Intravascular fasciitis: report of two intraoral cases and review of the literature

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Two unusual cases of intravascular fasciitis arising in a 25-year-old female and a 26-year-old male are presented here. The lesions apparently presented as firm, raised, submucosal nodules on the tongue. Intravascular fasciitis (IVF) shares the microscopic features of nodular fasciitis (NF), but with intraluminal, intramural, and extramural involvement of small- to medium-sized veins and arteries with a multinodular or serpentine growth pattern along the course of affected blood vessels. NF is a benign lesion occurring on the trunk and upper extremities with a strong predilection for young adults 20 to 40 years of age. Intraoral NF is uncommon, and intraoral IVF is extremely rare, with only sporadic reports in the literature. In both of our cases, the patient's main concern was rapid growth of the lesion, which was nontender, on the tongue. The clinical, histologic, and immunohistochemical features and treatment are presented, along with a review of the literature. (Oral Surg Oral Med Oral Pathol Oral Radiol 2016;121:e19-e25)

Nodular fasciitis (NF), also known as pseudosarcomatous fasciitis, is a common benign, self-limiting, reactive process composed of fibroblasts and myofibroblasts usually arising from subcutaneous tissues or muscle fascia.[1-3] NF is commonly misdiagnosed as a sarcoma because of rapid growth, rich cellularity, and the presence of mitotic figures.[1,3] Although NF is considered a benign process, the etiology is unknown. NF occurs more frequently on the upper extremities (>40%) followed by the head and neck region (7%-20%), and trunk area (12%-18%).[1,4] NF occurs equally in men and women and can occur over a wide age range, with the majority of cases diagnosed in the second to fourth decades. Rare morphologic subtypes of NF include ossifying fasciitis, cranial fasciitis, and intravascular fasciitis (IVF).[1] These subtypes differ clinically, grossly, and microscopically. Ossifying fasciitis is seen usually in females between the ages of 20 and 30 years and usually involves the upper and lower extremities. Cranial fasciitis is seen usually in infants during the first year of life. It involves the soft tissues of the scalp and can cause a lytic defect of the underlying skull.[1] IVF was first described by Patchefsky and Enzinger in 1981.[5] IVF is a rare and distinct variant of NF, with only six intraoral cases reported to date. We report two additional cases of IVF, one occurring in a 25-year-old female and the other in a 26-year-old male, and also discuss the histologic and immunohistochemical features of this lesion. In addition, we summarize the cases reported in the literature.

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CASE REPORT

The Temple University School of Medicine's Institutional Review Board approved these case reports.

Case 1

A 25-year-old female presented to an oral and maxillofacial surgeon for consultation of a rapidly enlarging swelling involving the left ventral surface of the tongue and the floor of the mouth of unknown duration. Clinical examination revealed a 1.0 × 0.6-cm, firm, multinodular, submucosal mass extending across the left floor of the mouth with a serpiginous tail-like extension going into the ventral surface of tongue superiorly (Figure 1). The lesion was mobile, firm to palpation, and nontender. There was no history of trauma to the area. The patient's medical history was positive for asthma, anxiety, and depression. A hemangioma in the trachea had been removed at birth. Medications included escitalopram (Lexapro), ethinyl estradiol, and levonorgestrel (Levora) on a daily basis, and albuterol inhaler. The patient had allergies to fentanyl and succinylcholine, which caused anaphylaxis. The differential diagnosis was a minor salivary gland neoplasm, a thickened blood vessel, or a mesenchymal neoplasm. Following anesthesia, using 2% xylocaine with epinephrine 1:100,000 by nerve block and 4% articaine with epinephrine 1:200,000 by local infiltration, surgical excision of the lesion in the left ventral tongue and sublingual area was done using sharp and blunt dissection. The deep aspect of the lesion was wrapped around the submandibular duct; the surgeon dissected as much of the lesion as possible without transecting the duct, and the tissue was submitted for microscopic examination.

Case 2

A healthy 26-year-old male presented to an oral and maxillofacial surgeon with the chief complaint of a rapidly growing mass on the dorsal surface of the tongue of 1 week's duration (Figure 2). There was no history of trauma, and the patient denied pain or numbness. On examination, a 1-cm, submucosal, slightly raised mass was found in the left mid-dorsal tongue with central ulceration because of secondary mechanical trauma caused by rapid growth. The patient's medical history was noncontributory. He took no medications

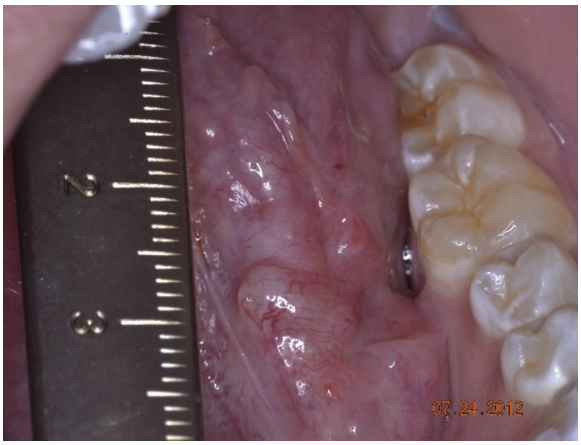

Fig. 1. Multinodular lesion with serpentine growth pattern on the left ventral surface of the tongue and the floor of the mouth.

on a daily basis and had no known drug allergies. He denied tobacco use and reported social alcohol use. His past surgical history was Lasik surgery in 2004. The clinical impression was a granular cell tumor or a thickened blood vessel. Following local anesthesia with 2% lidocaine and 1:100,000 epinephrine, the lesion was sharply dissected from underlying skeletal muscle. A central stalk that appeared to be a thickened blood vessel was identified; it was tied off with 3-0 Vicryl and transected using electrocautery. The specimen was submitted for microscopic examination.

