

Solitary Neurofibroma of External Aural Canal

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How to cite this paper: Ndiaye, C., Diouf, M.S., Dieye, A., Maiga, S., Ahmed, H., Mbaye, A., Ndiaye, M., Younes, H., Diom, E.S. and Tall, A. (2023) Solitary Neurofibroma of External Aural Canal. *International Journal of Otolaryngology and Head & Neck Surgery*, 12, 444-448.

<https://doi.org/10.4236/ijohns.2023.126047>

Received: October 2, 2023

Accepted: November 27, 2023

Published: November 30, 2023

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Abstract

Neurofibroma is a benign tumor of the connective tissue of the peripheral nerves, developed mainly at the endoneurium. The most common localizations are the extremities of the limbs and the head and neck region. Neurofibromas are often associated with neurofibromatosis type 1. An isolated localization in the auditory canal is exceptional. We report the case of a 45-year-old female patient presenting with a neurofibroma of the auditory canal which had previously caused hearing loss. Examination revealed a tissue mass firm, painless and covered with normal skin obstructing the external auditory canal. The patient's skin examination revealed no café-au-lait spots. A CT scan of the ear showed a hypodense tissue mass. Surgical removal via the ear canal gave good results. In conclusion, an isolated neurofibroma of the external auditory canal is a rare benign tumor with a good prognosis. The surgical approach depends on the exact location of the mass and the surgeon's experience.

Keywords

Tumor, Ear External Canal, Hearing Loss

1. Introduction

Neurofibromas are benign tumours of the connective tissue of peripheral nerves, arising mainly of the endoneurium [1]. The most common localizations are the extremities of the limbs and the head and neck region [2]. In the facial region, the mandible is the most common site of occurrence [3] [4]. Neurofibromas of the external auditory canal are rare [5]. It is often associated with neurofibromatosis type 1 (NF1) [6] [7]. Neurofibromatosis is one of the most common

netic disorders transmitted in an autosomal dominant fashion affecting more than 1 out of 3000 individuals caused due to deletions, insertions, or mutations. In neurofibromatosis type 1 gene is located in the pericentromeric region on chromosome 17 [8]. Isolated neurofibromas are called solitary neurofibromas. It is rarely described in the literature. Most reported cases are associated with NF1.

Our aim was to use this case report to discuss the rarity and difficulty of diagnosis and treatment to the neurofibromas of the external auditory canal.

2. Case Report

A 45-year-old female patient presented with complaints of right hearing loss accompanied by tinnitus and otalgia. She had noticed the presence of a swelling in her right ear that had been progressing over the course of three years. Examination revealed a tissue mass firm, painless and covered with normal skin obstructing the right external auditory canal. Skin examination was normal, with no café-au-lait spots. An ear CT scan indicated the presence of a hypodense tissue mass filling the external auditory canal without bone destruction. Pure tone audiometry showed conductive hearing loss of 35 dB.

The mass was surgically removed, through the canal approach, under general anaesthesia. The tumour appeared white upon contact with the cartilaginous duct (**Figure 1**). The postoperative period was uneventful. Pathological examination revealed a neurofibroma characterized by a tumor proliferation consisting of bundles of fairly regular, spindle-shaped cells. Their cytoplasm is eosinophilic and their nuclei elongated with tapered ends (**Figure 2**). After three years of follow up, there has been no recurrence.



Figure 1. Well-limited white mass after exposure.

