Solitary fibrous tumor of the parapharyngeal space

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Abstract

Solitary fibrous tumors are benign neoplasms of mesenchymal origin. They usually arise from the visceral or parietal pleura and peritoneum, although they have been found in many areas throughout the body. We report a case of solitary fibrous tumor of the parapharyngeal space. Microscopically, the tumor contained spindle cells with areas of marked hypercellularity without a definite pattern. Consistent with a benign lesion, there were few mitoses and no necrosis. The tumor cells stained strongly positive for CD34 and vimentin. At the 2-year follow-up, the patient was well and free of local and/or distant disease.

Introduction

Solitary fibrous tumors are rare spindle-cell neoplasms that usually arise from visceral and parietal pleura and peritoneum. Although they generally originate in serosa-lined tissue, these tumors can be found anywhere mesenchymal tissue is located, including the head and neck. Reported head and neck sites have included the parapharyngeal space,\(^1\) parotid gland,\(^2\) oral cavity,\(^3-7\) orbit,\(^8,9\) and larynx.\(^10\) Other sites have included the chest, arms, back, abdominal wall, liver, spinal cord, retroperitoneum, and adrenal gland. Epidemiologically, solitary fibrous tumors usually arise between the fourth and eighth decades of life; there is no predilection for either sex.\(^7\) They are typically slow growing, painless, well circumscribed, and nontender. Because of the relatively indolent and typically benign course of these tumors, patients may delay seeking medical attention for years. However, approximately 10 to 15% of tumors have exhibited malignant features, such as recurrence and distant metastasis.\(^7\)

The description of solitary fibrous tumors is somewhat confusing, as they have been previously reported under many names. The first reported tumor was described by Klemperer and Rabin in 1931 as a localized fibrous mesothelioma.\(^11\) Since then, other terms that have been used include submesothelioma, pleural fibroma, and hemangiopericytoma. The variety in nomenclature is a reflection of the presumed histologic origin of the deranged cells, the spectrum of the structural and the ultrastructural features of this neoplasm, and the broad differential diagnosis.\(^12-15\)

In this article, we report a new case of solitary fibrous tumor of the parapharyngeal space.

Case report

A 25-year-old man presented with a progressive, dull, right pharyngeal and mandibular pain of approximately 18 months’ duration. Facial asymmetry was evident (figure 1, A). Findings on bimanual examination were unremarkable. Computed tomography and magnetic resonance imaging (MRI) identified a 6.5 × 5.5-cm tumor in the right parapharyngeal space (figure 1, B). The mass had caused bowing of the ramus of the mandible and erosion of the posterior surface of the right maxillary sinus. It extended into the masticator space between the ascending ramus of the mandible and the pterygoid musculature.

In the operating room, a midline mandibulotomy with paralingual extension was performed to access the tumor. The medial and lateral pterygoid muscles were transected, at which point the tumor came into view (figure 2, A). The mass was freed from the mandible and deep surface of the parotid and dissected to the base of skull. The vascular supply, which emanated from the internal maxillary artery, was clipped and divided. The entire tumor was then removed. The resected specimen was smooth, encapsulated, tan-pink, and multinodular; it measured 8.3 × 5.6 × 4.5 cm (figure 2, B). Primary reconstruction of the neck was performed, and the mandible was repaired with a preshaped, six-hole titanium plate (figure 2, C). The patient tolerated the procedure well, and his postoperative course was routine.

Serial sections revealed a homogeneous tan-pink surface with scattered tan-yellow areas. Microscopic examination
revealed diffuse areas of hypercellularity, with some areas displaying variations in cellularity. The pleomorphism was associated with a patternless architecture, branching pericytoma-like vessels, and bland spindle-cell morphology (figure 3, A). There were few mitotic cells (<3 mitoses per 10 high-power field) and no necrosis. Immunohistochemistry showed reactivity to CD34 and vimentin (figure 3, B). There was no reactivity to keratins AE1/AE3, S-100 protein, smooth-muscle antibody, actin, and desmin. The morphologic and immunohistochemical features of this mass were highly consistent with the diagnosis of solitary fibrous tumor of the parapharyngeal space.

At follow-up 18 months postoperatively, the patient was free of tumor both clinically and radiologically, and the cosmetic result was excellent (figure 4). He remained free of disease at the 2-year follow-up.

