

Solitary fibrous tumour (previously termed hemangiopericytoma) tongue – a rare case report

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Abstract

Solitary Fibrous Tumor (SFT) is a rare soft tissue tumor which has fibroblastic origin. SFT constitutes a heterogeneous group of rare spindle-cell neoplasms that include benign and malignant entities. It was originally described in the pleura. They may arise in non serosal areas like orbit, thyroid, nasal cavity and soft tissues. Herein, we present a case of 35 year old male who came with a painless swelling on the tip of the tongue. Histopathological examination of the excised tissue showed hypercellular and hypocellular areas with spindle shaped cells arranged in whorls and fascicles with interspersed collagen. Hemangiopericytomatous growth pattern was evident. Immunohistochemistry revealed positivity for Vimentin, CD34 and BCL-2, negativity for S-100, cytokeratin and desmin. IHC for Ki 67 revealed a low proliferative index. A diagnosis of solitary fibrous tumor was made. The case is being presented due to its rarity.

Keywords: CD34, Hemangiopericytoma, Solitary Fibrous Tumor, Tongue

Introduction

SFTs were originally described as localized fibrous mesotheliomas, but later it was reported to be of non-mesothelial origin.⁽¹⁾ Their cell of origin is still debated. Although SFT frequently occurs in the pleura, growing number of SFT cases have been reported in extrapleural locations such as peritoneum, liver, meninges, orbit, thyroid, nasal cavity and soft tissues.⁽²⁾ SFTs are rare in the oral cavity and even rarer in the tongue. They mostly exhibit a benign behavior and malignant transformation is exceptional. We herein present a case of solitary fibrous tumor of tongue.

Case Report

A 35 year old male presented with a pedunculated soft tissue mass on the tip of the tongue for four months (Fig. 1A). The growth was firm, painless and gradually enlarging. There was no other finding on clinical examination. Past history and family history were not significant. The growth was surgically excised and sent for histopathological examination. Grossly, it was well encapsulated firm, measuring 2.5x2x1cms.

Cut surface showed glistening grey white areas along with foci of hemorrhage. Microscopically, the sections showed well circumscribed tumor tissue consisting of both hypocellular areas alternating with hypercellular areas (Fig. 1B). The hypercellular areas consisted of proliferation of mildly pleomorphic elongated spindle cells in sheets, fascicular and whorling distribution (Fig. 1C). Intercellular areas showed collagen formation (Fig. 1D). Haemangiopericytoma pattern was evident with the tumor cells arranged around the blood vessels (FIG. 1E). Mitoses was rare (2/10 HPF). There was no evidence of necrosis in any of the sections. Masson trichrome stain demonstrated collagen positivity (Fig. 1F).

Immunohistochemistry showed positivity for Vimentin, CD34 and BCL-2 (Fig. 2A-C). The Ki-67 labeling index was <10 % (Fig. 2D). The tumor was negative for S100, Desmin (Fig. 2E-F). VEGF showed positivity in endothelial cells (Fig. 2G). The final diagnosis based on morphology and immunohistochemistry was Solitary Fibrous Tumor of tongue.

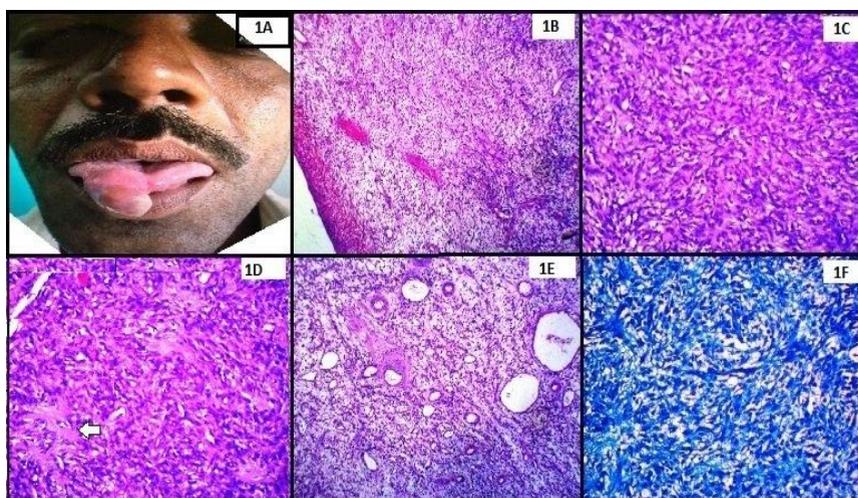


Fig. 1: Showing (a) Solitary mass present on tip of the tongue; (b) Hypercellular and hypocellular areas(H & E 40x); (c) Hypercellular area with proliferating spindle cells in whorling pattern(H&E 100x); (d) Collagenous bands in the intercellular area(H&E 100x); (e) Hemangiopericytoma pattern(H&E 100x) (f) collagen demonstrated with Masson Trichrome Stain (100x)

