Facial Nerve Neuroma : A Case Report

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Introduction

Facial nerve neuromas are uncommon tumors involving facial nerve. There are no typical patterns of presentation and can easily go untreated or misdiagnosed. Facial nerve palsy is the commonest mode of presentation. We present a case of facial nerve neuroma presenting with foul smelling otorrhoea followed by facial nerve palsy somewhat mimicking cholesteatomatous ear disease. Diagnosis was only possible peroperatively and later confirmed by histopathology.

Case Report

A 65 year old individual presented in the outpatient department with history of right sided, occasionally foul smelling, scanty ear discharge without any pain, deafness, tinnitus or vertigo of two years duration. He had not taken any active treatment for the same. On examination the right tympanic membrane was found intact but retracted. There were some crusts and a fleshy mass that seemed to be arising from the posterosuperior bony canal wall. Fistula test was negative and there was no mastoid tenderness or facial nerve palsy. At the outset, cholesteatomatous ear disease was suspected. He was prescribed antibiotic ear drops and oral antibiotics and asked to return with some relevant investigations. He returned two weeks later with an infrageniculate facial nerve palsy of the right side, which had progressed to House Brackman Grade -II within a week. The fleshy mass in the ear canal had completely disappeared. A small defect of about 2 mm was noticed in the posterosuperior bony canal wall close to the annulus. Microscopic examination did not reveal any ear drum perforation or cholesteatomatous flakes. In view of intact ear drum, the long standing foul smelling ear discharge, erosion of posterosuperior bony wall and onset of facial palsy a clinical diagnosis of congenital cholesteatoma behind an intact tympanic membrane was made.

On investigation, he was non-diabetic and all haematological and biochemical parameters were normal. Audiogram revealed early presbycusis affecting both the ears. X-ray mastoids showed partly sclerotic mastoid on the right with a small area of lucency containing wooly opacities.

He could not afford a CT scan. He was put on parenteral antibiotics along with oral prednisolone. He was taken up for exploration of the mastoid and middle ear. Under local anaesthesia, mastoid was approached through a standard post aural incision. While elevating the posterior meatal skin, a soft tumor like mass was encountered unexpectedly that was arising from within the bone in the region of the tympanomastoid suture. It was pale, firm and did not bleed on touch. The tumor was followed medially. A modified radical mastoidectomy was performed because part of the tumor extended into the mastoid antrum eroding the dome of the lateral semicircular canal. There was no fistula. The tumor was arising from the mastoid segment of the facial nerve and was meticulously dissected off from the nerve trunk. Thereafter the entire facial nerve from the stylomastoid foramen to the cochleariform process was conventionally exposed and decompressed. The nerve trunk was found intact. There was no trace of cholesteatoma anywhere in the middle ear or mastoid. The middle ear was normal. All the ossicles were intact. Post operatively the facial palsy became complete. Steroids were continued for four weeks and tapered off. The mastoid cavity healed well. Facial palsy recovered to House Brackman Grade-III in 16 weeks. After one year the palsy recovered to the preoperative level with minimal synkinesis. There was no worsening of hearing after surgery.

Fig. 1 : Microphotograph from the mass showing features of neuroma having spindle cells with wavy elongated nuclei with pointed ends (magnification 100 x)

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Microphotograph from the mass (Fig 1) showed features of neuroma.

Discussion:

Facial nerve neuromas can occur anywhere along the course of the facial nerve but reports suggest that the most common site is the geniculate ganglion [1].

Primary neurogenic tumors are rare. The majority are neuromas also known as schwannomas or neurilemmomas [2]. The clinical presentation and symptoms of neuroma of the facial nerve differ not only due to the location but also due to variability of involvement of adjacent anatomic structures. Horn et al, stated that presenting symptoms can be grouped into three basic categories: (a) facial nerve dysfunction; (b) hearing loss; and (c) facial pain or neuralgia.

The most common mode of presentation is the occurrence of progressive facial nerve dysfunction over a period of months. Jackson et al [3] pointed out that idiopathic facial nerve palsy progressing to complete dysfunction cannot simply be idiopathic. The paresis may be preceded by fasciculation or hemifacial spasm. Chorda tympani tumours are rare and may not cause facial nerve palsy [4]. Other associated findings help in localizing the site of tumor. Hearing loss is a common complaint. It may be conductive, cochlear or retrocochlear. Intracanalicular tumors may show retrocochlear hearing loss. Origin from the mastoid portion of the nerve leads to palsy, sometimes followed by the formation of an aural polyp located in the posterior wall of the EAC. Ear discharge may be present [5]. A relatively unusual presentation is otalgia. Diagnostic work up should include audiological tests including Brainstem Evoked Response Audiometry (BERA) as well as vestibular evaluation. Electrophysiological studies are of limited value. Tumor extent can be best studied by MRI with gadolinium. Both CT and MRI are complimentary to each other [6]. The treatment of facial nerve neuroma is surgical excision. The approach to treatment depends on tumor extent and degree of hearing.

Tumors of the vertical portions of the nerve are best managed by the transmastoid approach. The transmastoid extralabyrinthine approach is used for lesions extending to the geniculate ganglion. Total facial nerve exposure is offered by combined middle fossa and transmastoid approach in cases with good hearing. The trans labrynthine approach is used in cases with unserviceable hearing. The approach is appropriately modified as per tumor extension. The tumor has to be removed along with a segment of facial nerve followed by cable grafting. The graft is generally obtained from the post auricular nerve. There will be almost total paralysis for at least six months. Richard Liu [7] quoting

O'Donoghue et al has said that in 25% of his cases they were able to preserve at least 50% of the nerve intact. Regardless of the timing of surgery or the type of facial nerve repair, the eventual recovery of function will be no better than House-Brackman Grade-III. Our patient recovered to House Brackman Grade-II.

On comparing the outcomes of those who underwent definitive excision with those who were managed conservatively, the former group did well [7]. In our case the patient presented with ear discharge and a defect in posterior bony wall along with granulations and with a good hearing. Preoperative diagnosis of neuroma was not possible by the conventional investigations. Essem Saleh et al [8] obtained preoperative diagnosis in only 13 of 22 cases in a series.

The facial palsy in our case possibly developed due to pressure on the nerve aggravated by acute infection. We are of the opinion that pressure on the facial nerve due to any reason causes oedema of the nerve compromising blood supply and early institution of steroid is considered beneficial for a favourable outcome. Diagnosis was possible only at surgery. Tumor was excised from the mastoid portion of the nerve. We were able to preserve major part of the nerve due to which patient has shown good recovery, both functionally and cosmetically. Our case demonstrates an uncommon presentation of a relatively rare tumor in which the tumor has been excised with preservation of the facial nerve trunk with good functional recovery, which is also uncommon.

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