Case Report

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Congenital cholesteatoma tract presenting as a postaural swelling

Ganesh SRS, Farhat S, McCormick MS

Department of Otolaryngology, Royal Liverpool Children's Hospital NHS Trust, Liverpool, UK.

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ABSTRACT

The authors report a case of a three-year old boy, who presented with discharge from the site of a previously excised postaural lesion reported to have been an epidermoid cyst. It was found at operation that he had a fistulous connection between the site of the excised lesion and a congenital cholesteatoma sac in the mastoid bone. This case demonstrates the rare occurrence of congenital cholesteatoma eroding through the mastoid bone to the subcutaneous tissues and mimicking an epidermoid cyst. Such a presentation in a child has not previously been reported in the literature. The clinical, radiological, surgical and histological features of this case are discussed.

INTRODUCTION

Epidermoid cysts are ectoderm-lined inclusion cysts with only a squamous epithelium. They arise either from trapped pouches of ectoderm near normal folds, or from the failure of surface ectoderm to separate from the neural tube. Enlargement occurs by the desquamation of normal cells into a cystic cavity. These unilocular cystic masses expand slowly and produce only mild symptoms. Aural cholesteatomas can be defined as keratinizing squamous

epithelium growing in the middle ear cleft. This abnormal skin growth is characterized by a destructive process involving an accumulation of desquamated keratin arising from the squamous epithelium and pathologically affecting the middle ear or the mastoid process [1, 2].

CASE REPORT

A 3-year-old boy had previously been treated for otitis media with effusion with bilateral grommets nine months ago. He then had excision of a 0.75 cm in diameter of cystic swelling in the left postaural region at a district general hospital.

There were no cholesteatoma or retraction pockets noted in either ear. The histopathology of the cystic swelling removed was reported as an epidermoid cyst. He was then referred to our tertiary hospital, four weeks after his second surgery with a persistent discharging granulation at the site of the left post-aural wound. He was evaluated with pure tone audiometry showing 10db conductive hearing loss in left ear and normal tympanometry. Computerised tomography (CT) of the temporal bones (figure 1) was performed and showed destruction of ossicles of the left ear with a soft tissue shadow filling the left middle ear and the left mastoid cavity with a sinus extending through a defect in the mastoid bone to the subcutaneous tissues in left postaural region. The right ear appeared normal.

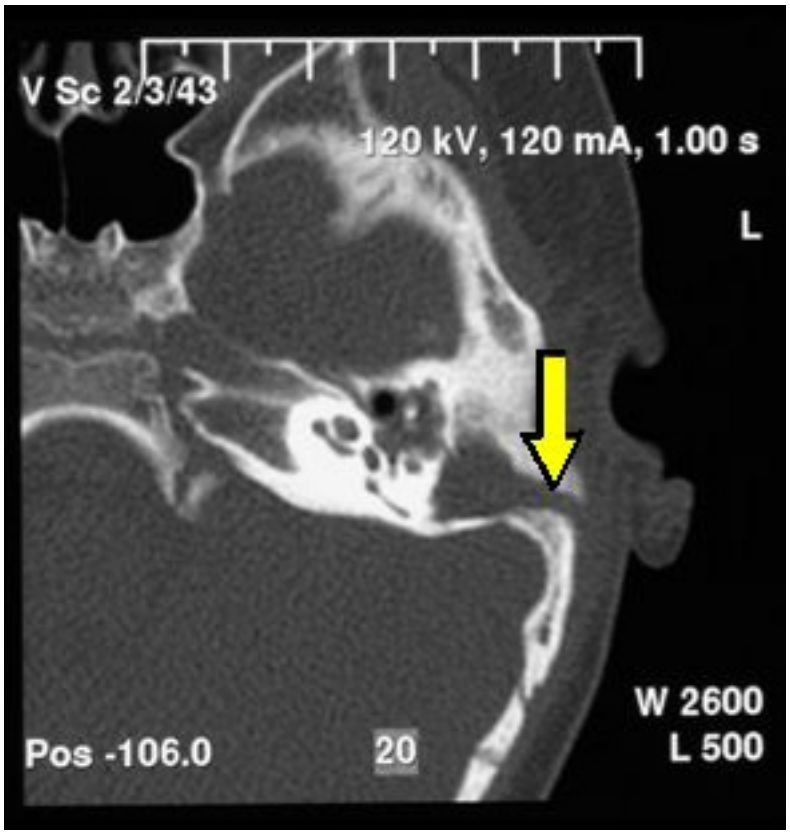

Figure 1: CT scan of Temporal bone. The thick arrow shows the bony discontinuity in the left temporal bone.

