

# Orbital Solitary Fibrous Tumor: Report of Two Cases and Literature Review

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**Background:** Solitary fibrous tumor (SFT) is a rare, benign, and very uncommon lesion in the orbit. Because of its complex and variable clinical and histological appearance the SFT is often misdiagnosed.

**Cases:** Two new cases of orbital SFT are reported, one in a man and the other in a woman, both unilateral and in the superomedial orbit.

**Observations:** Clinical and tomographical evaluations were conducted and the lesions were excised. The histological evaluation showed the tumors were composed of spindle-shaped cells within collagen bundles and vascular channels. Immunohistochemical staining was positive for CD 34 and negative for S-100 protein.

**Conclusion:** Immunohistochemical study is an important adjuvant in determining the SFT diagnosis. Long-term follow-up is necessary because of the possibility of SFT recurrence after excision. **Jpn J Ophthalmol 2003;47:415–418** © 2003 Japanese Ophthalmological Society

**Key Words:** Immunohistochemical study, orbit, solitary fibrous tumor, spindle cell neoplasm.

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## Introduction

Solitary fibrous tumor (SFT) is a rare spindle cell neoplasm derived from mesenchymal cells, first described by Klemperer and Rabin,<sup>1</sup> that arises most often in the pleura. Extrapleural sites are uncommonly reported, including orbit, meninges, nasal and oral cavity, thyroid and parotid glands, mediastinum, pericardium, retroperitoneum, liver, kidney, spinal cord and vulva.<sup>2</sup>

The mesenchymal structures of the orbit include extraocular muscles and Müller muscle, fibroblasts, adipocytes, smooth muscle of the vascular channels, septum, bone, and the cartilage of the trochea. Five percent to eight percent of the orbital tumors arise from these structures.<sup>3</sup> Because of the great morphological similarity between these entities, misdiagnosis is common and the immunohistochemical examination is an important evaluation for making the diagnosis.

The purpose of this study is to report two new cases of orbital SFT, describing the clinical, histological, and immunohistochemical aspects of these lesions.

## Cases Reports

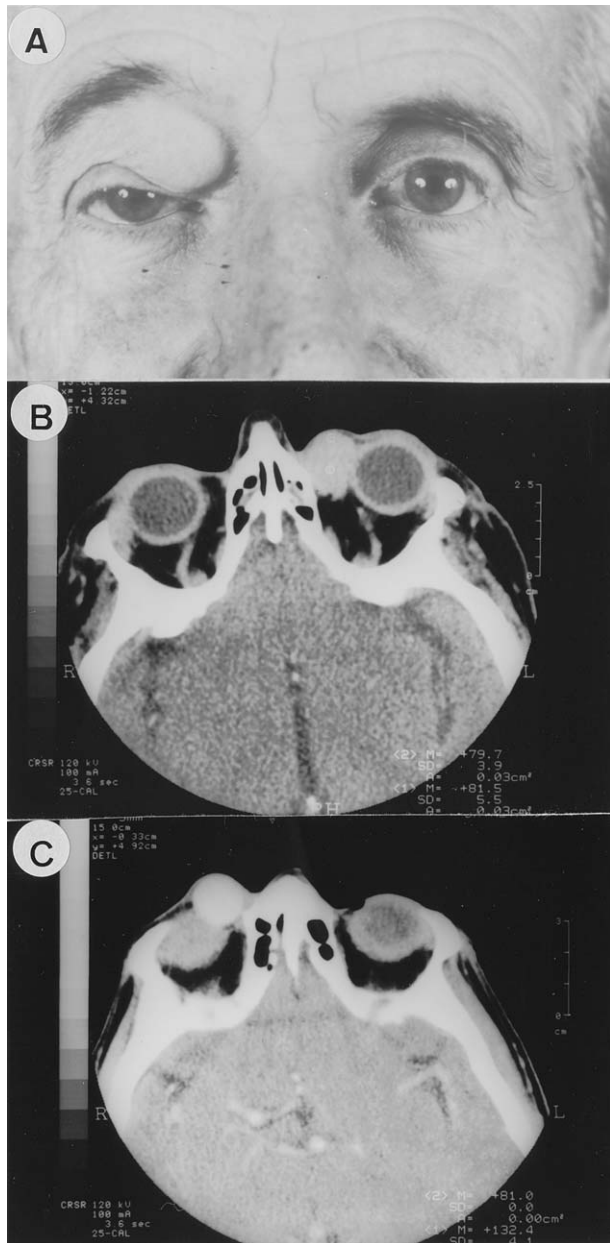
### Case 1

A 63-year-old white healthy man reported a slowly increasing painless mass for 15 years at the inner canthus of his right eye. A nontender and firm mass on his superomedial right orbit (Figure 1A) without proptosis or abnormal ocular motility was detected. On ophthalmic examination, visual acuity was 20/20 and intraocular pressure (IOP) 12 mm Hg in both eyes. A 2-cm, homogeneous, well-enhanced, soft-tissue density mass in the extraconal space of the anteromedial right orbit was detected by computed tomography (CT) scan and there was no evidence of bone destruction or calcification within the mass (Figure 1B). The tumor was removed by transcutaneous access with an incision at the superior eyelid crease. The specimen was a well-encapsulated, grayish-white

