STAT6 Reliably Distinguishes Solitary Fibrous Tumors from Myofibromas

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Abstract Solitary fibrous tumors (SFT) and myofibromas (MF) historically have belonged to the same morphologic spectrum and have been lumped together under the nonspecific umbrella term, “hemangiopericytoma” along with other pericytic/myoid tumors. While current evidence shows clear distinction between the two entities, they frequently remain in the same histopathologic differential diagnosis. This diagnostic dilemma especially is common for smaller incisional biopsies from the oral cavity. STAT6 immunohistochemistry (IHC) recently was established as a reliable method to detect solitary fibrous tumor; however, the literature is sparse regarding STAT6 reactivity in MFs. The authors report ten new cases of oral solitary fibrous tumor, discuss histopathologic similarities and differences between the two tumors, and list respective STAT6 IHC expressivity. After IRB approval, 10 cases diagnosed as SFT and 24 cases of MF were collected from the University of Florida Oral and Maxillofacial Pathology Biopsy Service between the years 1994 and 2016. The original hematoxylin and eosin slides and related IHC were reviewed. IHC with STAT6 antibody was performed on all 34 samples, and the findings were analyzed. All cases were from the oral cavity or perioral regions. 10/10 SFTs expressed STAT6 nuclear reactivity, while no cases of MF showed nuclear expression of STAT6. STAT6 is a dependable marker to differentiate SFTs from MFs.

Keywords STAT6 · Myofibroma · Solitary fibrous tumor · Hemangiopericytoma · Immunohistochemistry

Introduction

Solitary fibrous tumors (SFT) is an uncommon mesenchymal neoplasm with a variable histomorphologic spectrum. It was originally described in the pleura by Wagner in 1870 [1], but was not described as a distinct histopathologic entity until 1931 by Klemperer and Rabin [2]. In 1942, Stout and Murray introduced the term “hemangiopericytoma” (HPC) to designate a soft tissue tumor composed of endothelial cells that lacked the organoid features of glomus tumor [3]. The authors proposed that SFTs originated from Zimmerman’s pericytes [3]. SFT likely fell under the HPC category for decades. Finally, in the 2013 WHO Classification of Bone and Soft Tissue Tumors, SFT was considered to be of fibroblastic/myofibroblastic origin, and “hemangiopericytoma” was termed obsolete, as many tumors formally labeled HPC had been more specifically delineated [4].

Myofibromas (MF), another tumor characterized by decades of confusing names and unproven etiologies, is also an entity with uncertain histogenesis that has been included under the term HPC, most commonly as “infantile hemangiopericytoma.” It is currently considered a perivascular myoid neoplasm, along the same morphologic spectrum as myopericytoma [4]. SFT and MF often have overlapping histopathologic features, especially in small biopsy specimens. While differing immunohistochemical profiles of these neoplasms should resolve the issue, the results may not always be straightforward.

In 2013, SFTs were found to harbor recurrent genetic alterations involving chromosome 12q13, forming the

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NGFI-A binding protein 2 (NAB2)/STAT6 fusion gene [5–7]. Multiple researchers have confirmed that nuclear STAT6 immunohistochemical reactivity is highly specific for solitary fibrous tumor [8–13]; however, little remains known about the expressivity of STAT6 antibody in myofibromas.

The authors aim to report ten new cases of oral SFT and compare and contrast features of SFTs and MFs, including STAT6 expressivity in both.

Materials and Methods

All cases diagnosed as “solitary fibrous tumor” ($n = 10$) and “myofibroma” ($n = 24$) between the years 1994 and 2016 were culled from the University of Florida Oral and Maxillofacial Pathology Biopsy Service. The original hematoxylin and eosin (H&E)-stained slides as well as existing IHC slides were reviewed. Because the diagnoses made based on H&E and pre-existing IHC were unequivocal, no stains were repeated for diagnostic purposes of this study. All SFTs were reported by the clinicians as excisional, whole-tissue specimens, while 19/24 MFs were excisional biopsies. The remaining four cases were incisional biopsies. Diagnoses were established on current standard histopathological and IHC criteria provided by the World Health Organization (WHO) Classification of Soft Tissue Tumors [4].

Four-micron sections from paraffin blocks were made of each sample and mounted onto slides. IHC staining for anti-STAT6 antibody (1:100 dilution, Santa Cruz Biotechnology; Santa Cruz, CA, USA) was performed on all sections using manufacturer’s instructions. The quality of expression was graded and interpreted based on absent staining/non-specific blush, weak expression, or strong expression. The samples also were graded based on

diffuse ($>50\%$ of tumor cells) versus localized ($<50\%$ of tumor cells) staining, as well as localization of the stain within the cell (nuclear only, cytoplasmic only, or both nuclear and cytoplasmic).

