Solitary Fibrous Tumors of the Head and Neck

A Clinicopathologic and Radiologic Review

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Objective: To describe the clinicopathologic and radiologic features of solitary fibrous tumors of the head and neck.

Design: Retrospective analysis.

Setting: Tertiary referral center that performs head and neck surgical oncology.

Patients: Twelve patients with solitary fibrous tumors of the head and neck identified from the pathology and soft tissue tumor databases at Memorial Sloan-Kettering Cancer Center, New York, NY, from 1990 to 2004. All cases were reviewed by 3 experienced pathologists, 1 of whom is an experienced soft tissue tumor pathologist. The diagnosis was confirmed by microscopic features on hematoxylin-eosin staining and by positive staining for CD34 and Bcl2 on immunohistochemical analysis. Tumors were scored for mitotic activity, cellularity, nuclear pleomorphism, necrosis, and the presence of a malignant component. Details on patient characteristics, tumor characteristics, previous treatment and surgery, adjuvant treatment, and outcome were recorded from clinical records.

Results: Solitary fibrous tumors occurred in patients over a wide age range (27-78 years; median age, 52 years). Seven patients (58%) were women, and 5 (42%) were men. Most tumors presented as a slow-growing painless mass with a duration ranging from 2 months to 5 years. The tumors ranged from 1 × 1 cm to 6 × 5 cm. Patients presented with a subcutaneous mass of the scalp or face in 4 cases, intraoral mass in 4, sinonasal mass in 3, and paraspinal mass in 1. Computed tomographic and/or magnetic resonance imaging scans of 7 of the 12 patients showed well-circumscribed tumors that enhanced strongly with contrast. Treatment for all of the patients was surgical resection. Pathologic findings showed that 9 tumors were benign and 3 were malignant. Three patients had a positive surgical resection margin. All patients were alive at a median follow-up of 8 months (range, 1-76 months). Local recurrence occurred in 1 patient who had positive surgical margins 3 years after the initial surgery.

Conclusions: Solitary fibrous tumors of the head and neck region are rare and most commonly benign. The diagnosis depends on microscopic and immunohistochemical features, although imaging may help. Patients with these tumors can be safely treated with local excision, but tumors with positive margins require close follow-up over several years owing to the potential for late local recurrence.

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Solitary fibrous tumors (SFTs), first reported in the pleura by Klemperer and Rabin in 1931, are uncommon spindle cell neoplasms of mesenchymal origin. Over 50% of these tumors are located in the thoracic cavity, but extrathoracic tumors have been reported in many sites, including the liver, adrenal gland, skin, and the head and neck. In the head and neck, the most common site affected is the oral cavity, but case reports have been documented in the orbit, nose and paranasal sinuses, nasopharynx and parapharyngeal space, larynx, major salivary glands, and thyroid. Most of these reports have little data on the biological behavior of these rare tumors, but in general it is thought that they present as asymptomatic slow-growing masses that can be very difficult to distinguish from other soft tissue tumors, such as synovial sarcoma, benign fibrous histiocytoma, dermatofibrosarcoma protuberans, neurofibroma, schwannoma, fibromas, and myofibromas. The diagnosis is therefore strongly dependent on the microscopic appearance and characteristic immunohistochemical staining for CD34 and Bcl2. Clinically, these tumors are commonly treated by local excision without adjuvant therapy because they are believed to be relatively slow growing. However, there are few series that document the clinical presentation, surgical management, and outcome of these tumors in the head and neck region in any great detail. The present study was designed to analyze our experience with this rare tumor in...
the head and neck region by examining the medical records of 12 patients who were treated at a single head and neck unit from 1990 to 2004. Our objective is to report the clinicopathologic and radiologic features of these tumors in their diagnosis. In addition, we report the outcome of these tumors after surgical excision.

