Solitary Fibrous Tumor of the Auditory Canal

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• Solitary fibrous tumor (SFT) is an uncommon spindle cell neoplasm of increasing incidence that was originally described to be of pleural origin; however, more recently, SFT has been reported in extrapleural sites, including the orbit, liver, salivary glands, tongue, nose, paranasal sinuses, larynx, retroperitoneum, meninges, and thyroid. The increase in the number of SFTs does not necessarily mean increased incidence of this tumor but rather an increased understanding of this tumor, especially recognition of this tumor in extrapleural locations, which has been aided by immunohistochemical analysis. We report a case of SFT in the auditory canal, which to our knowledge has not been previously reported, as evident by morphologic findings and immunophenotype.

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REPORT OF A CASE

A 39-year-old woman with no significant past medical history presented with a 2-year history of decreased hearing in the left ear associated with mild pain and drainage. A hearing aid dealer referred her for evaluation after noting a masslike lesion in the left ear. Physical examination revealed an occlusion of the left ear canal by an aural polyp. Laboratory findings were normal. Coronal computed tomographic scan images obtained through the temporal bones demonstrated a soft tissue mass that expanded at the bony margins of the left external auditory canal without destroying it (Figure 1). An opacification was evident within the mastoid air cells. A tympanomastoidectomy was performed to remove the polyp.

During surgery, the bony canal appeared to have been widely eroded posteriorly and superiorly, lateral to the tympanic membrane. The mastoid cavity was filled with inflammatory-type tissue that did not appear to be frank cholesteatoma. The polyp appeared to be attached in the region of the tympanic membrane. The polyp was amputated and submitted for frozen section evaluation. A large perforation of the posterior portion of the tympanic membrane was noted after removing the polyp.

On gross examination, the tumor was well circumscribed, ovally shaped, and measured 1.7 × 1.1 × 0.6 cm. It had a tan-white, firm, elastic appearance, with a tan, homogenous cut surface. No lobulations or calcifications were identified. No hemorrhage or cystic changes were seen. A frozen section was used to make a diagnosis of benign spindle cell lesion, with a differential of perineurioma, schwannoma, and fibroma. On permanent sections of formalin-fixed tissue, the lesion was microscopically composed of spindle cell proliferation with focally myxoid stroma covered by squamous mucosa. The spindle cells were proliferating in a haphazard fashion with focal whirling. Few dilated vascular clefts and inflammatory cells were identified (Figure 2, a through c). No significant cytologic atypia, necrosis, or mitotic activity was seen. Immunophenotypically, the tumor cells were positive for CD34, CD99, and Bcl-2 (Figure 2, d through f). No immunoreactivity was noted with epithelial membrane antigen, S100 protein, smooth muscle actin, neurofilament, CD31, factor 8, and CD117. Based on these morphologic findings and immunophenotype, a final diagnosis of solitary fibrous tumor (SFT) was made.

COMMENT

Solitary fibrous tumor, previously known as benign or solitary mesothelioma, was thought to be of mesothelial origin, but the identification of SFT in extraserosal locations, along with its ultrastructural and immunophenotypic features, demonstrates that SFT has a mesenchymal rather than mesothelial origin.[1,2] The pleural SFTs mostly arise in the parietal pleura, some in the visceral pleura over the lung convexity and a small number in the pleura