Approval for the acquisition of tissue specimens and supporting case information, as well as permission to perform immunohistochemical studies, was obtained from the Institutional Review Board (IRB) of the University of Florida.

Results

Thirty-four total cases of SFTs and MFs were retrieved from the biopsy service: 10 SFTs and 24 MFs. The average age of diagnosis for the ten new SFT cases was 55.1 years with a range of 23–86 years and a male predilection (8/10). Four cases were obtained from the buccal mucosa, three from the labial mucosa, and one each from the floor of mouth, submandibular region, and gingiva (Table 1).

All ten SFTs were CD34 positive, while all other previously performed immunohistochemical stains were unreactive. STAT6 antibody nuclear reactivity was diffusely strong for 9/10 cases (Fig. 5a), while one case (Case #3, Fig. 6) exhibited weak and focal nuclear reactivity (Fig. 5b).

Case specifics for 20 of our 24 MFs have been reported previously in the literature (Smith et al. [14], cases 1–11, 13, 15–17, 19–21, 23, 24 are cases 1–20 in the present paper). Distinct nuclear positivity for STAT6 was lacking in all 24 MFs. Weak, cytoplasmic reactivity in one MF case was found (Fig. 7). Previously performed IHC results for MF cases are reported in Table 2. STAT6 results for both tumors are summarized in Table 3.

Table 1 Clinical and immunohistochemical properties of 10 oral SFTs

SFT	Age	Gender	Location	Previous IHC
1	81	M	Inferior border of mandible; right submandibular area	CD34+, S-100-, α SMA-, desmin-
2	57	M	Buccal mucosa	CD34+, S-100-, CD68-, α SMA-
3	23	M	Anterior floor of mouth	CD34+, S-100-, α SMA-
4	41	F	Buccal mucosa	CD34+, S-100-, α SMA-
5	56	M	Upper labial mucosa	CD34+, S-100-, p63-, GFAP-, α SMA-, EMA-
6	47	M	Buccal mucosa	CD34+, S-100-
7	86	M	Upper labial mucosa	CD34+, S-100-, keratins-
8	44	F	Lower labial mucosa	CD34+
9	57	M	Buccal mucosa	CD34+, S-100-
10	59	M	Anterior mandibular gingiva	CD34+, α SMA-

M male, F female, α SMA alpha-smooth muscle actin, GFAP glial fibrillary acidic protein

Table 2 Previously performed IHC results for 24 cases of MF; Cases 1–20 have been published prior in the literature [14]

MF	Previous IHC
1	αSMA+, desmin-, S100-, focal calponin+
2	αSMA+, desmin-, vimentin+, CD34-, S100-
3	αSMA+, desmin-, vimentin +, S100-
4	αSMA+, MSA+, desmin-, S100-, CD34-
5	αSMA+, MSA+, desmin-, vimentin +, S100-
6	αSMA+, desmin-, S100-
7	αSMA+, desmin-, S100-
8	αSMA+, desmin-, S100-, CD34-
9	αSMA+, MSA+, desmin-, H-caldesmon-, S100-
10	αSMA+, MSA+, desmin-, vimentin+, AE1/3-, S100-
11	αSMA+, desmin-, CD68-, AE1/3-, Cam 5.2-, S100-
12	αSMA+, MSA+, desmin-, vimentin +, S100-
13	αSMA+, desmin-, S100-, CD34-
14	αSMA+, desmin-, S100-, CD68-
15	αSMA+, MSA-, desmin-, S100-
16	αSMA+, desmin-, CD34-, S100-, CD68-
17	αSMA+, desmin-, CD34-, CD68-
18	αSMA+, MSA+, desmin-, vimentin +, S100-, Factor 8-, CD68-
19	αSMA+, desmin-, S100-, CD34-
20	αSMA+, MSA+, desmin-, vimentin+
21	αSMA+, MSA+, desmin-, CD68-, CD34-, S100-
22	αSMA+, desmin-, S100-, CD68-
23	αSMA+, desmin-, S100-
24	αSMA+, S100-

Table 3 STAT6 Comparative Immunohistochemistry, anti-STAT6 antibody, 1:100 dilution, Santa Cruz Biotechnology; Santa Cruz, CA, USA

Tumor type	Positive				Negative
	Strong nuclear	Weak nuclear	Strong cytoplasmic	Weak cytoplasmic	
SFT	9/10	1/10	0/10	0/10	0/10
MF	0/10	0/10	0/10	1/24	23/24

SFT solitary fibrous tumor, *MF* myofibroma

Occasional non-neoplastic structures present in our stained sections demonstrated weak cytoplasmic STAT6 staining, including endothelial and ductal cells.