**METHODS**

Twelve patients with SFTs were identified from the pathology and soft tissue tumor databases at Memorial Sloan-Kettering Cancer Center, New York, NY, from 1990 to 2004. Archived radiologic imaging scans (using computed tomography [CT] or magnetic resonance [MR] imaging or both) were available for 7 of the 12 patients and were reviewed by an experienced CAQ (certificate of added qualification) certified neuroradiologist (H.E.S.). All cases were reviewed by 3 experienced pathologists (R.G., M.E., and D.C.), 1 of whom (M.E.) is an experienced soft tissue tumor pathologist. The diagnosis was confirmed by microscopic features on hematoxylin-eosin staining. Characteristically, these tumors are well circumscribed, with spindle cells arranged in a haphazard fashion separated by strands of collagen. They are composed of alternating areas of hypercellularity (tumor rich) and hypocellularity (collagen rich). Prominent vascularity, with vessels with a staghorn pattern, may be seen. Diagnosis is aided by positive staining for CD34 and Bcl2 on immunohistochemical analysis, as well as CD99. Tumors were scored for mitotic activity (determined by counting the number

<table>
<thead>
<tr>
<th>Patient/Sex/ Age, y</th>
<th>Site</th>
<th>Primary Site</th>
<th>Size, cm</th>
<th>Clinical Presentation</th>
<th>Duration, mo</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/M/47</td>
<td>Intraoral</td>
<td>Left infratemporal fossa</td>
<td>5 x 3</td>
<td>Mass</td>
<td>4</td>
</tr>
<tr>
<td>2/F/67</td>
<td>Intraoral</td>
<td>Left anterior floor of mouth</td>
<td>1 x 1</td>
<td>Mass</td>
<td>4</td>
</tr>
<tr>
<td>3/M/44</td>
<td>Intraoral</td>
<td>Left upper gum</td>
<td>1 x 1</td>
<td>Mass</td>
<td>4</td>
</tr>
<tr>
<td>4/M/54</td>
<td>Intraoral</td>
<td>Right buccal space</td>
<td>2.5 x 2</td>
<td>Mass</td>
<td>4</td>
</tr>
<tr>
<td>5/F/57</td>
<td>Paraspinal</td>
<td>Right paraspinal mass</td>
<td>6 x 3.5</td>
<td>Arm pain</td>
<td>4</td>
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<tr>
<td>6/M/57</td>
<td>Sinonasal</td>
<td>Left ethmoid</td>
<td>6 x 6</td>
<td>Nasal obstruction</td>
<td>4</td>
</tr>
<tr>
<td>7/F/78</td>
<td>Sinonasal</td>
<td>Left frontoethmoid</td>
<td>6 x 5</td>
<td>Ptosis</td>
<td>36</td>
</tr>
<tr>
<td>8/F/51</td>
<td>Sinonasal</td>
<td>Right nasal</td>
<td>6 x 3</td>
<td>Nasal obstruction</td>
<td>2</td>
</tr>
<tr>
<td>9/F/59</td>
<td>Subcutaneous</td>
<td>Right suboccipital</td>
<td>4 x 3</td>
<td>Mass</td>
<td>4</td>
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<td>10/M/27</td>
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<td>Mass</td>
<td>60</td>
</tr>
<tr>
<td>11/F/39</td>
<td>Subcutaneous</td>
<td>Left frontoparietal scalp</td>
<td>2 x 2</td>
<td>Mass</td>
<td>48</td>
</tr>
<tr>
<td>12/F/40</td>
<td>Subcutaneous</td>
<td>Right infraorbital</td>
<td>1.5 x 1</td>
<td>Mass</td>
<td>12</td>
</tr>
</tbody>
</table>

