Case Report

The first case of proliferative fasciitis of tongue coexistent with squamous cell carcinoma: Case report of a rare lesion

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Abstract

Proliferative fasciitis (PF) is a rare pseudosarcomatous myofibroblastic benign tumor, a subcutaneous counterpart of proliferative myositis. Usually seen in upper extremities, no case has yet been documented in tongue or any other subsites in oral cavity. The present case becomes the first to be reported at this site as well as the first case of synchronous coexistent PF with squamous cell carcinoma (SCC) of tongue. The patient was 50 years male, having a polypoidal swelling at right lateral border of tongue with an ulcer adjacent to it. Histopathologically, the swelling was diagnosed as PF and ulcer as SCC; both the diagnoses were confirmed by immunohistochemistry. The polypoidal lesion was immunopositive for smooth muscle actin and calponin and immunonegative for pan cytokeratin, cytokeratins (5/6), P40 and P63, proving it to be a non-SCC lesion, different from its adjacent ulcerative one.

Keywords: Fasciitis, fibroblasts, myofibroblasts, proliferative fasciitis, proliferative myositis

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INTRODUCTION

Proliferative fasciitis (PF) is a very rare myofibroblastic proliferation – a benign tumor which is a mimic of sarcomas or malignancies.[1,2] It is considered a subcutaneous counterpart of proliferative myositis (PM), only uncommonly found in head, neck and face (HNF) region. After a review of English literature, the present case becomes the first documented case of PF in tongue (and oral cavity) and first reported concurrent PF with squamous cell carcinoma (SCC) in oral cavity and tongue. Only second primary SCC and a few granular cell tumor cases have been documented as synchronous tumors to SCC in oral cavity.[3-6]

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CASE REPORT

A 50-year-old male patient presented to Otolaryngology (ENT) Outpatient Department (OPD) with complaints of nonhealing ulcer over tongue. The patient, farmer by profession, was a bidi smoker. There was no history of any chronic condition. He had noticed a small ulcer over right lateral border of tongue 5–6 months ago, for which he was taking home remedies and alternative medicine, but without any healing. In the past month, he noticed a rapid increase in the size of the ulcer, and the lesion had become painful for last 2–3 weeks. This prompted him to seek medical treatment at a higher center, and hence he approached the

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ENT OPD of this hospital. Clinical examination revealed a roughly 3 cm × 3 cm ulceroproliferative lesion with a tender polypoidal lesion approximately 2 cm × 1.5 cm in size, and the mobility of tongue was restricted in the form that the patient was unable to protrude the tongue out of mouth [Figure 1]. One right level lb, lymph node was palpable, rest clinical examination was insignificant. Incisional biopsy of the ulcerative lesion and excisional biopsy of polypoidal one were performed under local anesthesia, and specimen was sent for histopathological examination.

Two biopsies were received at the Pathology Department. The first one (the excision biopsy of the polypoidal lesion) measured 2 cm × 1.5 cm × 0.5 cm, partly ulcerated mucosa externally, cut section solid, firm and grey-white. Histologically, it showed a tumor composed of plump fibroblastic and myofibroblastic spindle cells with bland cytological features [Figure 2], collagenous to myxoid, predominantly collagenous stroma [Figure 3a and b]. Furthermore, large ganglion-like cells were also seen. These characteristic cells had abundant basophilic cytoplasm, one or two vesicular nuclei [Figure 3c] with single prominent nucleoli. Few large lobulated nuclei in these cells were also observed [Figure 3d]. The clustering of ganglionic cells around the blood vessels was noted. Some mitoses (2–4 per 10 high-power fields) in both spindle and ganglionic cells [Figure 3e], plenty of acute inflammatory infiltrate and branching blood vessels were also present. Immunohistochemistry (IHC) showed immunopositivity for smooth muscle actin (SMA) and calponin [Figure 4]. Tumor cells were typically immunonegative for cytokeratins (CK 5/6), P40 and P63, thus confirming the diagnosis. The other biopsy showed the histopathological and IHC features of SCC [Figures 2 and 4]

The clinical stage of this tongue SCC was T2 N1 M0.