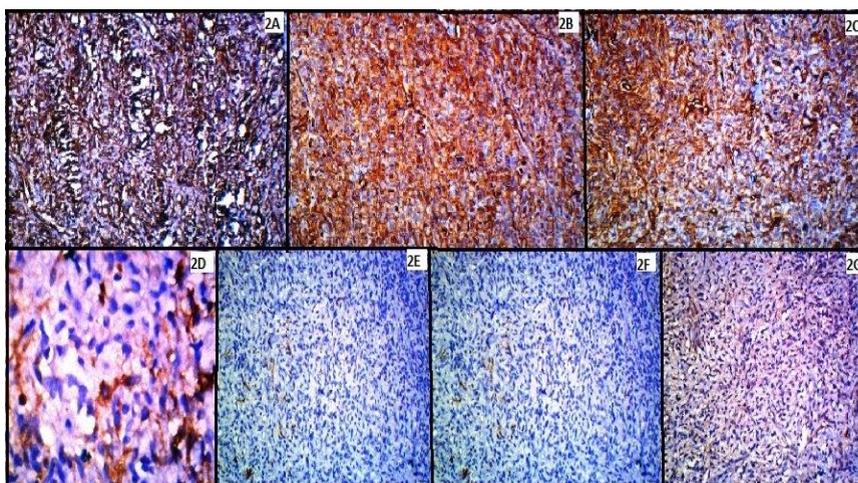


Fig. 2: Showing results of immunohistochemical markers applied on tissue sections (a) Vimentin positive; (b) BCL2 positive; (c) CD34 positive; (d)Ki67 proliferating index <10%; (e) S100 negative; (f) Desmin negative; (g) VEGF negative

Discussion

SFTs are a rare heterogeneous group of spindle cell neoplasms that include benign as well as malignant neoplasms. The cell of origin of SFTs has been the subject of debate since its conception. In 1931 Klemperer and Rabin first documented the occurrence of a distinctive localized pleural based tumour and proposed a submesothelial cell origin.⁽³⁾ Later, Stout and Murray carried out tissue culture experiments and claimed their derivation from mesothelial cells.⁽⁴⁾ SFTs have been known by a number of different names in the past viz localized fibrous tumor, localized fibrous mesothelioma, solitary fibrous mesothelioma, fibrous mesothelioma, subserosal fibroma and submesothelial fibroma because of its controversial cell of origin. With the advent of immunohistochemistry, it was further stated that SFTs have a fibroblastic origin, with

myofibroblastic differentiation at times. This formed the basis of hypothesis regarding occurrence of SFTs in extrapleural sites that are devoid of mesothelial cells.⁽⁵⁾ SFTs at various sites are associated with different epidemiological characteristics like age, sex predilection and incidence although gross and microscopic features remain the same at all the sites. The presenting symptoms are not specific. Soft tissue SFT represents <2% of all soft tissue tumors.

The vast majority of SFT are covered by a smooth glistening capsule. Most of the tumors are composed of uniform collagen forming spindle shaped cells, arranged in interlacing fascicles. Associated myxoid change, fibrosis, hyalinization, necrosis or rarely focal calcifications may be observed.⁽⁶⁾ The cells are negative for Cytokeratins and Epithelial membrane antigen, but stain in all cases with Vimentin and CD34, rarely with

muscle specific actin. The nonspecific morphological pattern (patternless pattern) and lack of distinctive immunohistochemical or electron microscopic features, makes it difficult to separate SFT from other spindle shaped neoplasms on routine staining alone.^(6,7) Cytogenetic and molecular studies do not have any additional role in making its diagnosis.

Although SFTs have been reported in various extrathoracic sites, its occurrence in the oral cavity is extremely rare and to the best of our knowledge, only 9 cases in tongue have been reported so far in English literature.⁽⁸⁻¹⁰⁾ There are no well defined criteria for labeling malignancy in SFT. However, infiltration margins, high mitotic counts (>4/HPF), pleomorphism and tumor necrosis have been reported in malignant forms.⁽¹¹⁾

Diagnosis in almost all the cases has been based on clinical, microscopic and immunohistochemical findings. In our case, the tumor showed CD 34, Vimentin and BCL-2 positivity which is similar to all the previously reported cases. The Ki-67 IHC revealed labeling index <10 % (i.e. having low proliferating activity) which is similar to the results of previous studies.⁽⁸⁻¹⁰⁾ Desmin and S100 were uniformly negative in all the previously reported cases as in the present case also.

Conclusion

Although rare but SFTs should be considered in the differential diagnosis of soft tissue tumors in oral cavity based on clinical and microscopic features and confirmed by immunohistochemical studies. Most SFTs behave in a benign fashion and recurrence is rare. Surgical excision with wide margin is the treatment of choice. However, their behavior is unpredictable, so long term follow up is mandatory.

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