**Table 2. Radiographic Findings**

<table>
<thead>
<tr>
<th>Patient</th>
<th>CT</th>
<th>MR Imaging</th>
<th>Post T1</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Non-contrast-enhanced; isodense to muscle</td>
<td>ND</td>
<td>ND</td>
<td>Well defined; 3.8 x 5.2 x 4.0 cm; regressive remodeling of anterior bony wall left maxillary sinus, supra, and infrazygomatic, temporalis, and lateral pterygoid muscle</td>
</tr>
<tr>
<td>2</td>
<td>Contrast-enhanced; moderate enhancement</td>
<td>Isointense to muscle</td>
<td>Isointense to muscle</td>
<td>ND</td>
</tr>
<tr>
<td>3</td>
<td>Not done</td>
<td>Isointense to muscle</td>
<td>ND</td>
<td>Fairly homogeneous; strong enhancement</td>
</tr>
<tr>
<td>4</td>
<td>Not done</td>
<td>Isointense to muscle</td>
<td>Isointense to muscle</td>
<td>ND</td>
</tr>
<tr>
<td>5</td>
<td>Not done</td>
<td>Isointense to muscle</td>
<td>Malignant hyperintense to muscle</td>
<td>Homogeneous; strong enhancement</td>
</tr>
<tr>
<td>6</td>
<td>Without contrast</td>
<td>Isointense to brain</td>
<td>Isointense and hypointense to brain</td>
<td>Heterogeneous; strong enhancement</td>
</tr>
<tr>
<td>7</td>
<td>Without contrast, isodense to brain, with contrast, mild enhancement</td>
<td>Isointense to brain</td>
<td>Isointense to brain</td>
<td>Homogeneous; strong enhancement</td>
</tr>
<tr>
<td>8</td>
<td>Without contrast, isodense to muscle</td>
<td>ND</td>
<td>ND</td>
<td>Well defined; 6.3 x 2.1 x 3.2 cm; expansile remodeling of bone</td>
</tr>
</tbody>
</table>

**Abbreviations:** CT, computed tomography; MR, magnetic resonance; ND, not done; TW1, T1-weighted image; TW2, T2-weighted image.  
*Neither CT nor MR imaging was performed on patients 9 to 12.
of mitotic figures per 10 high-power fields, cellularity, necrosis, and the presence of a malignant component. We considered SFTs to have a malignant component if there were areas of increased cellularity (>5%), an absence of alternating areas of sclerosis, and a mitotic rate of 4 or more mitoses per 10 high-power fields. Details on patient characteristics, tumor characteristics, previous treatment and surgery, adjuvant treatment, and outcome were obtained from clinical records.

**RESULTS**

**CLINICAL FEATURES**

Table 1 summarizes the patient and tumor characteristics. Seven patients (58%) were women, and 5 (42%) were men. Their ages ranged from 27 to 78 years (median age, 52 years). Ten of the 12 patients were examined at Memorial Sloan-Kettering Cancer Center for initial treatment but 2 (patients 8 and 10) had their initial surgery performed at a different institution. Patient 8 was referred for further treatment after endoscopic resection, and patient 10 was referred because of local recurrence. Most tumors presented as a slow-growing painless mass with duration ranging from 2 months to 5 years. The tumors ranged in size from 1 × 1 cm to 6 × 5 cm. Four patients presented with a subcutaneous mass of the scalp or face and 4 with an intraoral mass. Three patients with sinonasal SFTs presented with nasal obstruction in 2 cases and ptosis in 1. One patient with a paraspinal SFT presented with arm pain.

**RADIOGRAPHIC FINDINGS**

Table 2 is a summary of the radiographic features of the 7 patients whose imaging studies were available for review. All tumors were well defined with no infiltrative borders, even in the presence of bone destruction, and were isodense to muscle or brain on noncontrast CT scan (Figure 1). With MR imaging, all except 1 of the tumors were isointense to brain or muscle on precontrast T1-weighted (T1W) and T2-weighted (T2W) sequences (Figure 2). The tumor was compared with muscle or brain for signal characteristics depending on its location; lesions in the paranasal sinuses were therefore compared with brain parenchyma, whereas more remote lesions, such as those in the floor of the mouth or in the spine, were compared with adjacent musculature. Although most tumors were isointense to muscle or brain, the T2W signal showed the most variability of the MR imaging sequences. Some tumors exhibited scat-
tered hyperintense areas, and 1 tumor (in patient 5) was uniformly hyperintense relative to the adjacent paraspinal muscles.