A left modified radical mastoidectomy was performed through a postaural approach with an elliptical incision around the postaural granulation. The granulation was found to be in continuity with a tract that extended into a defect in the mastoid bone. The mastoid cavity was exposed to reveal that the tract was in continuity with a cholesteatoma sac within the mastoid (figure 2). The facial canal and the labyrinth were normal. The long process of incus was eroded and the cholesteatoma sac was bridging to preserve the sound conductive mechanism.

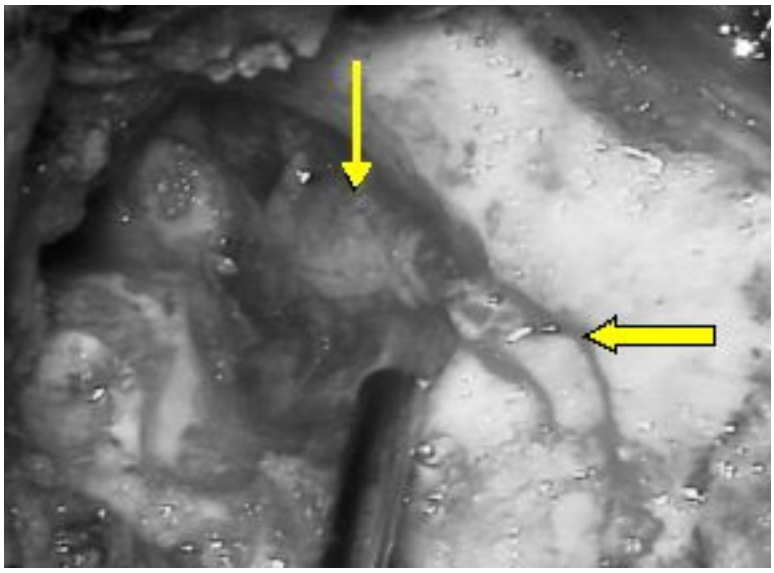

Figure 2: Left Modified Radical Mastoidectomy. The thin arrow indicates cholesteatoma sac and the thick arrow shows the tract communicating from the sac to the postaural skin.

Histopathology of the tissue removed from the mastoid cavity showed features consistent with inflammatory polyp and cholesteatoma. As the tympanic membrane appeared intact at the time of surgery, the cholesteatoma was considered to be congenital.

DISCUSSION

Cholesteatoma is an abnormal accumulation of keratin-producing squamous epithelium in the middle ear, epitympanum, mastoid or petrous apex. It has been further defined as a three-dimensional epidermoid structure exhibiting independent growth, replacing middle ear mucosa, and resorbing underlying bone. Although it is not a neoplastic lesion, it can be insidious and potentially dangerous to the patient. The cystic content is composed of fully-differentiated anucleate keratin squames. The matrix contains the keratinizing squamous epithelium lining a cyst-like structure. The perimatrix or lamina propria is the peripheral part of the cholesteatoma consisting of granulation tissue, which may contain cholesterol crystals.

Bone resorption is stimulated by a variety of factors, including inflammation, local pressure, keratin and specific cytokines, such as interleukins. In addition to destruction of the ossicles, bony erosion can lead to fistula of the lateral semi-circular canal, facial palsy, total sensorineural hearing loss, sinus thrombosis and intracranial invasion [5]. In acute mastoiditis it is common for the pus to erode the lateral wall of the mastoid bone leading to sub-periosteal and then subcutaneous spread of infection. However, this is a much less common mode of spread of cholesteatoma.

Epidermoid cysts and cholesteatomas are characterized by similar tissue layers, namely basal membrane, basal, intermediate and keratin layers. These two types of lesion have very similar histological features and are thus difficult to identify on the basis of microscopic examination in the absence of information on their origins [6].

Congenital cholesteatoma normally presents with only conductive hearing loss. This is the first reported case of congenital cholesteatoma presenting with a post aural subcutaneous cyst-like swelling in a child. A case of subcutaneous cholesteatoma has been described in a 71 year-old man who had previously had a radical mastoidectomy [7] and also another case of congenital cholesteatoma presenting as a preaural swelling with facial palsy [8] has been reported in the literature. In our patient the bony destruction was entirely due to the congenital cholesteatoma, as he had not previously had any mastoid surgery.

This case illustrates the destructive nature of cholesteatoma. The aerated nature of paediatric mastoid air cells system also allows easy extension of such lesion. What was initially thought to be an isolated benign cyst in the postaural region was in fact subcutaneous extension of an underlying cholesteatoma. This case demonstrates the value of pre-operative radiological investigation and the need for a high index of suspicion when excising lesions from unusual head and neck sites.

Corresponding Author:

Dr Ganesh, 44 Maynard Close Erith, Kent, UK. DA8 2BQ.

Email:suganshank@hotmail.co.uk

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