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CASE REPORT

Obliterative External Ear Canal Schwannoma

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Objective: To report a very unusual occurrence of obstructive external ear canal schwannoma.

Study Design: Case report and review of relevant literature.

Materials and Methods: Presentation of patient data, CT imaging, surgical therapy and histological findings.

Results: A schwannoma completely obstructed the opening of the external ear canal without tympanic membrane involvement. CT scan showed a well-defined tumor mass, which appeared benign. The tumor was completely excised using a postauricular approach.

Conclusion: Schwannoma should be considered in the differential diagnosis of benign or malignant tissue masses involving the external ear canal, although, in this location, the clinical and radiological findings are somewhat nonspecific and rare

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Symptoms such as hearing impairment or tinnitus are generally attributed to non-neoplastic causes, such as otitis media or trauma. Less commonly, auricular signs and symptoms are the result of neoplastic space-occupying lesions. The external auditory canal (EAC) can be the site of development of different benign tumors including ceruminous and pleiomorphic adenoma, squamous papilloma, osteoma, chondroma, haemangioma and keratoacanthoma.

Schwannoma is the most common neurogenic tumor of the auricular region, and the commonest nerve of origin is the vestibulocochlear nerve. Schwannomas of the external ear and tympanic membrane are exceedingly rare,[1,2] and therefore are often not immediately included in the differential diagnosis of EAC lesions. Here, we report a case of a giant schwannoma completely filled the opening and proximal part of the right EAC. Cure was achieved by surgical excision using a postauricular approach. To our knowledge, only seven previous cases have been reported to date in the world medical literature.[3] We discuss the clinical and radiographic appearances, surgical results and histopatological findings of this very rare lesion.

Case Report

A 59-year old male was admitted to the ENT department with a 5-year history of a sensation of fullness in his right ear and progressive right sided hearing loss. There was no otalgia or discharge from the ear. Macroscopically there was a smooth-surfaced ovoid mass obliterating the entrance of the external ear canal (Figure 1). Computed tomography (CT) showed a rounded soft tissue mass indicating a benign lesion (Figure 2). Surgical excision using a postauricular approach without cartilage resection was performed (Figure 3). A tumor mass arising from the posterior wall of the external ear canal was identified. It measured 3.5cm in largest diameter and was completely removed. Histological evaluation confirmed a benign schwannoma (Figure 4). The patients' recovery was uneventful, and no recurrence was observed during 8 months follow up.

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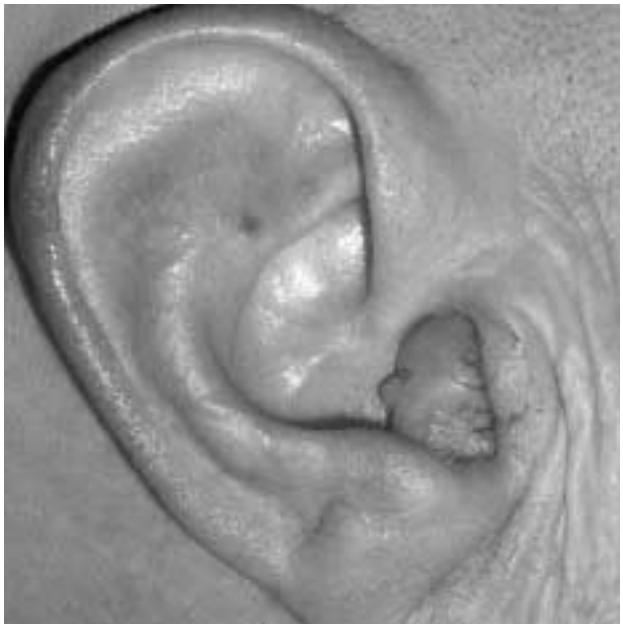

Figure 1. Tumor presenting as an obliterative ovoid mass involving the entrance of the external ear canal

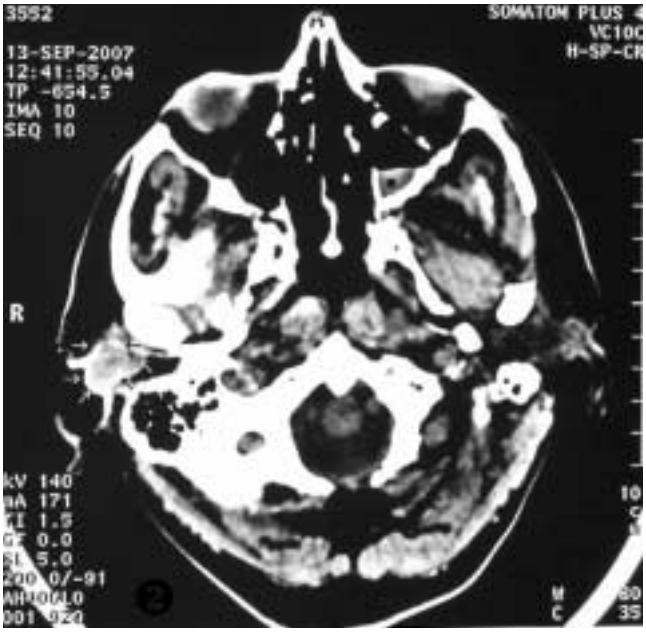

Figure 2. Axial CT scan showing a well-defined soft mass in the lateral part of the right external ear canal