Because SFTs are generally slow-growing benign lesions, they tend to cause regressive remodeling of adjacent bone (Figure 3). The benign appearance of SFTs on the bone windows of a CT scan can, however, be misleading. They can have the overall appearance of regressive remodeling, but there may be smaller foci of bone destruction that can be easily overlooked (Figure 4). Two of the 7 patients (patients 6 and 7) showed radiographic evidence of bone destruction. Pathologic evaluation of these 2 tumors showed a benign SFT in patient 6 and a malignant SFT in patient 7. Of the 3 patients with malignant SFTs, only 1 showed evidence of bone destruction on imaging. The other 2 tumors displayed no evidence of aggressive growth but rather showed regressive remodeling of adjacent bone only.

The most common distinctive feature was dense enhancement of the SFT on postcontrast CT scan (Figure 5) and MR imaging (Figure 6). Contrast enhancement was generally homogeneous, except for 1 patient who had a focus of low signal on all MR imaging sequences whereas the remainder of the tumor enhanced uniformly (Figure 7).

**PATHOLOGIC FINDINGS**

Figure 8 shows the gross appearance of a floor-of-mouth SFT excised from patient 2. The histopathologic features of the tumors are shown in Table 3. The microscopic appearance of a typical SFT stained with hematoxylin-eosin at low- and high-power fields is shown in Figure 9. This figure illustrates spindle cells in a characteristic “patternless pattern” with amorphous areas of collagen and haphazardly arranged cells. All tumors tested were positive for the presence of CD34 and Bcl2 (Figure 10 and Figure 11), which is characteristic for SFTs. A malignant component (Figure 12) was present in the tumors of patients 7, 8, and 10. Three patients had a positive surgical margin. An additional 3 patients (patients 6, 7, and 8) had piecemeal or endoscopic removal of their tumors and therefore margins could not be assessed. Patients 7 and 8 were later found to have malignant SFTs.

**TREATMENT AND OUTCOME**

Table 4 gives details of the surgery each patient underwent, the adjuvant treatment, and the outcome. All patients were treated with surgical resection. Pathologic findings showed that 9 tumors were benign and 3 were malignant (in patients 7, 8, and 10). In patient 7, an
anterior craniofacial resection had been performed. Because pathologic findings showed a malignant SFT and the margins were positive, the patient received adjuvant postoperative radiation therapy. Patient 8 underwent endoscopic resection of an SFT arising from the right nasal cavity and was referred for further treatment when the pathologic findings reported it as a malignant SFT. This patient has been observed closely for 12 months and has had no local recurrence. Patient 10 underwent a peroral excision at another hospital, pathologic findings of which also showed a malignant SFT with positive margins. This patient was initially managed by close observation, but 3 years after initial surgery he developed local recurrence and was referred to Memorial Sloan-Kettering Cancer Center for further treatment. The patient underwent a further wide local excision and at the time of this writing is alive and free of disease.

Follow-up periods ranged from 1 to 76 months (median, 8 months). Because of the limited number of patients, we did not attempt analysis of survival using the Kaplan-Meier method. All patients were alive at the time of article preparation, 11 with no evidence of disease and 1 patient (patient 1) with disease. This patient had residual disease after undergoing initial surgery (and revisional surgery) because complete resection of tumors located in the infratemporal fossa is not always possible. Two patients (patients 9 and 12) with positive re-

Figure 5. The most common distinctive feature was dense enhancement of the solitary fibrous tumor on a postcontrast computed tomographic scan. A left floor-of-mouth mass in patient 2 enhances strongly with contrast (arrow).

