Journal of Clinical Gerontology & Geriatrics 3 (2012) 42–44

Contents lists available at SciVerse ScienceDirect

Journal of Clinical Gerontology & Geriatrics

journal homepage: www.e-jcgg.com

Case report

Congenital aural stenosis with first branchial cleft fistula, presenting with recurrent facial nerve palsy, in an elderly patient

Shraddha Jain, MS [a,*], Sunil Kumar, MD, PGDGM [b], P.T. Deshmukh, MS [a], Dennis Yohannan, MD [c], Kavita Sudarshan, MBBS [a]

[a] Department of Otolaryngology and HNS, Jawaharlal Nehru Medical College, DMIMSU, Sawngi Meghe, Wardha, Maharastra, India
[b] Department of Medicine, Jawaharlal Nehru Medical College, DMIMSU, Sawngi Meghe, Wardha, Maharastra, India
[c] Department of Radiodiagnosis, Jawaharlal Nehru Medical College, DMIMSU, Sawngi Meghe, Wardha, Maharastra, India

ARTICLE INFO

Article history:
Received 12 August 2011
Received in revised form
10 November 2011
Accepted 14 November 2011

Keywords:
Branchial cleft
Cholesteatoma
Congenital ear deformity
Ear canal
Elderly
Fistula

ABSTRACT

Both duplication anomalies and external auditory canal stenosis can result in cholesteatoma due to retained epithelium. Despite their common origins, the coexistence of these anatomical abnormalities is quite unusual. This cholesteatoma may go unnoticed till adulthood and present with complications. Moreover, first branchial cleft malformations are often unrecognized or misdiagnosed for other inflammatory lesions in the periauricular region. We report a case of middle-ear cholesteatoma in a 68 year old male patient with grade II microtia with canal stenosis and first branchial cleft fistula in supra-auricular region presenting with recurrent infranuclear facial nerve palsy. The patient was managed surgically by excision of fistulous tract, canalplasty, auricular reconstruction and middle ear and mastoid exploration for cholesteatoma. This may probably be the oldest person to be managed surgically for cholesteatoma due to congenital ear anomalies and first such reported case.

1. Introduction

Both duplication anomalies of the external auditory canal (which include first branchial cleft cysts, sinuses and fistulae) and its congenital stenosis, can result in cholesteatoma due to retained epithelium.[1] Cholesteatoma behind the stenotic canal, or due to branchial cleft cyst, may go unnoticed until adulthood and present with complications due to local destruction of adjacent bony structures.

Microtia, with congenital aural stenosis, is known to be associated with postauricular fistulae and sinuses, because of the presence of primary cholesteatoma called "cholesteatoma auris congenita of atretic ear," due to retained epithelium behind the atretic canal.[2] These lesions invariably have part of the cartilaginous canal patent, but have an atrophic bony canal. Other causes of fistulae in the periauricular region could be the co-existence of duplication anomalies (first branchial cleft fistula) with canal stenosis. The first branchial cleft anomalies are rare and often unrecognized or misdiagnosed as tumors or other inflammatory lesions in the periauricular region. Surgical treatment is inadequate, leading to recurrence or secondary infection. These anomalies may have a close association with the facial nerve. Therefore, this may be a cause of recurrent facial nerve palsy and may lead to the risk of injury to the facial nerve while undergoing surgery.

We report a rare case of middle ear cholesteatoma in a 68-year-old male with grade 2 microtia and canal stenosis, with first branchial cleft fistula in the supra-auricular region, presenting with recurrent discharge from the fistula and recurrent painful swelling in the supra-auricular region of 45 years duration, with recurrent infranuclear facial palsy.

2. Case report

A 68-year-old male patient with grade II microtia and congenital aural stenosis of the right ear, presented to the otolaryngology department of our hospital, with a history of recurrent discharge from the preauricular region, associated with supra-auricular swelling and otalgia, spanning over a period of 50 years. He had grade V right infranuclear facial palsy at the time of presentation (Fig. 1) and had a history of recurrent facial palsy in the past, which improved with treatment. Keratin debris was noted within the stenotic canal, which was 2 mm in diameter and the tympanic membrane could not be visualized. There was a supra-auricular

* Corresponding author. Department of Otolaryngology and HNS, Jawaharlal Nehru Medical College, DMIMSU, Sawangi Meghe, Wardha, Maharastra, India.
E-mail address: sjain_med@yahoo.co.in (S. Jain).