Discussion
Hemangiopericytoma is the tumor that is most difficult to distinguish from solitary fibrous tumor because of its similar clinical and morphologic profiles. Some authors maintain that these two tumors may represent a single nosologic entity, others believe that they may exist along a continuum, and still others have proposed that they are distinct entities.12-15

Some authors have attempted to differentiate solitary fibrous tumor from hemangiopericytoma on the basis of histologic and immunohistochemical characteristics.5 Given the significant clinical implications, distinguishing between the two is imperative. But again, a distinction can be made only after intense scrutiny because the histologic and immunohistochemical differences are very fine.

The histogenesis of solitary fibrous tumor has been the subject of much controversy and debate. Two theories of its origin have been proposed. One theory15 implicates the multidirectional differentiation of fibroblasts, while the other16 is based on the presence of a specialized cell that is capable of differentiation into surface mesothelium. Current evidence favors a mesenchymal origin of this tumor on the basis of immunohistochemical findings.13

Macroscopically, solitary fibrous tumors are well circumscibed, uniform or nodular, soft to rubbery, and gray-whitish with whorls and streaks of fibrous tissue evident on the cut surface; the whorls and streaks can be very wide. The color and texture of the overlying mucosa are normal.

Goldsmith et al described solitary fibrous tumors cytologically as bland spindle-shaped cells with varying cellularity, variable dense collagenization, and prominent vascularity.15 The cells are arranged in no obvious pattern; focal storiform, fascicular, and herringbone patterns with nuclear palisading may all be observed. The tumor cells are often separated by thick collagen bands that demonstrate focal areas of keloid-like hyalinization. There is prominent vascularity, ranging from narrow vascular clefts to a hemangiopericytoma-like pattern admixed with areas of sclerosis; vessels with thick, hyalinized walls are often noted. Hemangiopericytomas, on the other hand, are...
characterized by a proliferation of monotonous spindle cells with no significant variation in cellularity and with a prominent staghorn-like vascular pattern.

Regardless of location, solitary fibrous tumors have a characteristic immunophenotype. The hematopoietic progenitor-cell antigen CD34 is present in normal and neoplastic endothelial cells, and it is strongly positive in most cases of solitary fibrous tumor. CD34 is a transmembrane glycoprophosphoprotein (molecular weight: ~110 kd) that is expressed on hematopoietic stem cells, small-vessel endothelial cells, and embryonic fibroblasts. The elaboration of this antigen suggests a vascular endothelial origin or induction of vascular structure formation by undifferentiated mesenchymal cells. Hemangiopericytomas may also be CD34-positive, but the pattern of reactivity is usually patchy and its intensity is much weaker.

Another difference between solitary fibrous tumors and hemangiopericytomas is the presence of mast cells in the former and their absence in the latter. Mast cells arise from CD34-positive pluripotential cells; it has been speculated that their presence can be explained by the cell-rich, CD34-positive makeup of solitary fibrous tumors.15 Positive immunostaining for Bcl-2 and CD99 is also observed in solitary fibrous tumors, but their presence is not uniform and positivity is often weak. Hemangiopericytomas do not stain for Bcl-2, and their pattern for CD99 is unknown. The presence of alpha-smooth-muscle actin in the wall vessels of solitary fibrous tumors is a distinguishing characteristic of this lesion; hemangiopericytomas do not contain this substance. Finally, solitary fibrous tumors are immunoreactive for mesenchymal markers such as vimentin and negative for desmin, epithelial markers (cytokeratin), vascular markers (factor VIII-related antigen), and neural markers (S-100 protein).6 Solitary fibrous tumors generally behave in a benign manner and do not metastasize. However, some reports in the literature have described patients with solitary fibrous tumors who developed recurrent disease with malignant characteristics, including distant metastasis.17 As mentioned, recurrence or distant metastasis has been reported in 10 to 15% of these tumors.7 Indicators of malignant potential include the presence of more than 4 mitoses per 10 high-power field, abnormal mitotic features, cellular pleomorphism, and tumor giant cells. A careful microscopic evaluation of cellularity, pleomorphism, and mitotic activity is important, although it does not reliably predict the clinical behavior of these tumors.

The mainstay of treatment is surgical resection. Several reports have described the use of neoadjuvant chemotherapy with doxorubicin and dacarbazine along with postoperative radiation therapy in cases of large tumors with positive surgical margins after resection.17 If the histologic evaluation suggests malignancy, diligent surveillance is crucial. Some recurrences and metastases have been reported years

Figure 2. **A:** The tumor is seen through the paralingual extension. The blue vessel loop encircles the lingual nerve. **B:** The excised mass measures 8.3 × 5.6 × 4.5 cm. **C:** The mandibulotomy is closed with the titanium plate.
The most important factor in the prognosis of patients with a solitary fibrous tumor is the completeness of the surgical resection.

References