Figure 6. Dense enhancement of a solitary fibrous tumor in the frontal sinus (patient 7) on postcontrast T1-weighted magnetic resonance imaging with associated postobstructive mucocele (arrow).

Figure 7. Contrast enhancement was generally homogeneous, except for 1 patient (patient 6) who had a focus of low signal on all magnetic resonance imaging sequences whereas the remainder of the tumor enhanced uniformly, as this postcontrast T1-weighted fat-saturated image demonstrates.

Figure 8. Gross appearance of solitary fibrous tumor from a floor-of-mouth resection (patient 2); the scale is in centimeters.
section margins have no evidence of local recurrence and are being managed by close observation. Three other patients (patients 6, 7, and 8) had piecemeal or endoscopic removal of their tumors and can therefore be considered to have minimal residual disease. Patient 6 had not experienced local recurrence at 4 months’ follow-up but is being observed closely. Patient 7 received adjuvant postoperative radiation therapy owing to the presence of a malignant component on histopathologic examination. At 16 months’ follow-up, this patient also had no evidence of local recurrence. Patient 8 did not receive adjuvant postoperative radiation therapy but she is being closely followed up and showed no evidence of local recurrence at 12 months postsurgery.

### Table 3. Pathologic Characteristics

<table>
<thead>
<tr>
<th>Patient</th>
<th>Mitoses per HPF</th>
<th>Necrosis</th>
<th>Increased Cellularity</th>
<th>Malignant Component</th>
<th>Margins</th>
<th>CD34</th>
<th>Bcl2</th>
</tr>
</thead>
<tbody>
<tr>
<td>1A</td>
<td>&lt;4/10</td>
<td>No</td>
<td>No</td>
<td>No</td>
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<td>Positive</td>
<td>Positive</td>
</tr>
<tr>
<td>1B</td>
<td>1</td>
<td>No</td>
<td>No</td>
<td>No</td>
<td>Positive</td>
<td>Positive</td>
<td>Positive</td>
</tr>
<tr>
<td>2</td>
<td>1</td>
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<td>No</td>
<td>No</td>
<td>Negative</td>
<td>Positive</td>
<td>Positive</td>
</tr>
<tr>
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<td>1</td>
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<td>No</td>
<td>No</td>
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<td>Positive</td>
<td>Positive</td>
</tr>
<tr>
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<td>0</td>
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<td>No</td>
<td>No</td>
<td>Negative</td>
<td>Positive</td>
<td>Positive</td>
</tr>
<tr>
<td>5</td>
<td>1</td>
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<td>No</td>
<td>No</td>
<td>Negative</td>
<td>Positive</td>
<td>Positive</td>
</tr>
<tr>
<td>6</td>
<td>3</td>
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<td>No</td>
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<td>Positive</td>
</tr>
<tr>
<td>7</td>
<td>9</td>
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<td>Yes</td>
<td>Yes</td>
<td>NA</td>
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<td>Positive</td>
</tr>
<tr>
<td>8</td>
<td>7</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
<td>NA</td>
<td>ND</td>
<td>ND</td>
</tr>
<tr>
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<td>Positive</td>
</tr>
<tr>
<td>10</td>
<td>6</td>
<td>No</td>
<td>Yes</td>
<td>Yes</td>
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<td>ND</td>
</tr>
<tr>
<td>11</td>
<td>2</td>
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<td>No</td>
<td>No</td>
<td>NA</td>
<td>ND</td>
<td>ND</td>
</tr>
<tr>
<td>12</td>
<td>3</td>
<td>No</td>
<td>Yes</td>
<td>No</td>
<td>Positive</td>
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<td>Positive</td>
</tr>
</tbody>
</table>

Abbreviations: HPF, high-power field; NA, not applicable (tumor in pieces); ND, not done; 1A, patient 1, primary occurrence; 1B, patient 1, recurrence.

