A SOLITARY FIBROUS TUMOR IN THE EXTERNAL AUDITORY CANAL- A RARE CASE REPORT

Rabindra Kumar Khatua¹, Anasuya Lenka², Alok Das³

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ABSTRACT: This paper reports a rare case of solitary fibrous tumor(SFT) arising independently from the external auditory canal. A 16-year-old female patient presented with mass in left external auditory canal and aural fullness. Physical examination revealed a large papillomatous mass coming out from left external auditory canal. The patient underwent surgical removal of tumor. A light microscopic study showed a spindle-cell proliferation with a collagenous stroma and a staghorn-like vascular pattern. The tumor cells were immune histochemically positive for vimentin. One week after resection aural fullness had resolved. Solitary fibrous tumor is a rare mesenchymal tumor which originates in the pleura or at virtually any site in the soft tissue. It has been observed that 78% to 88% of SFT's are benign and 12% to 22% are malignant. En-bloc resection remains the cornerstone of therapy. Although, several cases of extra pleural SFT have been reported, only one case of external auditory canal SFT has been reported so far. Excision of SFT of the external auditory canal is advisable, especially in the presence of symptoms, and should be preceded by confirmation of non-malignancy by biopsy, if possible.

KEYWORDS: Solitary fibrous tumor; Mesenchymal tumor; External auditory meatus; Immunohistochemistry; vimentin.

INTRODUCTION: Solitary fibrous tumor was first mentioned in the scientific literature by Wagner¹. The first discussion of its clinical and pathological properties was by Klemperer and Rabin². SFT is a rare mesenchymal tumor originating in the pleura² or at virtually any site in the soft tissue. It has been observed that 78% to 88% of SFT's are benign and 12% to 22% are malignant. In the head and neck area, there are many case reports on SFT of meninges⁴, orbit⁵, hypopharynx, parotid gland⁶ and only one SFT arising in the entrance of left external auditory canal⁷ has been reported in the literature. Therefore, we present this very rare case of large SFT in external auditory canal which has been treated by us recently.

CASE REPORT: A 16-year-old female patient presented to our OPD with complaints of mass in left external auditory canal and aural fullness. Physical examination revealed a large polypoidal mass coming out from left external auditory canal (figure-1). CT scan demonstrated a relatively homogeneous, low-density mass in the external auditory canal without invasion of adjacent structures. The rest of the ENT examination and results of blood tests yielded normal findings. Surgical excision of tumor was done under local anesthesia. Tumor was found to be attached to the posterior wall of external auditory canal. Microscopic examination of ear revealed intact tympanic membrane with no other obvious abnormalities.

Gross examination of specimen revealed a firm, polypoidal mass measuring 4.5 cm in diameter. Cut section of the tumor was pale and firm (figure-2). Histopathological investigation
revealed pattern less proliferation of bland spindle cells with a collagenous stroma associated with thin walled branching vascular spaces (figure-3). The tumor cells were reactive to vimentin in on immunohistochemistry (figure-4).

One week after resection of the mass, the postoperative wound at the external auditory canal had almost completely epithelialized and the chief complaint of aural fullness had resolved. No obvious evidence of recurrence has been seen at the resected area for a postoperative follow-up period of 10 weeks.

**DISCUSSION:** In previous studies, SFTs have been reported mainly in the pleura but SFTs in extrapleural sites have been rarely recognized. Very few cases in head and neck location and only one in external auditory canal have so far been reported. SFT was first mentioned in the scientific literature by Wagner. Nearly 61 years after that in 1931, the first discussion of its clinical and pathological properties was done by Klemperer and Rabin. Solitary fibrous tumor is a rare mesenchymal tumor originating in the pleura or at virtually any site in the soft tissue. Currently, however, most authors agree that they originate from submesothelial connective tissue. This hypothesis is based on immunohistochemical and ultrastructural findings as well as due to the occasional occurrence of this tumor in extra pleural sites.

The radiological features of SFT are the presence of a tumor of tissue with a regular contour sometimes consisting of calcifications or necrotic foci. If the contrast enhancement is low, then a benign course may be suggested. On MRI, SFT has been reported to show low signal intensity on the T1-, T2-weighted images correlated with their fibrous nature. The high-intensity central areas on T2-weighted images reflected ischemic necrosis or focal perivascular edema.

The macroscopic presentation of our case corresponds to its classical description in previous studies in the literature; firm, well-circumscribed and gray-whitish. SFT has been recently considered to originate from the mesenchymal cells of the submesothelial connective tissue of the pleura. According to immunohistochemical analysis, SFT of the pleura is positive for vimentin, CD34, CD99, and Bcl2, which are markers of mesenchymal cells; but it is negative for cytokeratin, which is found in mesotheliomas. These results indicate that SFT originates from mesenchymal cells rather than mesothelial cells.

A histological examination generally demonstrates the presence of spindle-cells in a collagenous background with a characteristic "pattern less pattern". Prominent vascularity resulting in hemangiopericytoma-like foci is also frequently seen. Most tumors have a variable appearance with alternating relatively hyper- and hypo-cellular regions. These features suggested the diagnosis of a benign nonepithelial tumor e.g. neurogenic tumor, fibroblastic tumor or hemangiopericytoma. At present, strong immunoreactivity for CD34 monoclonal antibodies allows the distinction of SFT from most of the other neoplasms. The degree of CD34 immunoreactivity observed in the neurofibroma and schwannoma varies, however, they are usually strongly positive for S-100 protein in contrast to SFT.

The morphologic distinction between hemangiopericytoma and SFT may be quite difficult because the latter sometimes has extensive areas with a growth pattern virtually indistinguishable from that of hemangiopericytoma. The pattern of CD34 immunoreactivity, which is rarely observed in hemangiopericytoma, is identical to that of SFT. Most SFTs behave in a benign fashion but sometimes they may be locally aggressive. The prognostic factors correlating with an aggressive
behavior are a large size, hypercellularity, a high mitotic activity and necrosis. The histological criteria of the tumor in the present case were compatible with those demonstrating a benign nature, a small degree of mitotic activity and an absence of necrosis. Surgical excision is usually the treatment of choice for SFT. A long term follow up is required to ensure prompt attention if by chance there is recurrence of the tumor.

REFERENCES:
Histological findings. The neoplastic cells have round to oval nuclei and are arranged in haphazardly oriented fascicles. H&E staining. Original magnification, 10×.

Immunohistochemical staining. The neoplastic cells showed a positive reaction for vimentin were detected. In addition, the endogenous control of vascular endothelial cells was positive for both. Original magnification, 100×.