

## ***Case Report***

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# **Benign fibrous histiocytoma of the external auditory canal: Case report and literature review**

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### **Abstract:**

**Background:** The benign fibrous histiocytoma (BFH) is a mainly dermal neoplasm composed of a mixture of fibroblastic and histiocytic cells. It is usually reported as occurring on the extremity, retroperitoneum or orbit but not commonly in the other head and neck regions.

**Methods:** A ten-year old girl presented with a right external auditory meatal mass for 6 months. Clinical examination showed a small, firm non-tender swelling on the antero-inferior part of the cartilagenous external auditory canal. An excisional biopsy was performed without complication.

**Results:** Clinical and histopathological examinations with immunohistochemical studies were consistent with the diagnosis of BFH of the external auditory canal. There was no local recurrence to date.

**Conclusions:** BFH is an uncommon benign neoplasm of the head and neck and none has been previously described occurring in the external auditory canal. Complete local excision is the mainstay of treatment with excellent prognosis.

**Key words:** *Benign fibrous histiocytoma; Dermatofibroma; Atypical fibrous histiocytoma; Fibrohistiocytic neoplasm; External auditory canal.*

### **Introduction:**

Benign fibrous histiocytoma (BFH) or dermatofibroma, is a benign neoplasm of fibroblasts and histiocytes in the soft tissues of the dermis and subcutaneous layers. It

generally presents as a slow growing solitary papule or nodule on the lower extremities of young to middle-aged adults. From review of the English medical literature there are a number of reported cases of benign fibrous histiocytoma of the head and neck. To the knowledge of the authors, this is the first reported case of a benign fibrous histiocytoma of the external auditory canal.

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### **Case report:**

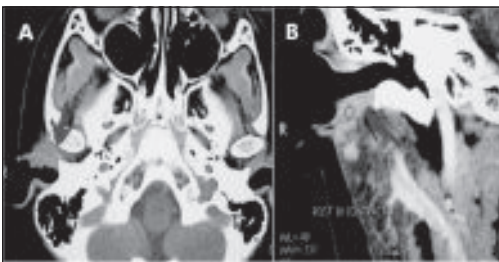
A ten-year-old girl presented with a right external auditory meatal mass for 6 months. There was no associated otalgia, otorrhea or impaired hearing. There was no preceding history of trauma or repeated infections.

Clinically there was a small, firm non tender swelling on the antero-inferior part of the cartilagenous external auditory canal measuring about 1.0 x 1.0cm. The overlying skin was normal (Figure 1).



**Figure 1:** A swelling on the antero-inferior cartilagenous part with normal overlying skin.

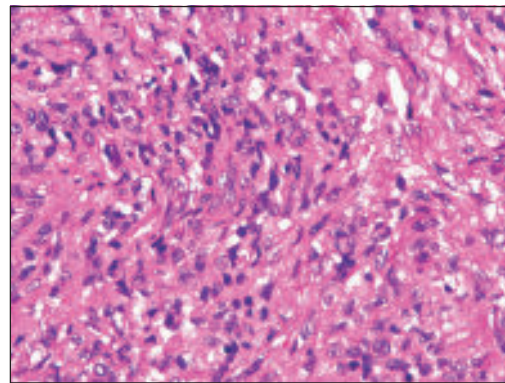
The mass obscured the view of the tympanic membrane. There was no evidence of cervical lymphadenopathy. Pure tone audiometry showed normal hearing. Computed tomography showed a superficial moderately enhancing mass in the ear canal with no involvement of the surrounding structures (Figures 2A & 2B).



**Figure 2:** A) Axial section, (B) Coronal section of a contrasted computed tomography, showing soft tissue mass in the cartilagenous ear canal wall (red arrows) with no infiltration into the surrounding structures.

The patient underwent excision of the mass under general anesthesia. Intra-operative examination showed that the external auditory meatal mass extended medially until the bony-cartilagenous junction while the lateral extent was the cartilagenous concha. The tympanic membrane was normal. The mass did not involve the underlying cartilage. The surgical plane between the mass and parotid gland was normal. The mass was removed in toto with skin. A split skin graft harvested from the right upper arm was laid over the exposed cartilage and dressed with rosette silk.

Histopathological examination demonstrated tumor within the dermis with overlying stratified squamous epithelium which was composed of spindle-shaped and rounded cells with coarse collagen bundles haphazardly arranged between the tumor cells. Scattered lymphocytes were also present (Figure 3). Immunohistochemical studies showed positivity for vimentin but were negative for S-100, SMA, CD34 and Desmin. A final diagnosis of dermatofibroma was confirmed.



**Figure 3:** Hematoxylin and Eosin (x 60). Tumor composed of spindle shaped and rounded cells with coarse collagen bundles haphazardly arranged in between.

The patient had an uneventful postoperative recovery. She was followed up regularly and at 12 months follow-up, the patient was well with no evidence of recurrence.