**Figure 9.** Microscopic appearance of typical solitary fibrous tumor stained with hematoxylin-eosin, at low- (A) (original magnification ×4) and high-power fields (B) (original magnification ×20).

**Figure 10.** CD34 immunohistochemistry of solitary fibrous tumor. The slides were stained with an antibody against either CD34 or Bcl2 followed by detection with avidin-diaminobenzidine, original magnification ×20.

**Figure 11.** Bcl2 immunohistochemistry of solitary fibrous tumor. The slides were stained with an antibody against either CD34 or Bcl2 followed by detection with avidin-diaminobenzidine, original magnification ×40.
Solitary fibrous tumors are rare and most commonly arise in the thoracic cavity. They occur with equal sex predilection over a wide age range. Most present as slow-growing painless masses. Rarely, larger tumors may be a source of paraneoplastic syndromes such as hypoglycemia owing to the production of insulin-like growth factor. In the head and neck region, SFTs have been reported in all sites, but the oral cavity is the most common site of occurrence.

In our study of 12 patients, all presented with a slow-growing mass of 2 to 60 months’ duration. Eight patients were asymptomatic, 1 presented with arm pain, 2 with nasal obstruction, and another with ptosis. These clinical presentations are consistent with previous case reports on SFTs of the head and neck. According to the literature, the oral cavity is the most common site of occurrence of a densely enhancing lesion revealed with CT and MR imaging. This is consistent with previous experiences reported in the literature. In our series of patients, all tumors were well defined and isodense when seen on CT. All tumors were isointense on T1W precontrast images, and most were isointense on T2W sequences, although some showed scattered foci of hyperintensity and one was diffusely hyperintense to adjacent musculature. Interestingly, a small portion of one tumor was hypointense in T2W and precontrast T1W MR imaging and did not enhance on a postcontrast T1W sequence. The precise implication of this observation is unclear, but it is likely due to the presence of areas of particularly dense fibrous tissue within the tumor. Tightly woven fibrous tissue is less likely to enhance postcontrast and shows as a hypointense area on T1W and T2W sequences on MR imaging. Accurate correlation of the radiographic hypodense focus to the histologic features in this tumor was not possible because the tumor was not resected en bloc. On histopathologic examination of the specimen, though, areas of hyalinization were indicative of dense fibrosis.

Regressive remodeling of adjacent bone secondary to long-standing pressure effect is the usual radiographic

Table 4. Details on Treatment and Outcome

<table>
<thead>
<tr>
<th>Patient</th>
<th>Type of Surgical Procedure</th>
<th>Surgical Margins</th>
<th>Pathologic Findings</th>
<th>Adjuvant Treatment</th>
<th>Recurrence</th>
<th>Follow up, mo</th>
<th>Status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Mandibulotomy, resection infratemporal fossa</td>
<td>Positive</td>
<td>Benign</td>
<td>None</td>
<td>None</td>
<td>76</td>
<td>AWD</td>
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<tr>
<td>2</td>
<td>Wide local excision</td>
<td>Negative</td>
<td>Benign</td>
<td>None</td>
<td>None</td>
<td>3</td>
<td>ANED</td>
</tr>
<tr>
<td>3</td>
<td>Wide local excision</td>
<td>Negative</td>
<td>Benign</td>
<td>None</td>
<td>None</td>
<td>28</td>
<td>ANED</td>
</tr>
<tr>
<td>4</td>
<td>Right cheek flap, marginal mandibulotomy, marginal maxillectomy, free radial forearm flap</td>
<td>Negative</td>
<td>Benign</td>
<td>None</td>
<td>None</td>
<td>3</td>
<td>ANED</td>
</tr>
<tr>
<td>5</td>
<td>Wide local excision of the paraspinal area from C2-T1</td>
<td>Negative</td>
<td>Benign</td>
<td>None</td>
<td>None</td>
<td>2</td>
<td>ANED</td>
</tr>
<tr>
<td>6</td>
<td>Anterior craniofacial resection, transcranial approach</td>
<td>NA</td>
<td>Benign</td>
<td>None</td>
<td>None</td>
<td>4</td>
<td>ANED</td>
</tr>
<tr>
<td>7</td>
<td>Anterior craniofacial resection (transcranial and transfacial approach), calvarial bone graft and pericranial flap</td>
<td>NA</td>
<td>Malignant</td>
<td>Radiation therapy</td>
<td>None</td>
<td>16</td>
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<tr>
<td>8</td>
<td>Endoscopic resection</td>
<td>NA</td>
<td>Malignant</td>
<td>None</td>
<td>None</td>
<td>12</td>
<td>ANED</td>
</tr>
<tr>
<td>9</td>
<td>Wide local excision</td>
<td>Positive</td>
<td>Benign</td>
<td>None</td>
<td>None</td>
<td>65</td>
<td>ANED</td>
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<tr>
<td>10</td>
<td>Peroral wide local excision</td>
<td>NA</td>
<td>Malignant</td>
<td>None</td>
<td>Local</td>
<td>39</td>
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</tr>
<tr>
<td>11</td>
<td>Wide local excision and split thickness skin graft</td>
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<td>Benign</td>
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<td>None</td>
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<tr>
<td>12</td>
<td>Wide local excision</td>
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<td>Benign</td>
<td>None</td>
<td>None</td>
<td>1</td>
<td>ANED</td>
</tr>
</tbody>
</table>