Microscopic examination

Microscopic examination of the specimen from Case 1 revealed a well-circumscribed nodular growth of fibroblasts and myofibroblasts in a collagenous stroma in large areas (Figure 3A). The lesion appeared elongated and serpiginous and varied in thickness. The thinner portion of the lesion was more collagenous, well encapsulated, and separated from the capsule and infiltrating thin-walled vein, consistent with an intravascular lesion (Figure 3B). The larger thicker portion of the lesion was nonencapsulated and located beneath the mucosa with scattered inflammatory cells (Figure 3C) and lesional cells with open vesicular nuclei with some scattered mitotic figures (Figure 3D). No giant cells were noted. The microscopic differential diagnosis was limited and included NF (including IVF), gastrointestinal stromal tumors, solitary fibrous tumor, schwannoma, and myofibroblastic tumor.

Immunohistochemically, the cells in the lesion and the blood vessel wall were diffusely positive for vimentin, smooth muscle actin (SMA), and muscle-specific actin (MSA) (Figures 4A, 4B, and 4D). Ki-67 staining revealed a low proliferative index (<5%). CD34 staining highlights both the intralesional vessels and blood vessel wall surrounding the lesional tissue (Figure 4C). C-kit, DOG1, S100, and GFAP stains were negative; thus, gastrointestinal stromal tumor and schwannoma were excluded. Desmin and AE1/3 were negative, and elastic stains revealed fragmented residual elastic fibers (Figure 5A), thus rendering a diagnosis of intravascular fasciitis based on morphology and immunohistochemistry. Complete excision of the remaining lesional tissue was recommended. To date (2 years and 4 months postoperatively), the patient is doing well, with no recurrence or complications.

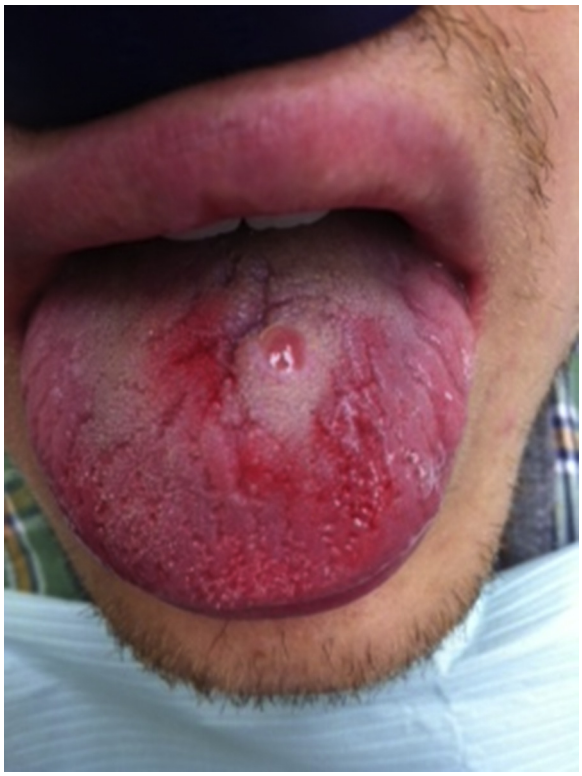

Fig. 2. Intraoral view from Case 2 reveals raised lesion on the dorsal surface of the tongue.

Microscopic examination of the specimen from Case 2 revealed a well-circumscribed mass with mainly extravascular and smaller areas of lesional tissue present between bundles of skeletal muscle with slitlike vessels (Figure 6A). Higher magnification of the spindle cells in a vessel is shown in Figure 6B. The lesion was ulcerated with exuberant growth of SMA and vimentin-positive myofibroblasts (Figure 6C). CD34 highlights the vessel wall (Figure 6D). Desmin, S100, and AE 1/3 were negative. Ki-67 staining revealed a low proliferative index (<10%). Elastic stains reveal fragmented residual elastic fibers (Figure 5B). Thus a diagnosis of myofibroblastic proliferation, consistent with intravascular fasciitis, was rendered. Excision was incomplete because lesional tissue extended into one margin. Sixteen days after surgery, the patient presented with a small, firm, ulcerated mass at the previous surgical site. The lesion was excised under local anesthesia and the tissue submitted for histologic evaluation. The microscopic features of the lesion were similar to the original lesion, with completed excision noted. To date (1 year and 5 months postoperatively), the patient is doing well, with no complications or recurrence.

DISCUSSION

The term “fasciitis” implies that the lesion originates in the muscle fascia and that it is of an inflammatory