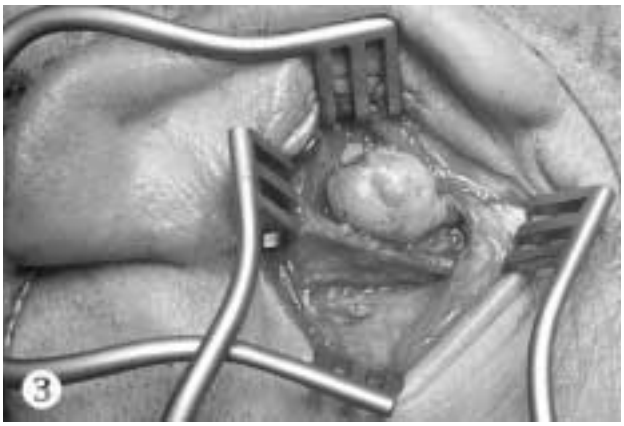

Figure 3. Postauricular minimal invasive approach to the tumor; incision of the posterior wall of the external ear canal exposes the tumor, which has an intact capsule

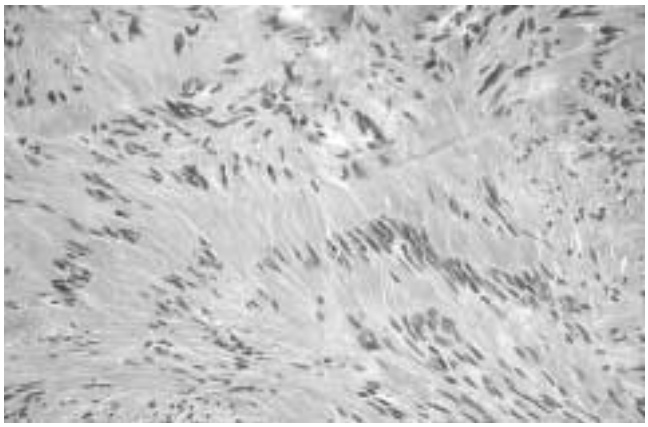

Figure 4. Histopathological section of the tumor demonstrating areas of compact spindle cells arranged in a palisade pattern (H&E staining, original magnification X40)

Discussion

The auricle and EAC are lined by keratinising squamous epithelium with accompanying cutaneous adnexal structures (such as hair follicles and sebaceous glands). Furthermore, the outer portion of the EAC contains variants of the apocrine gland known as ceruminous glands. The auricle and external auditory canal are, in turn, supported by fibrofatty soft tissue, elastic cartilage, and bone. The auriculotemporal branch of the mandibular division of cranial nerve (CN) V supplies the superior and anterior aspects of the EAC. The posterior and inferior aspects of the EAC are supplied by CN VII, IX, and X via the auricular branch of the vagus nerve (Arnold's nerve).

The epithelial elements may give rise to benign papilloma or keratoacanthoma[4]. The glandular appendages may be the source of ceruminous gland tumors and of pleiomorphic adenoma Such tumors were reported in less than 150 cases reported worldwide.[5,6] Benign mesenchymal tumors include

the very common osteomas[7] , but only 48 cases of chondroma of the external auditory canal have been documented in the literature to date.[8] Other benign tumors such as myxoma,[9] hemangioma,[10] myoma,[11] lipoma[12] and fibroma[13] have been published sporadically.

Benign nerve sheath neoplasms originate from Schwann cells and include schwannomas (neurilemmomas) and neurofibromas. Schwannoma of the external ear canal was first reported in an English language article in 1993 by Wu et al.[14] These are encapsulated lesions that grow away from the nerve trunk and can arise from any somatic or sympathetic nerve.

Although external ear schwannomas are not common, obliterative tumors of this size and in such a location could be considered as rare lesions. Like schwannomas in other sites, these tumors usually present as asymptomatic masses and often grow to a considerable size before they become obvious or clinically detectable. The symptoms and signs of previously reported cases were progressive otalgia and bloody otorrhea[15,16] related to pressure effects of the tumor and/or infection. The patient presented here felt only a sensation of blockage in his right ear and consequently progressive hypoacusis, but without pain or canal inflammation. These clinical features raise the possibility of degenerative changes or malignancy. Perineural infiltration and paralysis of the facial nerve, rapid growth, adjacent tissue infiltration and lymphatic metastases could also indicate malignancy. Nevertheless, there was no specific sign or symptom that identified a nerve origin pre-operatively: the diagnosis depended largely on the anatomopathologic description and only secondarily on clinical findings.