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**Figure 1.** (A) Case 1: Solitary fibrous tumor in the inner superior right orbit. (B) Case 1: Orbital computed tomograph (CT) showing the tumor in the extraconal space of the anteromedial right orbit. (C) Case 2: Orbital CT showing a well-circumscribed, soft-tissue density mass in the extraconal space in the medial left orbit.

mass, measuring  $2.5 \times 2.0 \times 1.0$  cm. Microscopic examination disclosed spindle-shaped cells with fusiform nuclei within collagen bundles without areas of atypia and some variably sized vascular channels (Figure 2A). Immunohistochemical stain (Figures 2B and 2C) exhibited positive immunoreactivity for CD34, focal staining

for collagen IV, and negative markers for S-100 protein and HNF-35. The patient has had no recurrence during the 18 months of follow-up.

### Case 2

A 46-year-old white woman reported a painless, slowly progressive tumor for 2 years at the inner canthus of her left eye. The patient had a non-tender mass on her superomedial left orbit without proptosis. Ocular motility was normal, as well as visual acuity (20/20) and IOP (10 mm Hg) in both eyes. CT scan of the orbit showed a well-circumscribed, homogeneous, well-enhanced, soft-tissue density mass in the extraconal space, measuring approximately  $2.0 \times 2.0 \times 1.5$  cm, in the superomedial left orbit with no bone destruction or calcification within the mass (Figure 1C). The patient underwent complete surgical excision of the lesion by transcutaneous access with an incision at the superior eyelid crease. Macroscopic examination showed a well-encapsulated, grayish-white mass, measuring  $2.0 \times 2.0 \times 1.5$  cm. Microscopically, there were clusters of spindle-shaped cells interspersed between dense collagen bundles and vascular channels, without atypic areas (Figure 2D). Immunohistochemical studies disclosed positive markers for CD34, focal staining for collagen IV and negative staining for S-100 protein, HNF-35, CD31, and cytokeratin (Figures 2E and 2F). The patient has had no signs of recurrence during 10 months of follow-up since the excision.

## Discussion

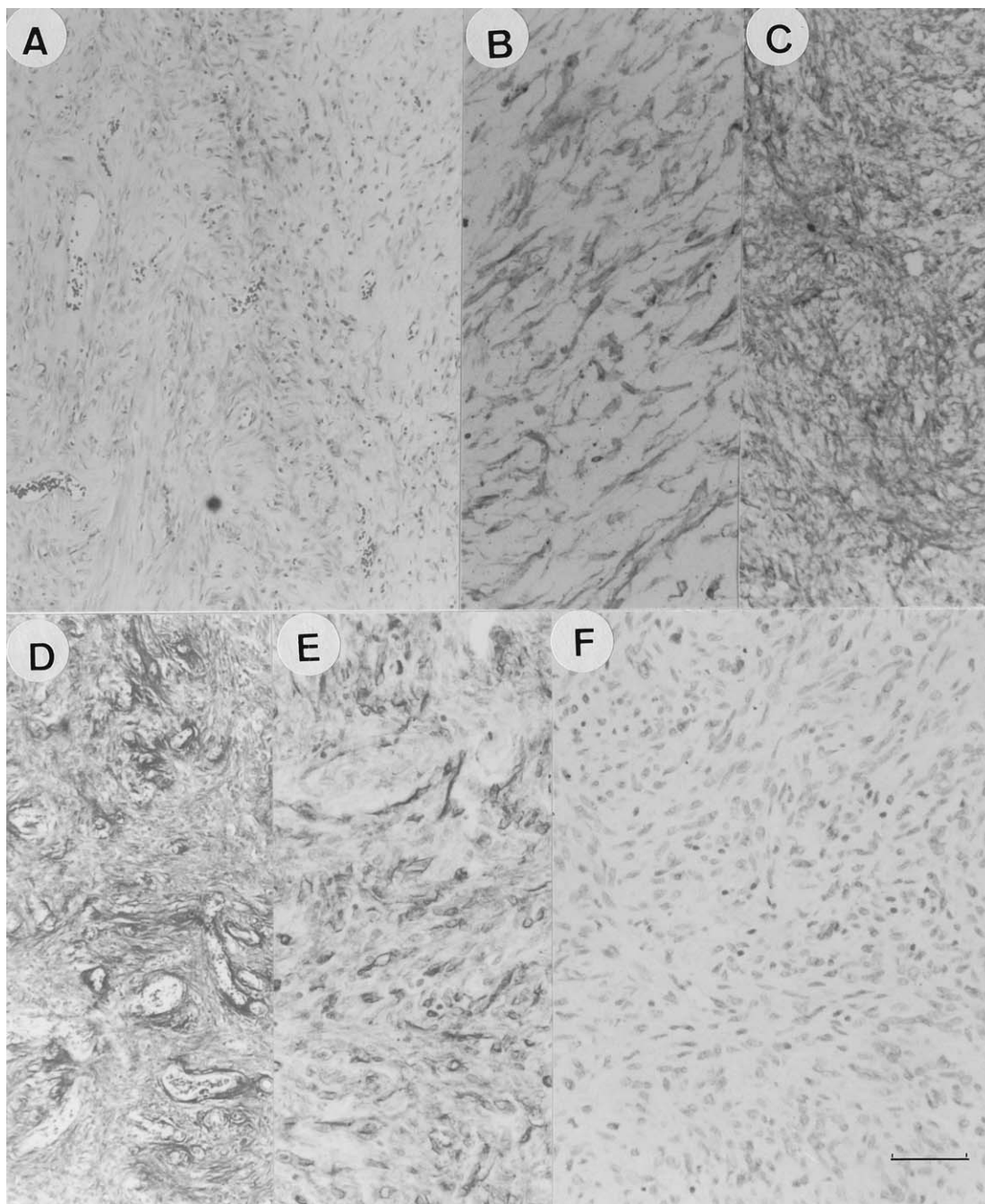
SFT is a rare neoplasm and its occurrence in extrapleural sites, especially in the orbit, was very unusual until now, only 24 orbital SFT have been described.