The patient was advised treatment in the form of wide local excision of lesion with neck dissection, followed by concurrent chemotherapy/radiotherapy depending on final postoperative histopathology report with follow-up once every 3 months for 1st year first. The preoperative workup was done, including the preanesthetic checkup, and computerized tomography neck, thorax and abdomen to rule out metastasis to (e.g., neck lymph nodes) or from any other site (e.g., lung). No other lesion was found proving the SCC to be primary to tongue. Thus, it became Stage 2 (T2 N0 M0) as the neck lymph node was reactive on radiology. Furthermore, no subcutaneous lesions were found on upper extremity, lower extremity, trunk or head

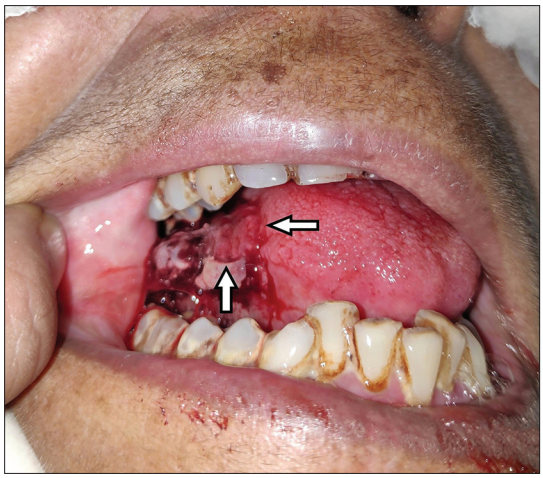

Figure 1: An ulceroproliferative lesion over right lateral border of tongue with a polypoidal lesion at the upper end of the ulcer (arrow)

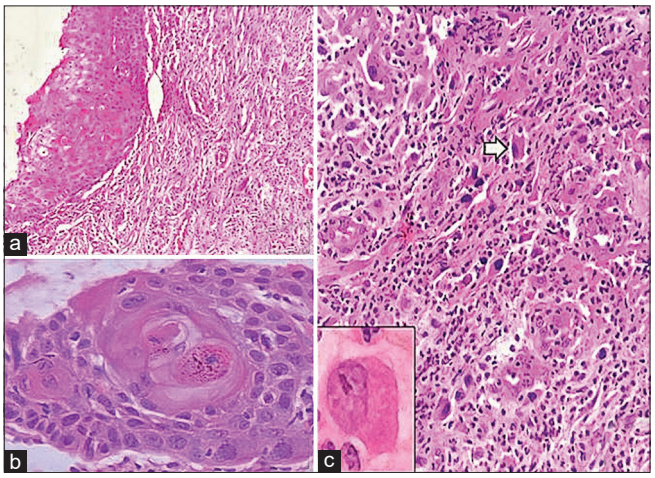

Figure 2: (a) Squamous cell carcinoma (SCC) with adjacent proliferative fasciitis (PF) (H&E, ×40), (b) SCC, other areas, showing keratin pearl and invasion (H&E, ×400), (c) PF showing fibroblastic and ganglionic (arrow and inset) spindle cells, and acute inflammatory infiltrate (H&E, ×100)

