Neurofibroma of the external ear - a case report

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Abstract Neurofibromas are relatively common tumours of the nervous system, but only a few cases involving the external ear have been reported. We are reporting here a case of a 20-year-old female with neurofibroma of the external ear. The primary complaint was cosmetic deformity. There was partial occlusion of the external auditory canal. The swelling was excised by postauricular approach. Surgery resulted in an excellent functional and cosmetic outcome.

Keywords Neurofibroma · External ear.

Introduction

Neurofibromas are circumscribed but nonencapsulated neoplasms of the nervous system. They can arise in all peripheral nerve elements, including Schwann's cells, neurons, fibroblasts and perineural cells. Neurofibromas are usually benign. Malignant transformation has been reported to occur in 2 to 16% of cases [1]. Neurofibromas of the head and neck are not uncommon, but they rarely affect the external ear and only a few such published reports are available.

Case report

A 20-year-old female patient attended the ENT department with a soft swelling at the concha extending into cartilaginous part of external auditory canal and at the postauricular area of left ear. The swelling occluded the external auditory canal partially. The bony part of the external canal and the tympanic membrane were normal. The size of the postauricular swelling was about 4 cm × 2.5 cm. There was no pain; but there was slight tenderness over the swelling. There was no history of bleeding. The patient first noticed the swelling six years back, when it was small. She was treated by a homeopathic doctor for 2 years. A local doctor performed incision and drainage twice. An incisional biopsy was done in the ENT department. The histopathology report was neurofibroma.

Most probably the tumour developed on the great auricular nerve. Pure tone audiometry showed no hearing loss. Her vision was normal. She had no family history of neurofibromatosis. The swelling was excised by postauricular approach under local anaesthesia. The mass was adherent to the bony cortex and to the skin of the meatus and ear canal. After removing a piece of conchal cartilage the entire mass was removed along with the excess skin which was adherent to it. Histopathological examination of the mass showed it to be neurofibroma. The lesion composed of spindle shaped cells having fusiform twisted nuclei arranged in fascicles. No significant cellular atypia was seen. The patient made an uneventful recovery, and a good cosmetic result was achieved.

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Fig. 1 Showing the swelling at the concha and postauricular area.

Discussion

Neurofibromatosis (NF) is one of the most common inherited disorders and can affect anyone, regardless of family history, race, gender, or ethnic background. There are two types of NF, type 1 (NF 1) and type 2 (NF2). NF 1 affects approximately one out of every 4,000 individuals. NF 2 affects one out of every 40,000 individuals [2]. Neurofibromas are tumours derived from Schwann cells, fibroblasts and the supporting cells known as perineural cells. They can grow anywhere in the body where there are nerve cells. This includes nerves just under the surface of the skin, as well as nerves deeper within the body, spinal cord, and / or brain. In NF 1 neurofibromas most commonly grow on the skin or on the nerve to the eye. NF 1 is caused by a change in a gene carried on chromosome 17. The neurofibromas may turn into malignant neurofibrosarcomas. In NF 2 neurofibromas most commonly grow within the spinal cord or brain. Bilateral schwannomas of the acoustic nerve and multiple meningiomas are characteristic. NF 2 is caused by a change in a gene carried on chromosome 22. Solitary lesions are not usually associated with systemic manifestations unlike multiple lesions which are commonly seen in patients with neurofibromatosis or von Recklinghausen's disease. There are many histological subtypes of neurofibroma including localized, plexiform and diffuse [3]. The change in the genetic material that causes NF 1 and NF 2 can be inherited from a parent, referred to as autosomal dominant inheritance, or it may occur as a result of a spontaneous mutation [4]. In our case there was no family history. A single or few neurofibromas alone do not indicate NF; some people who do not have NF

Fig. 2 Microphotograph showing spindle shaped cells having fusiform twisted nuclei. (H & E $\times 450$).

may have a neurofibroma [2]. The present case did not have any other features of NF 1. Most tumours caused by NF need no treatment. But tumours that are painful, disfiguring, growing rapidly, impairing function or compressing other body parts may need treatment. These tumours frequently recur and palliative treatment is usually required. It has been suggested that subtotal resection of neurofibromas is preferable to a complete excision as the latter procedure may result in a loss of normal tissue and still not prevent a recurrence [5]. Trevisani et al wrote that 'complete surgical excision of these lesions is virtually impossible; if not contraindicated' [6]. Crikelair and Cosman emphasized that amelioration rather than complete eradication should be the therapeutic goal [7]. Large lesions may require reconstruction and elevation of the pinna with dermal strips or marlex polypropylene mesh [6]. In our case we removed the entire lesion along with some skin and conchal cartilage. The patient made an uneventful recovery, and a good cosmetic result was achieved. During the one year follow-up period there has been no recurrence.

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