Solitary Fibrous Tumor of the Orbit
CASE REPORT AND REVIEW OF THE LITERATURE

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BACKGROUND
Solitary fibrous tumor (SFT), which usually presents in the pleura and is thought to be mesothelial in nature, has been recently discovered in extrapleural sites, including the orbit. Presently ultrastructural studies show absence of epithelial-mesothelial features, and reactivity of the tumor cells to CD34 antigen on immunohistochemical analysis suggests the mesenchymal origin of such tumors.

CASE DESCRIPTION
A 40-year-old woman had a 4-year history of progressive swelling of her right upper lid and a slow-growing palpable mass of the orbit. CT and MR imaging showed a well circumscribed, nonenhanced extraconal mass with mild erosion of the right orbital roof. The tumor was totally excised. Histological examination disclosed a spindle-cell tumor in a dense fibrous tissue. Immunohistochemistry showed positive staining for vimentin and CD34. We review the clinical, diagnostic, and surgical features of 22 orbital SFTs including the present case.

CONCLUSIONS
Orbital SFT generally pursues a slow, indolent, and non-aggressive course, reaches a size up to 4.5 cm, and can be cured by a single excision. It must be immunohistochemically differentiated from other spindle-cell tumors of the orbit. © 2001 by Elsevier Science Inc.

KEY WORDS
Solitary fibrous tumor, orbit, CD34 immunoreactivity, histogenesis.
(2.5 × 2 cm) was sharply defined with mild erosion of the adjacent bony orbital roof. At operation a right fronto-orbital craniotomy with unroofing of the orbit was performed. The tumor, which was extraconal and mildly adherent to the surrounding soft tissue, was totally excised.

**PATHOLOGICAL FINDINGS**

The lesion on gross examination was a well circumscribed but not encapsulated firm tumor, grey-blue in colour. The cut surface was relatively homogeneous. Microscopically, regions of hypercellularity alternated with areas of hypocellularity. The hypercellular areas were composed of spindle-shaped cells in a dense fibrous tissue (Figure 2A). We also observed hypocellular areas in which scattered single cells were surrounded by bands of hyalinized collagen. The cellular component was made up of spindle-shaped cells. No nuclear pleomorphism, mitotic activity, or invasion of adjacent structures were observed. The fibroblastic nature of these cells was confirmed by a strong positive reaction to vimentin (Figure 2B) and by a negative reaction to glial fibrillary acid protein, S-100 protein, and desmin. Moreover, focal immunoreactivity to CD34 was present (Figure 2C). Giant cells or hemosiderin deposits were observed.

**DISCUSSION**

Table 1 summarizes the clinical, diagnostic and surgical features of 22 cases of SFTs [1–5,7–14,16] including the presented one. Patients’ ages range

![Coronal contrast-enhanced T1-weighted MRI image of the tumor in the right orbit with erosion of the orbital roof.](image1)

![Photomicrograph showing histological features of the tumor: spindle-shaped cells in a dense fibrous tissue (H.E. × 400).](image2A)

![Immunohistochemistry: primary antibody vimentin revealed by peroxidase reaction (×400).](image2B)

![Immunohistochemistry: primary antibody CD34 revealed by peroxidase reaction (× 400).](image2C)
Clinical, Diagnostic, and Surgical Features of 22 Cases of SFTs Including the Present One

<table>
<thead>
<tr>
<th>CASE NO.</th>
<th>AUTHOR/YEAR</th>
<th>SEX</th>
<th>AGE (YRS)</th>
<th>SEX</th>
<th>DURATION OF SYMPTOMS</th>
<th>PROPTOSIS</th>
<th>VISUAL ACUITY</th>
<th>IMPAIRED OCULOMOTION</th>
<th>IMAGING</th>
<th>SURGICAL APPROACH</th>
<th>LOCATION IN ORBIT/SIZE (CM)</th>
<th>FOLLOW-UP</th>
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<tbody>
<tr>
<td>1</td>
<td>Westra et al 1994</td>
<td>M</td>
<td>60</td>
<td></td>
<td></td>
<td></td>
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<tr>
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<td></td>
<td>63</td>
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<tr>
<td>3</td>
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<td>44</td>
<td>R</td>
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<td></td>
<td>69</td>
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<td></td>
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<td>F</td>
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<td>20</td>
<td>R</td>
<td></td>
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<td>8</td>
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<tr>
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<td>Re-excision 3 mos later; NED after 10 mos</td>
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<td>F</td>
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<td>R</td>
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<td>N</td>
<td>Yes Ptosis</td>
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<td>Craniotomy, Extraconal Superolateral, 2.5 x 2</td>
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LGF = lacrimal gland fossa; N = normal; NED = no evidence of disease.
from 20 to 76 years (median age 48 years. No sex predilection is apparent. The left orbit seems more frequently involved than the right one but, because of the limited number of the cases, this is not statistically significant. Of the patients for whom the duration of symptoms is known, 15 had been symptomatic longer than 5 months (from 6 months to 5 years) and 3 had had symptoms for less than 5 months.

