

## Case Report

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## Solitary Fibrous Tumor of the Orbit

The occurrence of solitary fibrous tumor in the orbit is rare. The following is a report on the case of a 24-year-old man presented with painless, progressive proptosis of the right eye for the duration of 1 year. Computed tomography and magnetic resonance imaging demonstrated a well-circumscribed soft tissue mass with gadolinium enhancement located in the superomedial aspect of the right orbit. The patient underwent complete tumor removal through a right fronto-orbital approach, with a pathological diagnosis of solitary fibrous tumor. Postoperatively, the patient was symptom-free. From review of the literature on orbital solitary fibrous tumor, emphasis should be placed on complete tumor resection and continuous follow-up monitoring of the tumor.

### Key Words

orbital tumor;

proptosis;

solitary fibrous tumor

**S**olitary fibrous tumor (SFT) is a rare, but distinct, spindle cell neoplasm that was initially described in the pleura by Klemperer and Rabin in 1931.<sup>1</sup> For the past few years, this tumor has been documented in almost every anatomic location,<sup>2-5</sup> with both benign and malignant variants identified.<sup>3,4,6,7</sup> Orbital involvement was first recognized in 1994.<sup>8</sup> SFT of the orbit is a rare lesion, which may have been misdiagnosed as fibrous histiocytoma, hemangiopericytoma, or neurofibroma,<sup>9</sup> or other orbital lesions before the advent of immunohistochemical stains. Recently, increased orbital SFT cases have been recognized. Here, we report of a well documented orbital solitary fibrous tumor and review of this entity.

### CASE REPORT

The patient was a 24-year-old man presented with painless, progressive protrusion of the right eye for the duration of 1 year. An ophthalmologic assessment re-

vealed that the patient had right-sided eyelid swelling, obvious proptosis, and hypoglobus with limitation of upward gaze. Pupillary reflexes were preserved and remained symmetrical. The visual acuity was 20/20 in both eyes.

Computed tomography (CT) scan of the orbits showed an approximately 18 × 24 × 25 mm, well-enhanced, mass lesion in the right eye. The tumor produced a downward and a lateral displacement of the globe. There was no evidence of either invasion of adjacent structures or bony destruction (Fig. 1A).

Magnetic resonance imaging (MRI) demonstrated a right extraconal superomedial orbital mass with inferior displacement of the medial rectus muscle. The lesion exhibited isointense signal with respect to gray matter on T1-weighted images (Fig. 1B). On T2-weighted images, the lesion exhibited heterogeneously hypointense and isointense signal (Fig. 1C). The lesion showed strong enhancement after the injection of gadolinium (Fig. 1D). The first impression derived after the image study was

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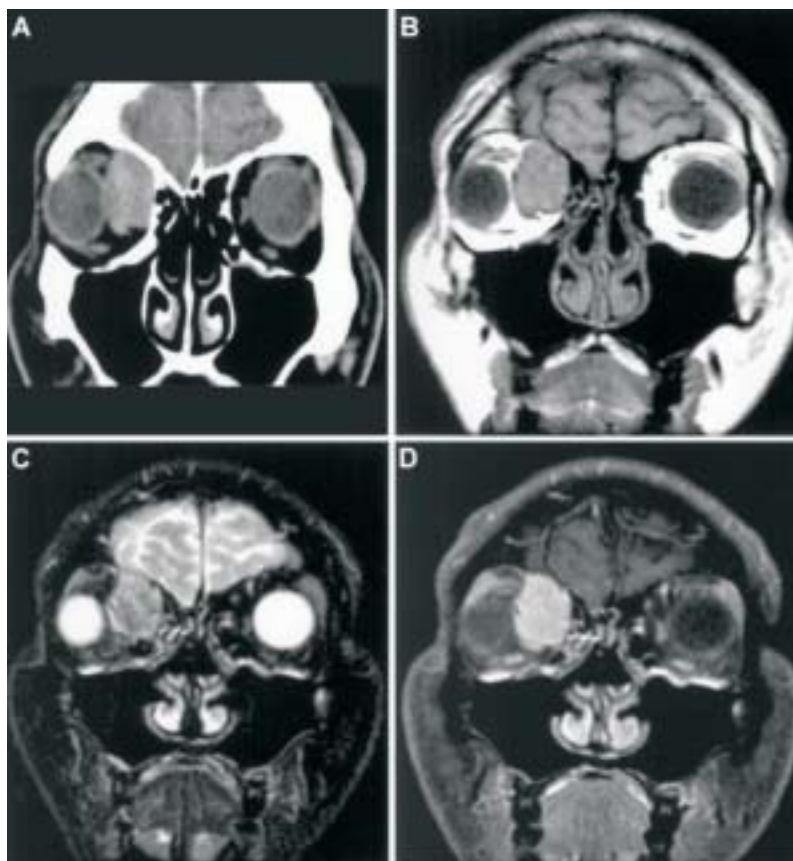
that of a hemangioma. The patient underwent a right fronto-orbital craniotomy with unroofing of the orbit. The tumor was encapsulated, with a good cleavage plane from the adjacent structures, and exhibited no evidence of bony destruction. The tumor was removed completely under microscope. After the removal, the patient's proptosis was resolved, ocular movement improved without diplopia, the visual acuity was 20/20 in both eyes, and still remained so 9 months after the operation (Figs. 2A and 2B). Histopathologically, the tumor showed cellular spindle cell areas arranged in the characteristic "patternless pattern". The mitotic rate was 1-2 per 10 high-power fields, in which moderate cellularity with minimal cellular atypia was seen. In certain areas, the tumor pattern resembled a highly vascular lesion, with some hemangiopericytic pattern and with some large di-