2210-8335
doi:10.1016/j.jcgg.2011.11.010

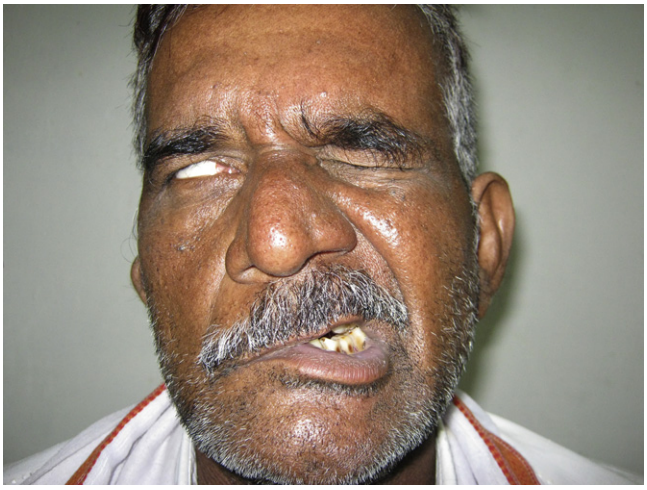

Fig. 1. Photo showing right infranuclear facial palsy.

fistula (Fig. 2) which was discharging pus in front of a supra-auricular swelling. Fistulography, followed by a CT scan of the temporal bone, revealed a fistulous tract extending from the middle ear cavity to the superior aspect of the ear lobe and complete atresia of the bony part of the external auditory canal and tympanic membrane. The middle ear cavity was found to be partially replaced by fat density lesions with absent ossicles. Scutum was eroded and a dehiscence of bony wall of the tympanic segment of the facial nerve was observed with its inferior displacement (Figs. 3 and 4). A diagnosis of congenital canal stenosis with cholesteatoma and first branchial cleft fistula were made. After adequate control of the discharge by an injection of ceftriaxone, the patient was managed by canalplasty, auricular reconstruction and middle ear and mastoid exploration for cholesteatoma, along with excision of the tract of the branchial cleft fistula. Intra-operative findings confirmed the radiological findings of cholesteatoma in the middle ear, with absence of bony canal, tympanic membrane and ossicles. Tympanic segment of the facial nerve was dehiscent and branching of the laterally placed mastoid segment was noted with loop formation towards the fistula site (Fig. 5). The supra-auricular fistula opened in the middle-ear and much supra-auricular tissue was seen, due to repeated infections, so the cyst could not be delineated. The excess conchal cartilage was excised for

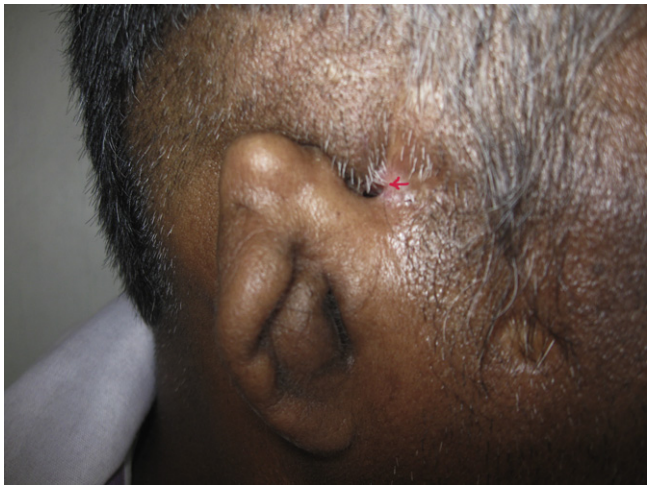

Fig. 2. Grade II microtia with canal stenosis with first branchial cleft fistula in the supra-auricular region (red arrow).

