Congenital Cholesteatoma of Temporal Bone with Bezold's Abscess: Case Report

Nara Janardhan · Janardhan Nara ·
Indeevar Peram · Suresh Palukuri ·
Arunkumar Chinta · Kuldeep Satna

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Abstract Congenital cholesteatoma is a rare entity. It may originate at various sites in the temporal bone, for example, in the petrous apex, the cerebellopontine angle, the middle ear cavity, the mastoid process or in the external auditory canal. The least common site being the mastoid process. Most common presentation is a retrotympanic pearly white mass with no previous history of ear discharge, perforation or any ear surgery. It can lead to various complications, both intracranial and extracranial, some of which may be life threatening. Bezold's abscess is an extracranial complication which is usually seen in children following acute otitis media with mastoiditis. Here we present a rare case of a 60 year old patient with congenital cholesteatoma complicating to Bezold's abscess. After necessary investigations patient underwent surgery for complete removal of cholesteatoma and the abscess drainage.

Keywords Congenital cholesteatoma · Bezold's abscess · Cortical mastoidectomy

Introduction

Congenital cholesteatoma or keratoma or epidermoid cyst is stratified squamous epithelial lined cyst filled with keratin debris. Though considered rare, the incidence of congenital cholesteatoma is on rise and now it accounts for 2 to 5% of all cholesteatomas [1]. Congenital cholesteatoma

arise from epithelial rests from faulty embryogenesis, and can also form as a result of inclusion, migration, invasion of squamous epithelium or metaplasia of normal epithelium [2]. These epithelium grow slowly and become clinically apparent in the early childhood. CC can present with variety of complaints like conductive hearing loss, ear pain, tinnitus, vertigo, facial nerve palsy, post auricular abscess and may rarely present as intracranial abscesses, but most commonly it is an incidental finding. On examination congenital cholesteatoma appears as a white round mass medial to an intact tympanic membrane, usually at the anterosuperior quadrant [3]. This epidermoid tissue is considered as congenital cholesteatoma only if it fulfill the criteria of Derlaki and Clemis [4] (i) mass medial to an intact tympanic membrane; (ii) a normal pars tensa and flaccida, (iii) no history of tympanic membrane perforation, otorrhoea, trauma or previous otological procedures, and (iv) prior bouts of otitis media or middle ear effusion are not grounds for excluding possibility of congenital cholesteatoma as it is very rare for a child to have no episodes of otitis media in first 5 years [5, 6]. The presentation of congenital cholesteatoma as Bezold's abscess is rare. As these abscesses may spread as far down as the larynx and even the mediastinum if untreated, the prognosis may be sometimes grave. Recognition of involvement of the mastoid process in the presence of a neck abscess dictates the need for concurrent surgical exploration of the neck and the temporal bone.

Case Report

A 60 years old male presented with pain behind the right ear, swelling in the right post auricular region extending down to the neck, fever, torticollis, and tinnitus since

N. Janardhan (✉) · J. Nara · I. Peram · S. Palukuri ·
A. Chinta · K. Satna
Narayana Medical College, Chinthareddypalem,
Nellore 524002, AP, India
e-mail: nara.janardhan@yahoo.com

10 days. There was history of dull intermittent pain in the right mastoid region since 5 years. There is no history of ear discharge.

On inspection an ill defined swelling measuring size of 10×5 cm, in the right post auricular region extending to the neck behind the sternocleidomastoid. Right post-auricular groove was obliterated. Skin over swelling is hyperemic, smooth and shiny. Tympanic membrane had no perforation and was dull, opaque, with loss of cone of light and decreased mobility. Tuning fork test suggested conductive type of hearing loss.

Ultrasound of neck revealed abscess in the right posterior triangle in intramuscular plane, with proximal 1/3rd of right sternocleidomastoid muscle. HRCT of right temporal bone (Fig. 1) showed soft tissue attenuation of right mastoid antrum with absence of air cells. There was absence of medial wall of mastoid antrum in region of Trautmann's triangle and sinus plate. Also a bony deficiency was seen in the posterior meatal wall. The patient was finally diagnosed as having congenital cholesteatoma complicating to mastoiditis and Bezold's abscess.

Surgery in the form of cortical mastoidectomy by post aural approach was done under general anesthesia. Cholesteatoma sac was found limited to mastoid antrum. Dura of sigmoid sinus, tegmen antri, and posterior cranial fossa was seen without any bony covering upto the lower jugular area and cholesteatoma sac was directly lying over the duramater. The cholesteatoma was delineated from its attachment and removed (Figs. 2, 3). Aditus and attic was free of squamous epithelium. A small bony deficiency found on the posterior canal wall. Anterior tympanotomy

done and lot of thick and viscid fluid was seen in middle ear which was sucked out. Middle ear mucosa was found healthy with all ossicles intact and mobile. Mastoid cavity obliteration was done using abdominal fat. Incision was extended on to the posterior triangle of neck and small quantity of pus was found deep to sternocleidomastoid muscle. Post operative period was uneventful. Patient is doing well after a follow up of 8 months.

Discussion

Congenital cholesteatoma is a stratified epithelial sac filled with keratin debris. As the histology is similar in congenital and acquired cholesteatoma, the distinction is made on clinical grounds. Here in this case we consider it as congenital by the presence of an intact tympanic membrane, no history of ear discharge or otological surgery and the sac limited to the mastoid antrum, so satisfying the criteria proposed by Derlaki and Clemis [4].

Cholesteatoma grows slowly with accumulation of desquamated keratin arising from the squamous epithelium and pathologically affecting the middle ear or the mastoid process [7]. Bone destruction caused by CC is thought to be caused by two predominant mechanisms: pressure induced bone resorption and enzymatic dissolution of bone by cytokine mediated inflammation [8]. The age of the patients ranged from 2 to 51 years (mean, 13.3 years) [9]. High resolution CT scan and MRI are most commonly used modalities to know the extend of disease and bony destruction. Early diagnosis and treatment can lead to better outcomes and decreased likelihood of recurrence.

This case shows that congenital cholesteatoma may present in an older age without a typical white mass behind the tympanic membrane appearance with the cholesteatoma

Fig. 1 Absence of medial wall of mastoid antrum in the region of Trautman's triangle and sinus plate bony deficiency in the posterior meatal wall

Fig. 2 Intra operative photo clip showing delineated cholesteatomatous sac from dura

Fig. 3 The pointer showing wide area of exposed dura

limited to mastoid antrum, which is considered the least common site of CC [10]. Here the patient was unaware of possible lethal nature of the disease because of the mild symptoms like dull pain in the temporal region. Later he developed mastoiditis and Bezold's abscess, which is very rare with cholesteatoma and this kind of presentation of congenital cholesteatoma is not seen in the literature.

This case illustrates that a thorough examination of a case presenting with swelling around the ear with or

without discharge is mandatory to exclude cholesteatoma so as to prevent life threatening complications.

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