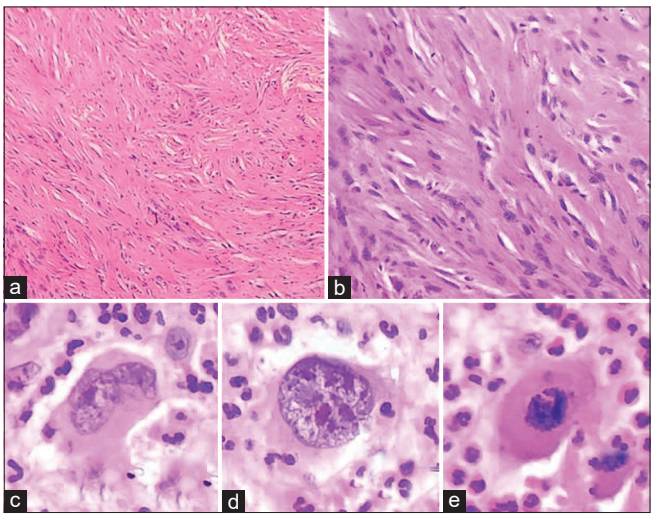

Figure 3: Collagenous stroma in proliferative fasciitis ((a) H&E, ×40, (b) H&E, ×100). Nuclear features of ganglionic cells – Binucleation (c), large and lobulated (d) and mitosis (e) (H&E, ×400)

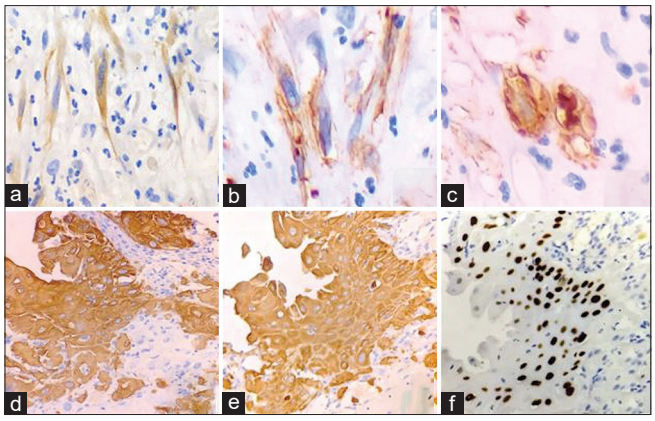

Figure 4: Immunohistochemistry – Calponin (a) and smooth muscle actin (SMA) (b) positivity in the fibroblastic spindle cells in proliferative fasciitis (PF) (H&E, ×100). SMA positive ganglionic cells in PF ([c] H&E, ×400). Pancytokeratin (d), Cytokeratin CK5/6 (e) and p63 (f) positivity in adjacent squamous cell carcinoma (H&E, ×100)

and neck clinically or radiologically. This indicated primary nature of the PF tongue as well.

Fearing surgery and disfigurement, the patient refused the surgical management and went to try conservative treatment by alternative medicine.

DISCUSSION

PF is a rare pseudosarcomatous benign soft tissue lesion, considered subcutaneous counterpart of PM. Chung and Enzinger established sound morphological criteria for the diagnosis of PF while first reporting 53 cases in 1975, delineating it from nodular fasciitis (NF) and PM.[1] Earlier in 1960, PM was first described by Kern formally, and characterized further by Enzinger and Dulcey in 1967.[7,8]

PF has not been reported earlier in oral cavity. In HNF region as well only 11 cases have been documented.[9] Furthermore, till now, only three cases of PM of tongue have been described, one of them at lateral border.[10-12]

Oral cavity is one of the common sites for field cancerization, where there is occurrence of second or multiple primary malignancies (SCC). Only a few cases of synchronous second tumor with SCC tongue other than another primary SCC have been reported, which was granular cell tumor,[3-6] few of them with both tumors at same site are also described.[4-6] However, the present case becomes first synchronous coexistent PF with SCC tongue.

PF is benign proliferation of myofibroblasts along with PM and NF. PF is subcutaneous whereas PM is intramuscular, both with same histology. PF microscopically resembles NF with additional ganglionic cells.[13] All of them are considered pseudosarcomatous due to their rapid growth rate and histological features.

The usual age for affection by PF is 40–70 years with a roughly equal sex distribution.[9] A pediatric subtype is also described. It involves most commonly the extremities and usually forearm.[13] The lesion is nodular, firm, palpable subcutaneously, freely movable as is unattached to the overlying skin. Majority of cases have complaints of pain or tenderness, the exact reason being unknown. However, in the present case, it could have been due to the adjacent malignant ulcer. They are usually <5 cm in size.