Orbital SFTs, like SFTs elsewhere, are slow-growing masses that pursue an indolent course. Most patients have a history of gradual painless proptosis; a few have swelling of the orbit, with visual acuity usually normal or mildly impaired. These tumors arise anywhere inside the orbit; none of them have shown evidence of extension into bone or extra-orbital soft tissue. They are easily detected by neuroimaging as circumscribed masses of significant volume (from 1 to 4 or 5 cm in diameter).

As do SFTs in most sites, orbital SFTs generally pursue a benign, nonaggressive course and are usually cured by a simple excision. Nevertheless, in one case [7] the SFT was reported to involve the entire orbit and require enucleation.

So far, two cases out of 22 orbital SFTs have been documented to recur [2,3]; in the third case reported by Dorfman et al [3], the tumor was incompletely excised at the second recurrence also. In the case of de Saint Aubain et al [2] the course was uneventful after the re-excision. There have been no deaths related to orbital SFTs.

A wide range of mesenchymal tumors occur in the orbit. The differential diagnosis of orbital SFT includes fibrous histiocytoma (the most common mesenchymal orbital tumor in adults), rare fibroblastic tumors such as the juvenile fibromatosis of the orbit, and other uncommon spindle-cell tumors such as hemangiopericytoma, nerve sheath tumors, and meningioma. Therefore, it is possible that previous orbital SFTs have been misdiagnosed. Recently, strong and consistent CD34 immunoreactivity has been found in SFTs and this marker facilitates histopathological differentiation of this lesion from other spindle-cell tumors of the orbit.

CD34 antigen is expressed by bone marrow progenitor cells and in vascular endothelium. In addition to acute leukemia and vascular tumors, the spectrum of CD34-positive tumors has expanded to include primitive tumors [14]. Therefore, the strong CD34 immunoreactivity of SFTs is indicative of a mesenchymal origin of these tumors that, moreover, show negative immunoreactivity to epithelial markers and, ultrastructurally, absence of mesothelial features.

Conclusions

In recent years, there has been a steady increase in the number of extrapleural sites of occurrence of SFTs, reported, including the orbit. Orbital SFT seem to pursue a benign nonaggressive course. However, because of the small number of reported cases, as well the limited follow-up, it is not possible at present to assert that tumors in extrapleural sites behave differently from those in the pleura. Careful long-term follow-up is necessary because recurrence may appear long after excision of the primary tumor.

REFERENCES

16. Westra WH, Gerald WL, Rosai J. Solitary fibrous tu-

COMMENTARY
Solitary fibrous tumor of the orbit is indeed a rare lesion, but the 22 cases compiled by the authors indicate that it is being recognized more frequently. It is certainly possible that in the past without the immunochemistry used in the present case report, errors in diagnosis could have been made. The generally benign nature of the lesion with its potential for complete resectability supports an aggressive approach to its removal.

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We asked doctors what they do or plan to do to increase income. Among the answers: “Moonlight.” “Work more hours.” “Keep the office open longer.” “Expand my side income from CME teaching.” Some young physicians operate nonmedical sideline businesses.

—Brad Burg, “Young doctors face a steep climb”
“Medical Economics,” August 20, 2001

Forty percent of doctors under 34 years of age owe $200,000 or more, while 42% of doctors 35 to 39 years of age owe $200,000 or more.

—Brad Burg, “Young doctors face a steep climb”
“Medical Economics,” August 20, 2001