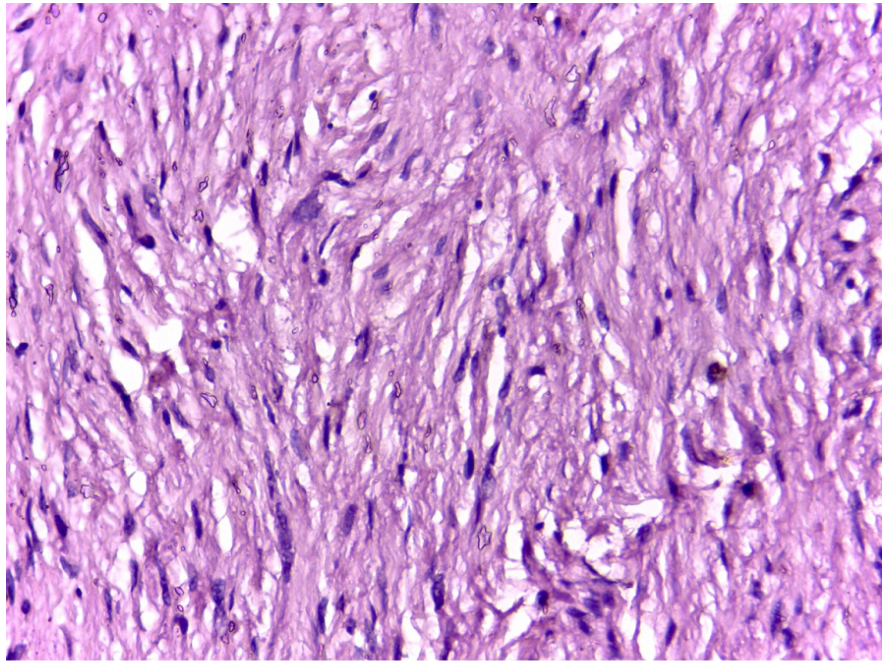


Figure 2. Hematoxyline eosin $\times 200$. Image showing a tumor proliferation consisting of bundles of fairly regular, spindle-shaped cells. Their cytoplasm is eosinophilic and their nuclei elongated with tapered ends.

3. Discussion

Neurofibromas are benign tumors of the connective tissue of peripheral nerves, arising mainly of the endoneurium [1]. The mandible is the most common site in the facial region [3] [4]. Neurofibromas of the external auditory canal are rare [5]. It may be isolated or occur in the context of neurofibromatosis type I [6] [7] [8]. Isolated neurofibromas are called solitary neurofibromas.

Ear canal neurofibromas can develop on the auricular branch of the X or branches of the VIIth and IXth pairs of cranial nerves [9]. The nerves are responsible for innervation of the auditory canal. Alyono reports a series of 4 patients with neurofibromas of the ear developed on the facial nerve. Of these, 3 presented with preoperative facial paralysis. About this situation, the author recommends imaging before the biopsy, and underlines the therapeutic dilemma of preserving the facial nerve [10].

Neurofibromas are most commonly found in males aged between 14 and 45 years [3]. However, in the ear, the female sex seems to be more affected: 4 out of 7 reported cases are female [5]-[9]. Our patient was a young woman aged 45.

Clinical signs are non-specific, but conductive hearing loss is consistently reported in all cases [5] [6] [8] [9]. The hearing threshold typically range between 30 and 40 dB and depends on the degree of ear canal obstruction [9]. A CT scan reveals a heterogeneous contrast-enhanced tissue mass that is not specific to neurofibroma. The diagnosis is obtained by histological examination after biopsy. In our case, we removed the mass straight away, as it was mobile and well limited. In addition, we live in a poor country, and it is often the patients

who pay directly for medical care.

Complete and large surgical excision without causing postoperative functional loss is the treatment of choice for neurofibromas, because recurrence is possible [5] [11] [12]. The retroauricular approach is the most commonly used because the scar behind the ear is more esthetic. We preferred the canal approach, which was easier and quicker, with fewer sutures. Dissection was facilitated by the use of a stripper.

In pediatric NF1 patients, complete or near complete excision of the neurofibromas ensures recurrence rates of <20% and 40%, respectively, whereas subtotal resection (removal of 90% or less) leads to a recurrence rate of >60% [13]. We therefore believe that isolated neurofibromas may have a better prognosis than neurofibromas occurring in the context of NF1. Cases of malignant degeneration have been described in the literature [1] [14]. The possibility of recurrence and malignant degeneration requires a long term follow up of patients.

4. Conclusion

Solitary neurofibromas in the external auditory canal are rare. Preoperative diagnosis is difficult, as symptoms are non-specific. The histological result often comes as a surprise to the surgeon.

Informed Consent

Informed consent was obtained from the patient to report this case.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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