Both SFT and MF demonstrated significant histopathologic variability within each tumor, even from field-to-field. Common features (even in focal areas) included circumscription, alternating hypo- and hypercellular zones (Fig. 1a, d), variable areas of collagenized or hyalinized stroma (ropey collagen) (Fig. 1b, e), “staghorn” (also referred to as “HPC-like”) vessels (Fig. 1c, f), giant cells

(Fig. 2a, d), perinuclear clearing (Fig. 1b, e), spindle to ovoid cells with indistinct cell borders, and myxoid stroma. Even dense, “smudgy,” hyalinized areas (sometimes referred to as pseudocondroid areas), which is a classic feature of MFs, could be distinguished in certain foci of SFTs (Fig. 2c, f). It should be noted that while both tumors exhibited variable cellularity, the hypocellular zones in SFT demonstrated a loose, mucoid-appearing background, while the hypocellular areas in MF were more solid or hyalinized.

Significant differences were noted in that no cases of MF exhibited prominent perivascular hyalinization, which was seen in 6/10 SFT cases (Fig. 3). No cases of SFT exhibited infiltrative margins nor a lobular architecture (“myoid ball” structures) (Fig. 4), as seen in some MFs. Additionally, while many MFs were well circumscribed, none exhibited a full or partial fibrous capsule, while 7/10 SFTs exhibited at least a partial capsule, which is helpful when evaluating whole tissue specimens. The “patternless pattern” typically observed in SFT was present in the majority of our SFT cases, but it was not diffusely observed in all cases, particularly case #3 (Fig. 6) where the pattern was predominantly fascicular. The “patternless pattern” was not overtly observed in the MFs.

Due to IRB restrictions, no clinical follow-up information was obtained for any of our cases.

Discussion

The cell of origin for SFT has long been debated. Since its description as an entity in 1931, proposals for cell origin have included mesothelial, serosal, submesothelial, pericytic, and fibroblastic. It is now generally agreed upon that the cell type is fibroblastic [4, 15, 16], although it may demonstrate myofibroblastic differentiation or contain more primitive mesenchymal cells which lack fibroblastic features [15–19]. This potential for myofibroblastic differentiation adds to the confusion between SFTs and MFs. Furthermore, while CD34 reactivity is one of the hallmarks for SFT, 5–10% are reported to be negative, and 20–35% of tumors may show positivity for alpha-smooth muscle actin (αSMA) or epithelial membrane antigen (EMA) [1, 4]. Rare cases of CD34 positivity in MF also have been reported [20]. While CD99 and Bcl-2 are helpful in diagnosis of SFT, they are non-specific, and tumors in its differential diagnosis may also demonstrate positivity to the same markers [21]. These conflicting IHC findings along with the all-encompassing historical concept of HPC may lead to diagnostic confusion between the two entities.

Since the discovery of NAB2/STAT6’s role in SFT tumorigenesis [5–7] and the subsequent advent of STAT6 immunohistochemical marker, SFTs are more easily

Fig. 1 Histopathologic similarities between solitary fibrous tumors and myofibromas (*white arrow* hypocellular region, *black arrow* hypercellular region)

Fig. 2 Histopathologic similarities between solitary fibrous tumors and myofibromas, continued

Fig. 3 Prominent perivascular hyalinization noted in SFT case #1, 10×

Fig. 4 Lobular and infiltrating architecture of MF, with structures often referred to as "myoid balls," 10×

differentiated from other soft tissue tumors. While multiple researchers have reported the sensitivity and specificity of STAT6 expression in SFTs and its histopathologic mimickers [8–13], we were only able to find information about its expressivity in MFs for five cases in the literature: one case in a large series by Demicco et al. (which reported negative STAT6 expressivity) [8] and four cases in a study by Yoshida et al. [13]. In the four MF cases published by Yoshida, the authors reported that 2/4 demonstrated cytoplasmic and nuclear staining of STAT6 antibody. They explained that both nuclear and cytoplasmic reactivity was seen in 74/155 of their non-SFTs, and that this pattern of staining was different from the pure nuclear staining found in SFTs; however, this staining pattern found in 50% of their MF cases, combined with the paucity of STAT6 results in MF in the literature led the authors to question whether STAT6 truly is a valuable marker to distinguish the two tumors. Additionally, Demicco et al. [8] reported that 4/17 cases of scar tissue expressed nuclear STAT6 antibody. Because trauma vaguely has been associated with MF [14], STAT6 expression in scar tissue further begs the question whether MFs may demonstrate STAT6 expression, as well.