lated angular vessels (Fig. 3A). The tumor cells stained positively for CD34 (Fig. 3B).

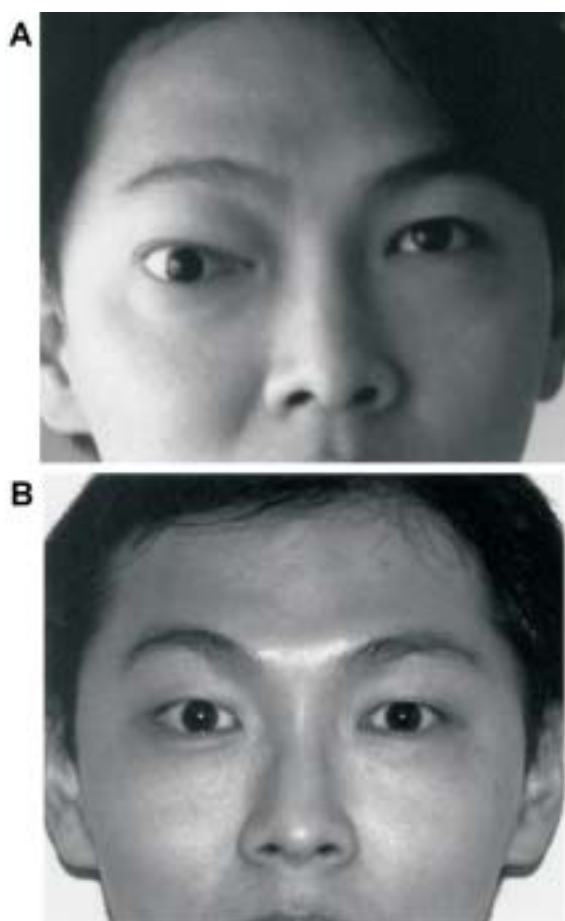
## DISCUSSION

SFT is a rare neoplasm that was originally thought to occur exclusively in the intrathoracic region. However, with increasing number of reported cases in a variety of extrapleural sites, it was recently suggested that extrapleural SFTs may actually develop more frequently than pleural tumors. Histogenesis of this neoplasm, originally thought to be of mesothelial origin, is now considered to be of mesenchymal, and possibly of fibroblastic origin.<sup>5,10</sup>

The clinical behavior is variable. Most SFTs are be-



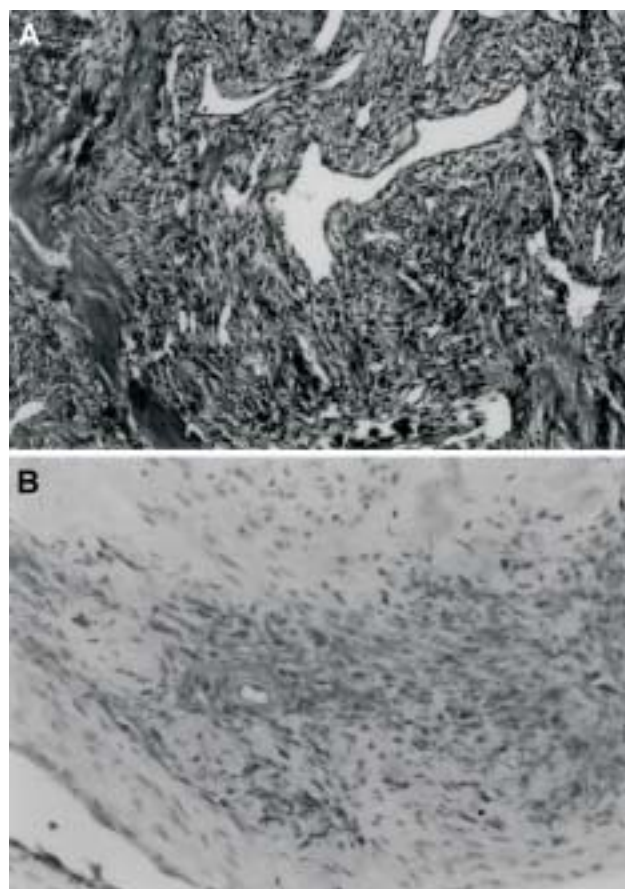
**Fig. 1.** (A) Precontrast computed tomographic coronal image reveals a homogeneous extraconal soft tissue tumor in the right superior medial orbit region with inferior displacement of medial rectus muscle and lateral inferior displacement of eyeball. (B) T1-weighted coronal MRI reveals an extraconal mass that appears isointense to gray matter tumor in the right superior medial orbit region with inferior displacement of medial rectus muscle and lateral inferior displacement of the eyeball. (C) T2-weighted MRI shows the mass is mixed iso- and hypo-intense. (D) T1-weighted MRI after intravenous gadolinium-DTPA shows good enhancement of the mass.



