



## Introduction

Solitary fibrous tumour (SFT)/ haemangiopericytoma (HPC) is a rare, slow growing vascular tumour that arises from the pericapillary pericyte cells of Zimmermann<sup>1</sup>. It accounts for approximately 1% of all vascular tumours, and 13-25% of them presents in the head and neck region<sup>2</sup>. In the head and neck region, solitary fibrous tumour has been reported in the oral cavity, tongue, nasal cavity, paranasal sinuses, larynx, salivary gland, and thyroid<sup>3</sup>. To the best of our knowledge '5' such cases were published in the past, Izumaru et al<sup>4</sup> and Rezk et al<sup>5</sup> in 2004, Castillogone et al<sup>2</sup> in 2015, Lee et al<sup>6</sup> in 2016, and Rijo-Cedeno et al<sup>6</sup> in 2019. We report sixth case of solitary fibrous tumour/haemangiopericytoma in the external auditory canal of a 32-year-old male from Bangladesh.

## Case Report

A 32-year-old man visited in the Department of Otolaryngology – Head and Neck Surgery, Bangabandhu Sheikh Mujib Medical University, Dhaka, Bangladesh with the complaint of progressive painless swelling in the left ear canal, blockage sensation and decreased sensation of hearing for 7 months. He did not notice any discharge from the ear. A physical examination revealed a smooth surfaced, firm swelling seemed to be attached with the inferior and post wall of external auditory canal (EAC). It completely occluded the canal for which tympanic membrane could not be visualized. Prominent vascular marking was noticed over the swelling under the skin. Tuning Fork test showed Rinne negative the left ear and Weber lateralized to same side correlated with conductive hearing loss. Pure tone audiogram revealed 30.75dB ABG (air-bone gap) in 500,1000,2000 and 4000 Hz. Computed tomography (CT) showed a round, homogenous soft tissue mass involved the posterior and inferior wall of EAC and was enhanced with contrast. The lesion seemed to be eroded the bony EAC wall but there was no invasion of middle and mastoid or adjacent structures.

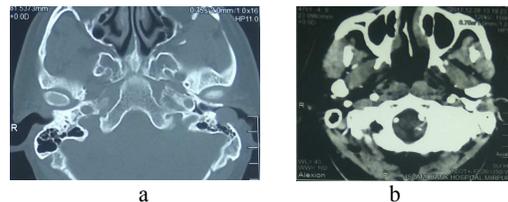


Fig.1 CT scan showed a. Isodense mass seems eroding posterior wall, b. Contrast enhanced mass

The tumour was excised completely with preserving the skin of EAC with a standard postauricular approach under general anaesthesia on 1st February 2018. The anterior and superior walls of the EAC, and tympanic membrane were intact and healthy. After removal of the tumour, an erosion was noticed in the tympanic part at the floor of EAC which was reconstructed with cortical bone chips and conchal cartilage.

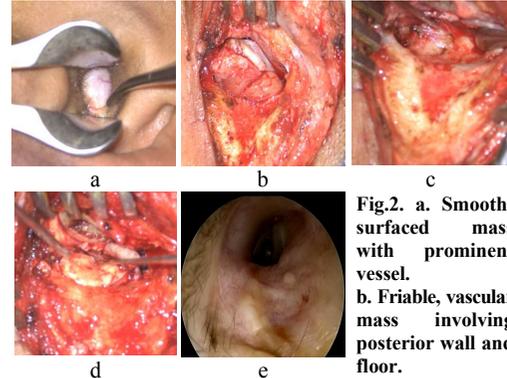


Fig.2. a. Smooth-surfaced mass with prominent vessel. b. Friable, vascular mass involving posterior wall and floor. c. Erosion of tympanic bone at the floor, d. Bony defect was filled with bone chips and conchal cartilage e. Healed floor and posterior wall

Macroscopically the tumour was grey-white in colour. Microscopically the tumour was composed of sheets of cells with uniform, monomorphous spindle cell with a round to oval nuclei. These cells showed moderate pleomorphism with a moderate amount of eosinophilic cytoplasm and indistinct cytoplasmic borders. Intratumoral staghorn type well developed, branching, thick-walled blood vessels were present. The focal area showed storiform pattern. Mitoses were infrequent. Based on these findings the tumour was diagnosed as (solitary fibrous tumor/ haemangiopericytoma)-grade II.