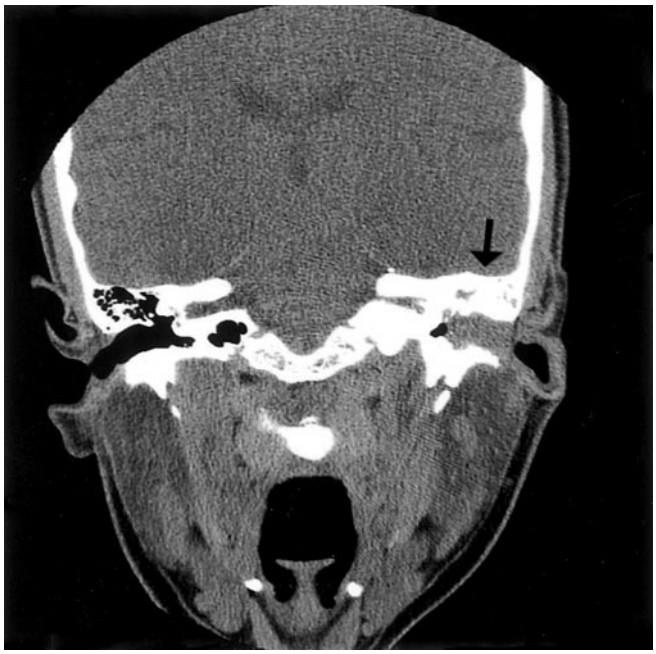

Figure 1. *Coronal computed tomographic scan that shows a soft tissue mass (arrow) that expands at the bony margins of the left external auditory canal.*

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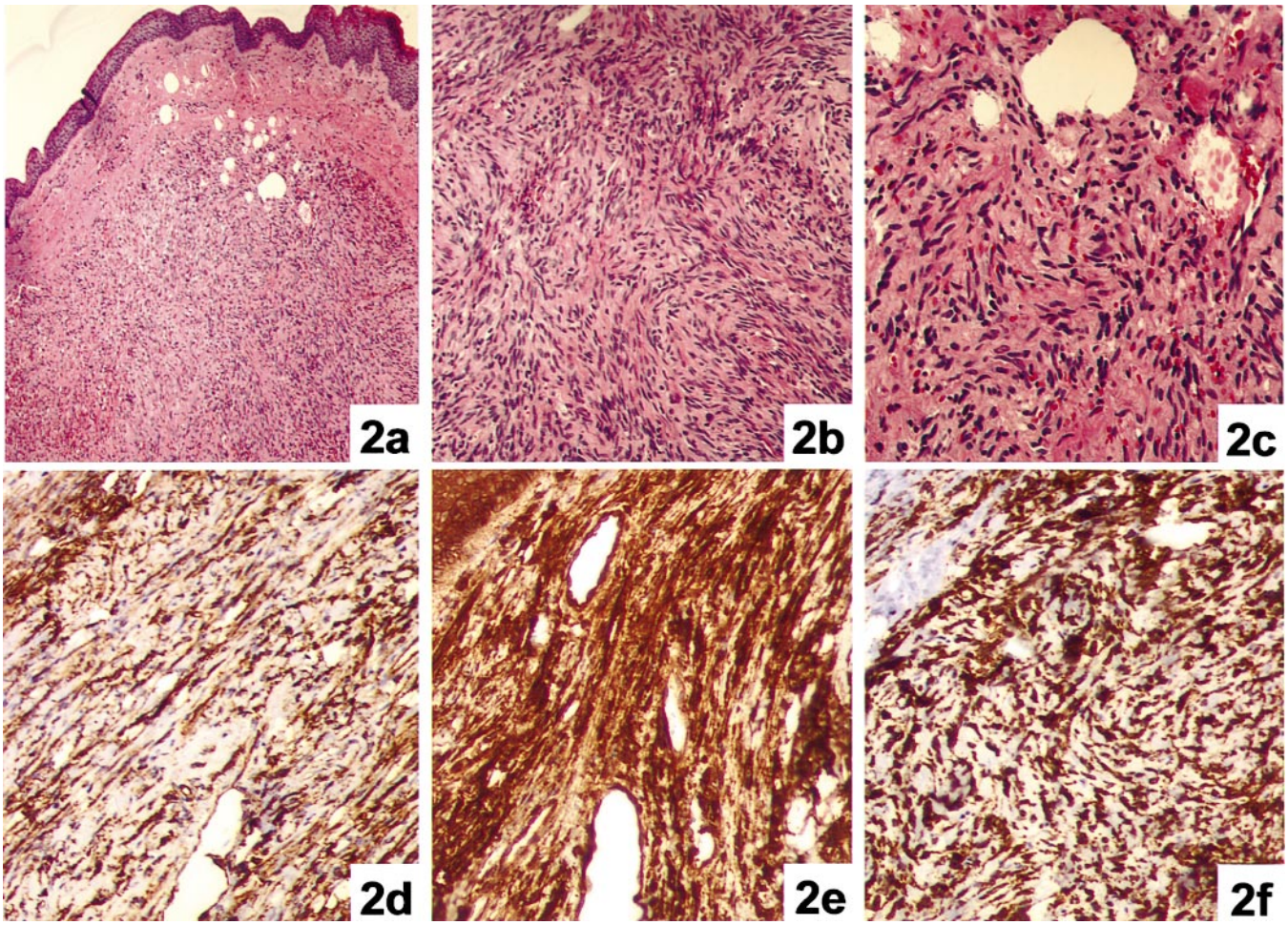