**Fig 2.** (A) Preoperative case photograph showing hypoglobus, right eyelid swelling, and proptosis. (B) Postoperative case photograph showing normalization of globe and resolution of right proptosis.

nign, but local invasion or recurrence of the lesion has been demonstrated; in a few cases, distant metastases have been reported in pleural cases.<sup>7</sup> Whether the orbital SFT has malignant behavior or not is unclear. To our knowledge, there are 8 cases that have reported recurrences,<sup>6,8,9,11,12</sup> but no cases of metastasis have been described. This corresponds with the idea that orbital SFT behaves in a non-aggressive manner. Nevertheless, in order to further reinforce this conclusion, a long-term follow-up of these patients is essential.

The CT and MRI characteristics of SFTs of the orbit illustrate a well-defined, enhanced mass lesion. The CT-specific features include a smoothly surfaced, well-delineated mass with heterogenous contrast enhancement. Bone remodeling without frank destruction reflects the tumor's relatively slow growth pattern and its



**Fig 3.** (A) Solitary fibrous tumor characterized by patternless architecture of spindle cells, branching hemangiopericytoma-like vessels, and keloidal type collagen deposition. H&E X100. (B). Tumor cells showing immunoreactivity for CD34. X200.

lack of biological aggressiveness.<sup>13</sup> The MRI demonstrates a hypointense to isointense signal on T1-weighted image, a hypointense signal with heterogeneity on T2-weighted images, and variable gadolinium contrast enhancement. Other spindle cell tumors, such as fibrous histiocytoma and hemangiopericytoma, typically demonstrate a hyperintense T2-weighted signal.

The diagnosis of SFT rests on the histological appearance and the immunohistochemical confirmation. One classic feature is the presence of spindle cells that grow in a haphazard fashion in a variably cellular stroma, known as the "patternless pattern". The stroma is often heavily collagenized, with bands of collagen interspersed between the tumor cells. Branching staghorn vascular channels is a prominent feature, similar to the vascular spaces seen in hamangiopericytoma.<sup>4,5</sup> Orbital

solitary fibrous tumors can mimic other spindle cell tumors of the orbit, which include fibrous histiocytoma, meningioma, schwannoma and hemangiopericytoma. In light of the tumor's varied and potentially confusing histologic appearance, it is possible that previous, unrecognized cases of orbital SFTs may have been classified under 1 of these diagnostic headings. Recently, CD34 immunoreactivity was found to be a highly sensitive marker for SFTs.<sup>10</sup> All of the SFTs demonstrated strong cytoplasmic staining with a monoclonal antibody to CD34. CD34 is an antigen expressed on the surface of vascular endothelium and hematopoietic progenitor cells.<sup>9,11</sup> Intense CD34 immunohistochemical staining helps to differentiate SFTs from hemangiopericytoma, which shows weak and patchy CD34 staining, and also from fibrous histiocytoma, which shows CD34 negative.<sup>10</sup> Schwannoma is also CD34-positive, but it is very focal, and it will be strongly positive for neural markers such as S-100 protein.<sup>14</sup>

Treatment of an SFT of the orbit consists of complete surgical excision with long-term follow-up. Various surgical approaches are available for resecting orbital tumors: fronto-orbital approach; the pterional approach; lateral orbitotomy; and medial orbitotomy. Lesions with intracranial extension, involving the optic canal, or medial to the optic nerve in the apex, are addressed via the transcranial fronto-orbital approach.<sup>15</sup> A long-term follow-up is requisite because recurrences may occur several years after the excision of the primary tumor<sup>9</sup> and the efficacy of either radiotherapy or chemotherapy in the management of residual SFT remains unclear.

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