Abbreviations: ANED, alive with no evidence of disease; AWD, alive with disease; NA, not applicable (tumor in pieces).
osseous finding because most SFTs are benign and slow growing. As demonstrated by the radiographic findings in patient 7, who had minimal bone destruction, and patient 8, who had no bone destruction, accurate prediction of the nature of the tumor is not always possible. The presence of obvious bone destruction, on the other hand, is generally associated with more aggressive tumors. One of the 2 patients in our series who had radiographic evidence of bone destruction had a malignant SFT. Interestingly, the most obvious destructive bone changes were present in patient 6, who had a benign tumor. Of the 3 pathologically proven malignant SFTs, only patient 7 showed a radiographic focus of bone destruction. The overall effect on the remaining bone in this tumor was of regressive remodeling. The other 2 malignant tumors showed only regressive remodeling of adjacent bone. The benign appearance of SFTs on bone window CT scans can therefore be misleading. Although the presence of bone destruction should prompt suspicion for a malignant tumor, the absence of bone destruction does not negate the possibility of malignancy.

The gross macroscopic appearance of a tannish-yellow mass with a smooth, glistening capsule and a cut surface showing a whorled appearance is typical, although not diagnostic, of SFTs. Histologically, they consist of a patternless arrangement of spindle cells in a collagenous background with a prominent vascularity that results in a hemangiopericytoma-like pattern. The diagnosis of SFTs is based on the characteristic microscopic appearance as reported by Chan²⁰ in conjunction with immunohistochemical features. CD34, CD99, and Bcl2 immunohistochemistry helps in the distinction of SFTs from other soft tissue tumors, including benign fibrous histiocytoma, dermatofibrosarcoma protuberosa, myofibroma, fibroma, and neurogenic tumors. CD34 is a marker for healthy endothelium and vascular neoplasms and has been found to stain primitive mesenchymal stromal cells and several mesenchymal tumors.²¹ All malignant SFTs and 77% of benign SFTs stained positive for the presence of CD34 in one study.²¹ Benign fibrous histiocytomas typically stain negative for the presence of CD34 and Bcl2. Neurofibromas and schwannomas may be reactive for CD34 and Bcl2, but these tumors also stain positive for the presence of S100. In addition, CD34 is weakly or focally positive. Myofibroma and fibromas stain strongly for the presence of vimentin, smooth muscle actin, and muscle-specific actin but stain negative for CD34. Dermatofibrosarcoma protuberosa is frequently CD34 positive but often Bcl2 negative. Solitary fibrous tumors can be difficult to distinguish from other vascular soft tissue tumors, such as hemangiopericytomas and synovial sarcomas. Like SFTs, hemangiopericytomas also stain positive for CD34. Some authors²²,²³ report that the presence of a basement membrane in hemangiopericytomas also allows them to be distinguished from SFTs. However, it is now generally accepted that the delineation of hemangiopericytomas as a separate entity from SFTs may become obsolete because their histopathologic features so closely resemble cellular areas of SFTs.²⁴ Differentiating SFTs from synovial sarcoma is aided by the fact that over 80% of synovial sarcomas have a specific chromosomal rearrangement, t(X;18)(p11.2;q11.2).²⁵,²⁶ which is demonstrable by cytogenetics, fluorescence in situ hybridization,²⁷ and reverse transcriptase–polymerase chain reaction looking for the SYT–SSX1 or SYT–SSX2 fusion transcripts.²⁸