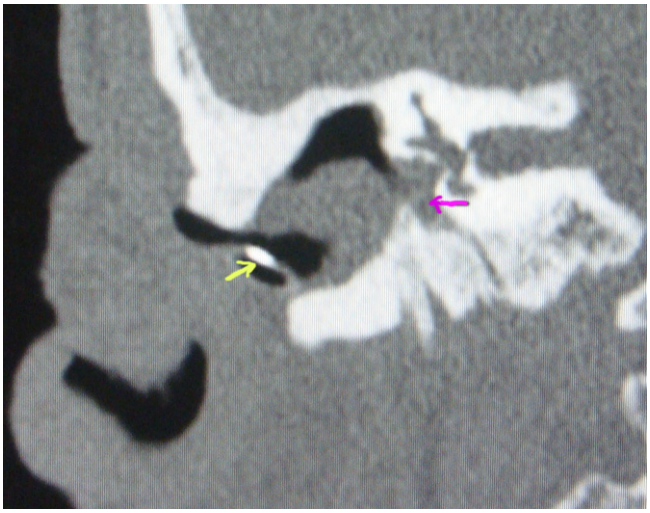

Fig. 3. High resolution computed tomography (HRCT) of the temporal bone (coronal section at the level of the oval window). The image demonstrates a cannulated fistulous tract (yellow arrow) communicating with the middle ear cavity occupied by a soft tissue mass causing smooth lining of the mesotympanum and hypotympanum with erosion of scutum. The ossicular chain is not visualized. The bony canal of the tympanic (horizontal) segment of the facial nerve shows erosion in its inferior aspect (pink arrow).

canaloplasty and it was used for auricular reconstruction. The patient was doing well 6 months after surgery, with no signs of recurrence of the cholesteatoma or branchial cleft cyst, but he still had persistent grade IV facial palsy, which was being managed by physiotherapy and eye care. EMG performed 1 month after surgery demonstrated the presence of voluntary motor unit action potentials, indicating a favorable outcome for the patient.

3. Discussion

The first branchial cleft gives rise to the external auditory canal and the epithelium of the tympanic membrane. Developmental errors of the first branchial cleft can result in duplication anomalies of the external auditory canal, as well as external auditory canal stenosis and atresia. A classification system has been presented by Blevins that addresses the full spectrum of first branchial cleft anomalies, which includes both absence and duplication of the external auditory canal according to the stage of development at

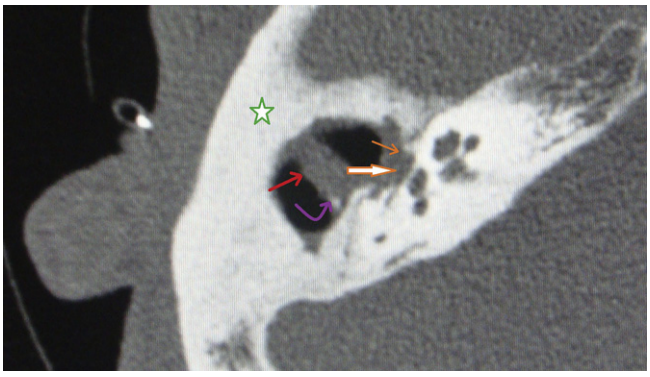

Fig. 4. High resolution computed tomography (HRCT) of the temporal bone (axial section at the level of the superior portion of the cochlea and vestibule). The temporal bone shows sclerotic changes (green star) with fat density lesion (red arrow) in the middle ear cavity. A remnant of the ossicular chain is visualized (purple curved arrow). In this image, the lateral wall of the horizontal segment of the facial canal is eroded (orange arrows).

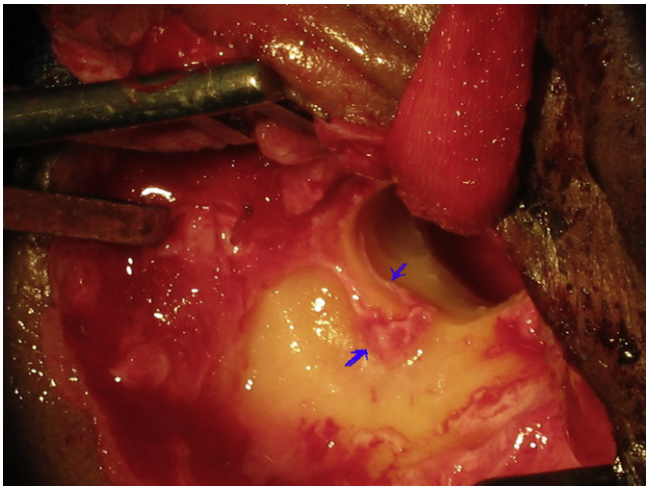

Fig. 5. An operative photograph showing a laterally placed mastoid segment of the facial nerve with loop formation (blue arrows).

