immunohistochemical staining once again became crucial in establishing the diagnosis of SFT.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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