Some of the histopathologic overlap between SFTs and MFs includes the presence of ovoid to spindled cells, "staghorn"-like vessels, variable areas of collagenized or hyalinized stroma (also known as ropey collagen, keloid-like collagen, or amianthoid fibers), multinucleated giant cells, perinuclear clearing, densely hyalinized areas (also known as pseudochondroid areas) and mixed hyper- and paucicellular zones. However, none of our MF cases demonstrated distinct perivascular hyalinization, as seen in 6/10 of our SFTs (Fig. 3). Another feature unique in our series was the presence of encapsulation; none of the MFs demonstrated a complete or partial capsule, while 7/10 of the SFTs did. The SFTs also did not demonstrate

Fig. 5 **a** Strong nuclear STAT6 antibody reactivity from SFT case #2, 40×. **b** Weak nuclear STAT6 reactivity in SFT case #3, 40×

Fig. 6 SFT, case #3 **a** Low power image demonstrating circumscription and partial encapsulation of the tumor. **b** Spindled-to-ovoid cells are arranged in a fascicular pattern, 9×. **c** Positive reactivity for CD34, 10×. **d** Clinical image of tumor in the anterior floor of mouth

Fig. 7 Weak cytoplasmic STAT6 antibody reactivity in one MF, 20× (**a**) 40× (**b**)

infiltrative margins nor lobular architecture, as is sometimes seen in MFs. Because the histopathologic presentation of both of these tumors often varies from field-to-field, distinguishing between them on histopathologic

patterns alone can prove difficult, especially on small, incisional biopsies.

All ten of our SFTs demonstrated STAT6 nuclear expression. Nine cases showed strong, diffuse nuclear

expressivity (Fig. 5a), while one case demonstrated weak and focal nuclear staining (Case #3) (Fig. 5b). The latter case demonstrated classic histopathologic features of a cellular SFT: variable hypo- and hypercellular zones, partial encapsulation, and myxoid stroma with round to ovoid nuclei arranged in a predominantly fascicular arrangement (Fig. 6a, b). The tumor exhibited strong CD34 positivity (Fig. 6c). It was only unusual in that the floor of the mouth is a rare location for SFT, with less than five cases reported in the literature to date (Fig. 6d) [22–24].

None of our 24 MF cases revealed distinct nuclear positivity for STAT6. Weak, cytoplasmic reactivity in one MF case was found (Fig. 7). Of note, weak cytoplasmic staining was also present in scattered endothelial cells in one of our specimens, as well as scattered ductal cells from adjacent minor salivary glands in another case.

Clinically, SFTs have a predilection for middle-aged adults and are rare in children. Our series found an age range of 23–86 years with an average age of 55.1 years. They can be found in almost any anatomic site, most commonly in the deep soft tissues of the pleura and abdomen [16]. When they present within the oral cavity, buccal mucosa is the most common location [23]. While this tumor does not tend to show a gender predilection [16], 80% of our cases were found in males.

Distinguishing MFs from SFTs is clinically important for four reasons. First, SFTs have been known to act aggressively, with 10% demonstrating local or distant metastasis [4]. While some SFTs demonstrate outright malignant features, other metastasizing or clinically aggressive SFTs appear microscopically banal [16]. One study suggests, however, that oral SFTs may not demonstrate the same malignant behaviors as some of those in extraoral sites [23]. Secondly, MFs have the potential to be associated with multiple lesions, and when found on the viscera, they may be life-threatening [25]. Thirdly, SFTs rarely can be associated with Doege-Potter syndrome, hypoglycemia due to secretion of insulin-like growth factor II by the tumor. This syndrome may occur with either benign or malignant SFTs [26]. Finally, recurrent PDGFRB mutations have been found in the majority of MFs (but not all) [27, 28]. There is some evidence to prove that tyrosine kinase inhibitors may target activated PDGFRB [29, 30], leading to a potential therapy that may prove useful in difficult-to-excite lesions as in the oral cavity or jawbones. More research is needed to determine the prevalence of this mutation in oral MFs and its role in therapeutic outcomes.

Conclusion

In conclusion, distinguishing between SFT and MF has important clinical implications. While distinguishing the

two entities often is straightforward on large, excisional samples, it may prove difficult on small biopsy specimens, as in the oral cavity. We presented ten new cases of oral SFT and reviewed the histopathologic similarities and differences between SFTs and MFs. Additionally, we demonstrated that STAT6 immunohistochemical marker is a dependable marker to discriminate the two entities.

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