Radiological features are not specific or characteristic. This makes histopathological examination essential tool for confirmatory diagnosis in these cases. Histologically, a biphasic cell population of proliferating spindle myofibroblasts like tissue culture and characteristic ganglion-like cells is present.[13] The later cells are large, polygonal, with abundant basophilic-to-amphophilic cytoplasm, large vesicular one or two nuclei, prominent nucleoli. The present case also showed few large and some lobulated nuclei in the ganglionic cells. Variable but typical mitosis in both cell types may be found. Multinucleated giant cells are rare, unlike NF. The stromal component varies from myxoid to collagenous. Pediatric cases have acute inflammatory infiltrate, necrosis in addition to higher cellularity and mitotic rate, and more solid growth pattern.[14] These features are distinctly unusual for the adult tumors. However, the presence of ulcerated surface or adjacent ulceroproliferative SCC might have been the cause for plenty of neutrophils seen in the present case.

Differential diagnosis of adult cases includes NF, PM, inflammatory myofibroblastic tumor (IMT) and some malignancies; and of pediatric PF includes rhabdomyosarcoma (RMS), epithelioid sarcoma (ES) and other malignancies.[14] In the past, PF has been misinterpreted as embryonal RMS, ganglioneuroblastoma and others.[13] The cross striations of rhabdomyoblasts and IHC help to rule out RMS.[15] The nuclear atypia of the spindle cell component noted in RMS is also absent in PF.[13] ES is cytokeratin positive unlike PF. Ganglioneuroblastoma cells are reactive to neural markers including S100 protein in contrast to PF. There is prominent and more conspicuous inflammatory component consisting of plasma cells and lymphocytes in the inflammatory myofibroblastic tumor along with zonality consisting of superficial (submucosal) myxoid zones juxtaposed to deep cellular zones; whereas inflammatory component if seen in PF (as in pediatric cases) is composed of acute inflammatory infiltrate, i.e.,

neutrophils. In most cases, IMT shows diffuse cytokeratin positivity in contrast to PF.

A low-grade malignant tumor myofibroblastic sarcoma has been added to the WHO classification of the oral cavity and mobile tongue (2017 fourth edition), soft tissue and neural tumors. Oral cavity including tongue is known to be the preferred site. Small size of the polypoidal lesion, the absence of the nuclear atypia, pleomorphism of the spindle cells and atypical mitosis were the features notwithstanding with the diagnosis of myofibroblastic sarcoma.[16]

In IHC of PF the spindle-shaped cells stain positive for SMA and muscle specific actin. CK, desmin, myogenin, β-catenin and S-100 protein immunostains are usually negative. The ganglion-like cells are often negative for SMA. This profile is similar to those of NF and PM.[13] Calponin stains the myofibroblasts. The present case was even immunonegative for CK (5/6), P40 and P63, proving it to be a non-SCC lesion, different from its adjacent ulcerative SCC.

Antecedent injury plays a probable role in the pathogenesis of PF, which could have been the possible cause at this site also. Recently, adult cases have been shown to harbor FOS gene rearrangement in the ganglionic cells, not in spindle cell component. All PF/PMs lack USP6 rearrangement, unlike NF.

The treatment and prognosis in the present case would be dictated by SCC tongue. Complete excision in solitary PF cases is known to be curative with only rare recurrences. Instead, the adjacent malignant lesion warrants large resection with neck dissection here. The T2 SCC tongue has >60% of 5-year survival.

CONCLUSION

PF is rare in HNF, with the present case being first in tongue and oral cavity. It should be included as a differential diagnosis of polypoidal lesions of oral cavity. As it is difficult to distinguish from some malignancies based on clinical or radiological findings, histopathological examination and if required IHC becomes mandatory to make a correct diagnosis.

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 initial s 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.

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