The tumor locations in the orbit have been inner superior canthus retro-ocular, intraconal space, eyelid, and conjunctiva.<sup>4</sup>

Clinical findings in patients with orbital SFT are variable, and age at the diagnosis has ranged from 14 to 76 (median: 44.38) years. Fourteen women and 10 men exhibited slow or abrupt progression of the orbital mass, edema, or proptosis, and symptoms were reported some years before the initial evaluation, as in our cases in which the lesions described were first noted 15 and 2 years earlier.<sup>2</sup>

Visual acuity and IOP are usually normal and optic nerve or globe compression is uncommon.

Diagnosis of orbital SFT requires CT scans and magnetic resonance imaging. Typically, CT scan shows a high-density mass with no evidence of bone destruction, and a hypointense lesion is seen in magnetic resonance.<sup>4</sup> Benign fibrous tumors of the orbit are well-circumscribed



**Figure 2.** Case 1: (A) Spindle-shaped cells with fusiform nuclei within collagen bundles without areas of atypia and some variably sized vascular channels. (B) Immunohistochemical positive markers for CD34; (C) negative markers for S-100 protein. Case 2: (D) Clusters of spindle-shaped cells interspersed between dense collagen bundles and vascular channels. (E) Immunohistochemical positive staining for CD 34; (F) negative for S-100 protein markers (Bars = 25 $\mu$ m).

lesions that appear as non specific, variably enhancing soft tissue mass.

Macroscopically, tumors appeared as single, well-circumscribed, firm, whitish-yellowish tissue very similar to those described in the pleura and other sites.<sup>2</sup>

SFT is an often misdiagnosed entity because of its complex and variable histological appearance. Histological features are nuclear palisading resembling rudimentary Verocay bodies, which are considered typical of nerve sheath tumors. The immunohistochemical markers for

neural and meningeal differentiation (S-100 protein, neuron-specific enolase, glial fibrillary acidic protein, myelinic basic protein, Leu-7, and epithelial membrane antigen) are very important for final diagnosis.

Immunohistochemical study was probably the factor that increased the diagnosis of SFT after 1994, although the entity had been recognized since 1931. The immunohistochemical study typically includes positive staining for CD34 (endothelial cell marker) and vimentin (mesenchymal cell marker) and negative staining for S-100 protein (neural differentiation marker).<sup>5</sup>

SFT has, usually, a nonaggressive, benign clinical course, evolving to cure after complete excision, and there is no report of metastatic disease derived from an orbital lesion. However, some cases may present malignant behavior, showing a more infiltrative pattern with adherence to the optic nerve and the surrounding retro-orbital soft tissues, or developing recurrence after incomplete removal months to 20 years after the tumor excision.<sup>4</sup> No death secondary to orbital tumor has been

reported.<sup>6</sup> However, because of the possibility of local recurrence, metastasis, and later complications, long-term follow-up is essential.

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