which arrest occurs. The failure of obliteration of the ventral aspect of the first cleft during weeks 5 and 6, results in a duplication anomaly of the external auditory canal, which can manifest as a cyst, sinus or fistula. Sinuses and fistulae arise from incomplete closure of the first branchial groove and if the failure occurs during the time of formation of the closing membrane, a fistula extending into the middle ear (a first pouch derivative) may result, as occurred in our patient.[3] Failure of the process of recanalization in the 6th and 7th months of gestation is manifested as aural atresia or stenosis. Occasionally, squamous epithelium may be trapped medially within a stenotic canal, resulting in a canal cholesteatoma. This does not occur with grade III microtia, as it is associated with absent canal or canal atresia, due to the arrest of development at an earlier stage at the 4th week of gestation. The cholesteatoma occurs more commonly if the diameter of the external auditory canal (EAC) is < 2 mm.[4] In our patient, there was incomplete recanalization, with a stenotic external auditory canal which ended blindly and an absent bony canal. Furthermore, the ventral aspect of the external auditory canal failed to involute, persisting as a first branchial cleft cyst with fistulous opening in the middle ear cavity, which got repeatedly infected. The retained epithelium due to both these conditions led to the occurrence of middle ear cholesteatoma, with recurrent facial nerve palsy due to both dehiscent tympanic segment and abnormal loop of facial nerve in association with the fistula tract.

Despite their common origins, the coexistence of these anatomical abnormalities is quite unusual. Only few cases have been reported.[1,5,6] In our patient, presentation at 68 years of age makes it a unique occurrence.

The distinct clinical features of recurrent painful swellings around the ear and sometimes associated with pus discharge from fistulous or sinus openings, in association with malformations of the pinna and the external auditory canal, should make us suspect first branchial cleft anomalies. Treatment, in the form of excision of the cyst and tract, should be followed by subjecting the tissue for histopathological diagnosis.

The surgeon should be wary of close association of these anomalies with the facial nerve and try to identify and preserve the nerve at the time of surgery (loop formation of the facial nerve and opening into the normal ear canal or the loop of the facial nerve around the tract).[7]

In conclusion, while evaluating patients with congenital ears, otologists should always rule out the presence of cholesteatoma by high resolution computed tomography (HRCT) of the temporal bone in every case. The surgical plan in a patient with congenital anomalies of the external and middle ear should be tailored to individual defects, along with the management of complications in each patient. In elderly patients, the main aim of treatment is clearance of the disease and prevention of repeated infection more than the cosmetic aspect.

References

1. Barkdull G, Carvalho D. Goldenhar syndrome with external auditory canal stenosis complicated by canal cholesteatoma and first branchial cleft cyst. *Int J Pediatr Otorhinolaryngol Extra* 2007;**2**:128–32.
2. Nishimura Y, Minatogawa T, Kumoi T. Intractable retroauricular abscess associated with microtia and aural atresia – some views in relation to the congenital cholesteatoma and microtia. *Ann Plast Surg* 1989;**23**:74–80.
3. Tham YS, Low WK. First branchial cleft anomalies have relevance in otology and more. *Ann Acad Med Singap* 2005;**34**:335–8.
4. Cole RR, Jahrsdoerfer RA. The risk of cholesteatoma in congenital aural stenosis. *Laryngoscope* 1990;**100**:576–8.
5. Yalcin S, Karlidag T, Kaygusuz I, Demirbag E. First branchial cleft sinus presenting with cholesteatoma and external auditory canal atresia. *Int J Pediatr Otorhinolaryngol* 2003;**67**:811–4.
6. Blevins NH, Byahatti SV, Karmody CS. External auditory canal duplication anomalies associated with congenital aural atresia. *J Laryngol Otol* 2003;**117**:32–8.
7. Mokhtari N, Sharifi N. Preservation of facial nerve in surgery of first branchial cleft anomaly: five cases of duplicated external ear canal. *IJMS* 2006;**31**:50–2.