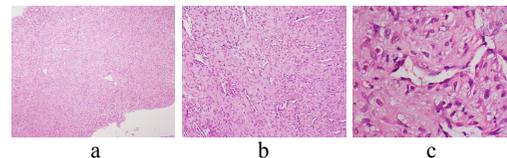


Fig.3. Histopathological (a) Sections show a cellular neoplasm, the stroma shows intratumoral staghorn type well developed, branching, and thick walled blood vessels. Focal area shows storiform pattern (H&E, X40). (b) The tumor cells are spindle in shape and distributed in a prominent vascular network (H&E, X100). (c) The neoplastic cells are uniform, monomorphous spindle cell with round to oval nuclei. These cells show moderate

pleomorphism with moderate amount of eosinophilic cytoplasm and indistinct cytoplasmic borders. Mitoses are infrequent (H&E, X400).

The patient passed an uneventful postoperative course. The complete healing of the external auditory canal was achieved within 2- 3 months. He got complete relief of symptoms with the improvement of hearing. The patient was in regular follow-up in every 5-6 months interval. The last follow-up otoendoscopy and audiogram recorded on 25th July 2021. The follow-up pure tone audiogram showed 6 dB ABG (air-bone gap) in 500,1000,2000 and 4000 Hz. There is no recurrence seen after 41 months of follow-up.

## Discussion

Solitary fibrous tumour (SFT)/ haemangiopericytoma (HPC) usually presents between fourth and fifth decade with no sex preponderance<sup>3,5</sup>. Clinical symptoms and signs depend on site of origin and extension of the lesion<sup>3</sup>. In case of external auditory canal SFT/HPC, progressive swelling with conductive hearing loss is the predominant clinical presentation. Pain and otorrhoea may be due to otitis externa developed by self-cleaning or surface ulceration.

Radiological features of SFT/HPC on CT and MRI cannot differentiate it from other benign soft tissue neoplasm such as angioma, meningioma, fibroma, chordoma etc. CT scan of SFT demonstrates as isodense to hyperdense depending on the collagen content. The characteristic features of SFT at MRI are the presence of low-signal -intensity foci on T1- and T2- weighted images, corresponding to the collagen content. SFT are highly vascular and are avidly enhancing on both CT and MR images. This combination of features produces a chocolate chip cookie appearance, which can help in diagnosis<sup>7</sup>. But if there is increased contrast enhancement, possibility of malignancy is higher. The CT scan of the current case revealed contrast enhanced lesion involving floor and posterior wall with a bony defect at the tympanic bone. The bony defect might be due to resorption of bone by expansion of the tumour.

Immunohistochemically, cells in SFT/HPC are reactive with CD34, vimentin, and Bcl-2. In addition, STAT6 nuclear immunoreactivity is the most diagnostic marker for SFT<sup>8</sup>.

SFT/HPC are usually benign in nature. Although, approximately 15-20% of SFT are malignant, SFT/HPC in the head and neck are more likely to be benign than are SFT elsewhere, and have a good prognosis<sup>7</sup>.

Complete resection is the treatment of choice for both benign and malignant SFT/HPC. If the post-op

histopathological report confirmed malignancy or positive margin, adjuvant radiotherapy is recommended. Prognostic factors of malignant tumour are related with high cellularity, increased mitotic activity (>4 per 10 high-power fields), pleomorphism extensive haemorrhage and necrosis<sup>3</sup>. Considering these criteria, our reported case is benign (grade-II) in nature.

Though no recurrence is demonstrated in current case after 41 months of follow-up, long term follow-up (at least 10 years) is recommended to detect late recurrence in a case with unpredictable behaviour.

## Conclusion

Though rare in presentation, external auditory canal SFT/HPC may be considered as a differential diagnosis of any smooth-surfaced slowly growing lesion seems to arise from EAC. Even with modern imaging technique, differentiation of SFT/HPC from other benign soft tissue neoplasm such as angioma, meningioma, fibroma, chordoma etc is quite difficult. In addition to CD34, vimentin, and Bcl-2, STAT6 nuclear immunoreactivity is the most diagnostic marker for SFT. Because of unpredictable behaviour, after complete resection, benign SFT/HPC should be followed up for at least 10 years to notify late recurrence.

## References

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