Figure 2. *Histologic section that shows a polypoid spindle cell lesion covered by stratified squamous epithelium (hematoxylin-eosin, original magnification ×10) (a), which in some areas shows a fascicular and whorled pattern (hematoxylin-eosin, original magnification ×20) (b) and vascular clefts (hematoxylin-eosin, original magnification ×40) (c). On immunohistochemical analysis, the tumor cells are positive for CD34 (d), CD99 (e), and Bcl-2 (f) (immunoperoxidase stain, original magnification ×20).*

within one of the interlobar fissures. Extrapleural tumors have been recently reported in many sites, such as in orbit,[1,2] liver,[1,2] salivary glands,[1,2] parotid gland,[3] retroperitoneum,[1] meninges,[1] oral cavity,[4] nose,[4] tongue,[1,5] paranasal sinuses,[4,6] mediastinum,[2,7] hypoglossal nerve,[8] breast,[9] larynx,[10] and thyroid.[1]

Solitary fibrous tumor is a slow-growing tumor that most often occurs in the fourth and fifth decades of life.[11] No sex preponderance has been established, although some studies have reported slight female predilection. Clinical symptoms depend on the size and location of the tumor, where symptoms are usually related to compression rather than infiltration of the adjacent tissue. Solitary fibrous tumor most often manifests as a benign lesion; however, malignant transformation and metastasis have been documented in a number of both pleural and extrapleural tumors.[1,4,12]

Because of the occasional aggressive behavior of this tumor, it should be considered potentially malignant and surgical resection performed.[1] The recurrence rate is very low after resection, with a slight increased risk of recurrence in the extrathoracic tumors. Prognostic factors for a malignant potential include high cellularity, mitotic activity (>4 per 10 high-power fields), pleomorphism, extensive hemorrhage, and necrosis. Also, tumors larger than 10 cm should be closely monitored.

Morphologically, SFT consists of proliferating ovoid or spindle-shaped cells that are arranged haphazardly, often in a fascicular and whorled pattern, in a stroma that can have myxoid or dense areas with collagen deposition. The so-called patternless pattern refers to the haphazardly arranged spindle cells.[1–5] Inflammatory infiltrates and vascular clefts are usual findings. Mild nuclear atypia and few mitotic figures can sometimes be seen and do not denote a malignant potential. Immunohistochemically, SFT cells are positive for vimentin, CD34, Bcl-2, and CD99.[1,13,14] Smooth muscle actin may be negative or show focal variable staining.[1,13,14] They are negative for S100 protein, epithelial membrane antigen, cytokeratin, CD31, and neurofilament.[1,13,14] The broad range of morphologic features shared with other spindle cell neoplasms, along with the unusual extrapleural locations of the tumor, can make the diagnosis of SFT at times challenging.

In the head and neck region, SFT has been previously reported in the oral cavity, tongue, nasal cavity, paranasal sinuses, orbit, larynx, salivary glands, and thyroid,[1–4,14] but to the best of our knowledge SFT has not been previously reported in the auditory canal. In our case, the unusual

and unreported auditory location of the tumor made SFT not the primary candidate in the initial differential diagnosis at frozen section. A diagnosis of schwannoma, fibroma, or perineurioma was first entertained on cryostat sections. However, after correlating the morphologic features on permanent sections of formalin-fixed tissue and the immunophenotypic results, SFT was the most appropriate diagnosis. Although our case did not have histologically malignant potential, follow-up was recommended due to the unpredictable behavior.

In conclusion, we report a case of SFT occurring in the auditory canal, which, to our knowledge, has not been previously reported. Solitary fibrous tumor must be considered in the differential diagnosis of spindle cell lesions of the auditory canal, and immunohistochemical analysis can be of great help in their diagnosis.

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