**Discussion:**

Benign fibrous histiocytoma (BFH) is a benign tumor composed of a mixture of fibroblastic and histiocytic cells. It is mainly a dermal neoplasm but can also be found in soft tissue and parenchymal organs.

The commonest sites affected by BFH are the soft tissues of the lower extremities (50%), less frequently in the upper extremities (20%), retroperitoneum (20%) and orbit<sup>1</sup>. BFH occurring in the head and neck is not common. In a review of recent English medical literature, the sites of where BFH have been found to occur was in the oral cavity (buccal mucosa, gingival or alveolar ridges, soft palate and floor of mouth), upper and lower lips, mandible, maxilla, nasal cavity and paranasal sinuses<sup>2</sup>. Other sites include the temporomandibular joint, the submandibular and parotid glands, the larynx, trachea and supraclavicular fossa<sup>2,3</sup>. To the knowledge of the authors, this is the first reported case of BFH occurring in the external auditory canal.

A large case series found that the typical patient was more frequently female than male, 40 years of age with a lesion on the lower extremities located in the dermis, often with infiltration of the subcutaneous fat tissue<sup>4</sup>. Another review of literature found the ages of patients ranged from 1 to 70 years with an average age of 37 years but the male to female ratio was 2.5:1. This tumor usually develops as a painless mass where specific symptoms arising once there is interference with the normal anatomy and physiology of the area in which it is found. This tumor has been associated with preceding trauma, sun exposure and chronic infection, suggesting a reactive proliferation of benign cells<sup>3,5,6</sup>. Our patient presented with an external auditory canal painless mass without other associated symptoms.

The most widely used classification for histiocytic lesions is based on clinical

histopathological criteria, the two main categories being benign and malignant (Table 1). This classification is based on the age of the patient, location of the lesion and biological behavior<sup>7</sup>. Atypical fibrous histiocytoma is a clinically benign lesion with malignant pathologic features, occurring as a solitary, ulcerated nodule of facial skin in fair-haired, blue-eyed persons in their seventh decade of life<sup>7</sup>. Inflammatory fibrous histiocytoma affects the internal organs and body cavities with a diffuse neutrophilic response, commonly occurring in patients in the sixth decade of life<sup>3</sup>. It behaves as a low-to intermediate-grade sarcoma whereas malignant fibrous histiocytoma acts as a high grade sarcoma. Aggressive oncologic management is required for optimal treatment of patients with the malignant variants of fibrous histiocytoma, including radiation therapy, chemotherapy and radical surgery.

BFH has been described macroscopically as appearing very well circumscribed and the cut surface ranged from pale to yellowish brown in color with a range of maximum diameter of two to 12cm (mean 5.3; median 4.0)<sup>6</sup>.

Histologically, BFH is composed of a biphasic cell population of histiocytes and fibroblasts. There are spindle-shaped cells with elongated nuclei arranged in a storiform pattern with rich vascularization, varying numbers of giant cells and xanthomatous cell (lipid-laden foamy histiocytes)<sup>3</sup>. However, another author reported from his series that secondary elements such as giant cells or xanthoma cells were usually lacking and the vascular pattern were commonly indistinct<sup>6</sup>. Positivity for CD68 and vimentin on immunohistochemical staining can demonstrate histiocytic cells and fibroblast-like cells respectively. Negativity for SMA and S-100 could correspondingly differentiate the tumor from leiomyosarcoma and neurogenic tumours<sup>5</sup>.

The most important diagnostic distinction is the separation of this tumor from aggressive forms of fibrohistiocytic neoplasms such as dermatofibrosarcoma protuberans and MFH. In contrast to BFH, MFH is composed of malignant pleomorphic sarcomatous cells, bizarre giant cells and frequent mitotic figures. It often demonstrates perineural, vascular, mucosal or bony invasion as well as distant metastases<sup>8</sup>. Dermatofibrosarcoma protuberans also occurs in the dermis and subcutis but is more prone to show extensive subcutaneous involvement compared to BFH<sup>5</sup>. Its margins are infiltrative in contrast to the well-defined margins of BFH. Immunohistochemically it is positive for CD34 while BFH is not.

The treatment of choice is local excision without sacrificing structures that would cause major functional or cosmetic morbidity. Of the cases with follow-up reported in the literature, 11% had a recurrence after local excision. The reason for these recurrences is unknown, as is the adequacy of the margins of resection. Incomplete excision or enucleation may result in a recurrence. Radiation therapy or chemotherapy has no role in the management of BFH<sup>3</sup>. These tumors have no metastatic potential and generally have good prognosis.

In conclusion, BFH is an uncommon benign neoplasm of the head and neck and in the English medical literature; none has been previously described occurring in the external auditory canal. Complete local excision is the mainstay of treatment with excellent prognosis.

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