Grossly, the mass was well defined, but this did not help us to make a diagnosis preoperatively. This difficulty could have been eliminated by presurgical biopsy. However, biopsy may induce bleeding from a vascular tumor or sometimes there is a risk of creating a fibrous adherence between the capsule of the tumor and the wall of the EAC leading to difficulties with the later removal of the tumor. However, cavernous hemangiomas generally do not exhibit bone invasion but rather arise most frequently in the posterior portion of the EAC, sometimes extending onto the tympanic membrane.[17] An osteoma manifests as a discrete, pedunculated bony mass that arises from a different site - the tympanosquamous suture line adjacent to the bone-cartilage junction. Keratoacanthomas arise from the hair follicles and represent epithelial tumors, which are characterized by a keratin-filled crater, rapid growth in the proliferation stage, and the potential for spontaneous regression. The lesion presents as an elevated mass that appears flesh colored or slightly reddish. Keratoacanthoma should be considered in the differential diagnosis of squamous cell carcinoma. Pleiomorphic adenoma is a neoplasia of the ceruminous glands of the EAC, which are located in the lower two-thirds of the dermis in the cartilaginous segment of the EAC.[18] Grossly, it is a skin-covered, circumscribed, polypoid or rounded, usually not ulcerated, gray-white-pink mass.

As the CT scan indicated that the mass was most likely benign, a presurgical or fine needle aspiration (FNA) biopsy was not performed. Polyposition CT of the temporal bone including axial and coronal projections is the most informative method of visualization of the bony part of the EAC. Axial projection covers the anterior and posterior walls of the canal whilst coronal projection covers the upper and lower walls. Hence, CT scanning is able to characterize EAC changes (soft tissue, bone, size, position). Radiologic imaging by CT shows schwannomas to be well-circumscribed, homogenous masses that enhance with contrast. CT is also mandatory to rule out the extension of a schwannoma from other temporal bone sites (middle ear, mastoid or internal auditory canal) to present as an external ear canal mass.[19]

Definite diagnosis should be based on the histological and immunohistochemical findings. Typically, histological analysis demonstrates that a schwannoma is composed of S-100 protein positive Schwann cells arranged in 2 growth patterns, namely Antoni A and B. The former is a highly cellular pattern and composed of elongated Schwann cells, which exhibit nuclear

palisading. The Antoni B areas are also composed of elongated Schwann cells, but these are arranged in a less dense myxoid pattern and are more disorganized than Antoni A areas. In contrast to schwannomas, neurofibromas frequently occur in multiples and are not encapsulated. This along with the fact that they are less likely than schwannomas to undergo cystic degeneration may also help in radiologically differentiating the two tumors.

Surgical removal of an obliterative schwannoma arising in the external ear canal is sometimes technically difficult because of limited operative exposure. Although schwannomas are usually benign, their resection can be associated with significant postoperative morbidity such as cicatricial stenosis or infection. Appropriate therapy requires complete excision with minimal injury to uninvolved areas. However, different surgical approaches are possible. Generally, two options have been proposed most frequently: transmeatal[20] and postauricular approaches.[21] When surgery is planned, the best approach will inevitably depend on the tumor location, size and its relations to surrounding structures. Most cases of schwannoma reported to date have only partially occupied the external ear canal cavity and the largest tumor did not exceed 3 cm in diameter. Consequently, transmeatal or endaural approaches have been employed most frequently. In this case, the extensive size of the lesion made endaural enucleation impossible. Using a postauricular approach, we found that a small epithelial incision in the posterior wall of the external ear canal allowed careful surgical dissection, which gradually exposed the tumor. This approach led to only modest intraoperative bleeding which was easily controlled. With this technique, which provides a wider operative field, the tumor can often be enucleated from the nerve trunk. However, it is of more importance that the fibrous capsule is left intact to facilitate resection. By dissecting along the capsule, scarring and granulation tissue formation can be avoided. As the chosen approach allowed excellent external ear canal exposure, there was no interference with cartilaginous or bony ear structures and post-operative recovery time was reduced. Such total tumor resection reduces the chances of recurrence: in this case, there is no sign of recurrence after 8 months follow up. Incomplete excision may be of concern because some tumors can undergo malignant change and cause local destruction or may result in slow local recurrence over several months to years.

Schwannoma should be considered in the differential diagnosis of benign or malignant tissue masses involving the external ear canal although, in this location, the clinical and radiological findings are somewhat nonspecific and rare. CT scanning is mandatory for identifying the benign nature and true extension of this lesion. The retroauricular approach allows excellent exposure for surgical resection with good healing and minimal cosmetic disability.

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