An estimated 5% to 20% of thoracic SFTs may have malignant features, but malignant extrathoracic tumors are rare. The diagnosis of malignancy is based on both clinical features and histologic findings. Atypical histologic results include a lack of circumscription, nuclear atypia, hypercellularity, and a mitotic count of 4 or more per 10 high-power fields and necrosis. England et al²⁹ reported that most malignant fibrous tumors of the pleura had mitotic counts of more than 4 per 10 high-power fields, were larger than 10 cm in diameter, and were hemorrhagic and necrotic. England et al³⁰ and Witkin and Rosai³¹ also reported that the most important prognostic factor was resectability. In our study, only 3 cases were malignant (patients 7, 8, and 10). In patient 7, a malignant tumor was suspected at initial presentation because of the appearance of bone destruction on CT scan. This patient underwent an anterior craniofacial resection. She had positive resection margins and therefore received radiation therapy as postoperative adjuvant therapy. She is currently free of disease, although her follow-up is only 3 months’ duration.

Surgery is recognized as the treatment of choice. Factors that predispose to local recurrence in non–head and neck SFTs are a tumor diameter larger than 10 cm, the presence of a malignant component to the histologic findings, and microscopically positive surgical margins.³ The study by Gold et al³ also showed that positive margins correlated with a large tumor. As might be expected in the head and neck region, all tumors were smaller than 10 cm in diameter in our patients. However, positive resection margins were found in 3 patients. In contrast to tumors found in other anatomic locations, such as the trunk and lower extremities, tumors of the head and neck present a unique anatomic challenge by virtue of their relationship to adjoining structures. Therefore, this high rate of positive margins most likely reflects the tumor location rather than its biological features. One such patient (patient 10) experienced local recurrence, but not until 3 years after initial surgery, which suggests that local recurrence can be slow to occur. Patients with positive margins should therefore be closely followed up for several years. Gold et al³ also reported that a tumor larger than 10 cm in diameter was closely associated with the presence of a malignant component (P<.01). In our study, patient 7 had a 6 × 5-cm tumor and patient 8, a 6 × 3-cm tumor, both of which were subsequently found to be malignant. Patient 10 also had a malignant tumor, but it was much smaller, 2 × 2 cm. Thus, the size of a tumor in the head and neck region may not be as important in the progression to malignancy. However, because of the small number of cases, it is not possible to make any meaningful conclusions with regard to malignant phenotype and tumor size.

Solitary fibrous tumors of the head and neck region are extremely rare. The characteristic features of these tumors seen on CT or MR imaging in conjunction with a benign, slow-growing clinical history may help in the diagnosis, but definitive diagnosis is usually made only
after tumor resection. These tumors can be safely surgically excised, and patients who undergo complete surgical resection and do not have any malignant component can expect a favorable outcome. However, patients with positive surgical margins or whose tumors have a malignant component benefit from adjuvant postoperative radiation therapy and require close follow-up because they are at increased risk for local recurrence.

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