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Porażenie Bella czy perlak wrodzony? Opis przypadku

Bell's palsy or congenital cholesteatoma? A case report

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Streszczenie

Perlak wrodzony jest poważną chorobą dotyczącą populacji pediatrycznej, która znacząco wpływa na jakość życia pacjentów. Prawidłowe rozpoznanie schorzenia może stanowić wyzwanie, ponieważ często przebiega ono bezobjawowo i bywa, że jest stwierdzane przypadkowo. W większości przypadków diagnoza stawiana jest dopiero po pojawieniu się powikłań. Powikłania pozaczaszkowe obejmują m.in. zapalenie wyrostka sutkowatego, ropień podokostnowy i porażenie nerwu twarzowego. Główną metodą leczenia perlaka wrodzonego jest interwencja chirurgiczna (mastoidektomia). Celem jest zwalczenie choroby i zapobieganie późniejszym powikłaniom. Poza zabiegiem operacyjnym pacjenci wymagają długotrwałego leczenia dożylnie podawanymi antybiotykami.

Słowa kluczowe: perlak wrodzony, porażenie nerwu twarzowego

Abstract

Congenital cholesteatoma is a debilitating disease occurring in the paediatric population and affecting the quality of life of patients. However, diagnosing the disease remains a challenge. It might be asymptomatic at presentation, and diagnosed only accidentally. In the majority of cases, it is not diagnosed until complications arise. Extracranial complications include mastoiditis, subperiosteal mastoid abscess, and facial nerve palsy. The main treatment for congenital cholesteatoma is surgical intervention, namely mastoidectomy. The treatment aims to eradicate the disease and prevent subsequent complications. In addition to the surgical procedure, patients require a prolonged course of intravenous antibiotics.

Keywords: congenital cholesteatoma, facial nerve palsy

INTRODUCTION

Cholesteatoma is defined as an accumulation of abnormal keratinising squamous epithelium with a collection of keratin debris and a surrounding inflammatory reaction.

This abnormal growth is locally invasive and has the potential of destroying the middle ear cleft structures. It is categorised as either congenital or acquired. The congenital type forms before birth, occupying the space medial and intact eardrum and has no history of otitis media or previous otologic procedures. Acquired cholesteatomas most commonly begin after birth with a retraction pocket in the eardrum, usually as a result of chronic middle ear disease.

We report the case of a boy aged 2.5 years who first presented with sudden onset right facial nerve palsy for one day, with no other otological symptoms or previous otological procedures. He presented with right otalgia complicated by acute mastoiditis two weeks after facial nerve palsy.

CASE REPORT

A boy aged 2.5 years, with a background history of cleft lip and palate operated on in 2015 was first referred for right facial nerve palsy persisting for one day. His facial asymmetry was noticed by his parents when he was crying, and claimed the boy had a history of upper respiratory tract infection three days before the onset of facial asymmetry. Clinically, he had right facial nerve palsy House–Brackmann (HB) grade 4, and his otoscopic examination revealed tympanic membranes intact bilaterally, with no suspicious masses. At that time, he was diagnosed with Bell's palsy and treated with oral prednisolone at tapering doses and was scheduled a follow-up in one week's time.

Upon the next review in the clinic, the parents complained that the child was crying upon touching his ear and had foul-smelling ear discharge for three days. Clinically, his right facial nerve palsy remained HB grade 4 with his right pinna had displaced forward and laterally, inflamed and tender over the mastoid region. Otoscopic examination showed minimal mucopus in the external auditory canal, with no posterior wall sagging or perforation seen on the tympanic membrane.

The boy was admitted to the ward and treated for right acute mastoiditis with right facial nerve palsy. Despite five days on high doses of intravenous amoxicillin-clavulanic acid, his overall condition did not improve.

High resolution computed tomography of the petrous bone was performed to look for disease extension and disclosed the destruction of the right mastoid air cells involving the external auditory canal and middle ear cavity. There was a soft tissue density lesion within the right external auditory canal, middle ear cavity, and mastoid air cells. The right tegmen tympani, scutum, and ossicles were eroded. The tympanic segment of the right facial canal was seen to be partly eroded; however, the rest of the facial bony canal was intact (Fig. 1).

The patient was scheduled for an emergency right cortical mastoidectomy the following day. Intraoperative findings showed sagging of right posterior wall, with a polyp arising from the middle ear and filling the external canal. Tympanic membrane perforation was seen in the posterior part. Granulation tissue had occupied the middle ear cavity, enveloping the malleus and incus, antrum, and mastoid cavity. Nonetheless, there was no apparent cholesteatoma sac seen. Granulation tissue was sent for histopathological examination.

Postoperatively, the antibiotic was changed to intravenous ceftazidime for three weeks. Upon completion of the antibiotic course, the boy's facial nerve palsy improved to HB grade 3. He was then discharged home.

Upon review at one week after his discharge (one month post cortical mastoidectomy), brainstem evoked response (BSER) was performed to assess his hearing. The result showed right ear moderate HL at least at high frequency, and normal hearing in his left ear. Histopathological examination from granulation tissue revealed cholesteatoma.

Two months post mastoidectomy, the boy's right ear remains dry, his facial nerve has improved markedly to HB grade 1, and the tympanic membrane is intact.

DISCUSSION

Cholesteatoma affects about 10% in chronic otitis media in the paediatric population[1]. Amongst the 10% of the incidence, around 70–96% of cases are acquired in nature[2].

Congenital cholesteatoma presents as a nidus of trapped squamous epithelium medial to an intact tympanic membrane without history of otitis media or previous otologic surgery. The appearance was described as a pearl-like mass seen medial to an intact tympanic membrane. Congenital cholesteatoma can go undiagnosed for many years until the patients notice their hearing is deteriorating[3].

Acquired cholesteatoma is divided into primary and secondary. In primary acquired cholesteatoma, it is observed

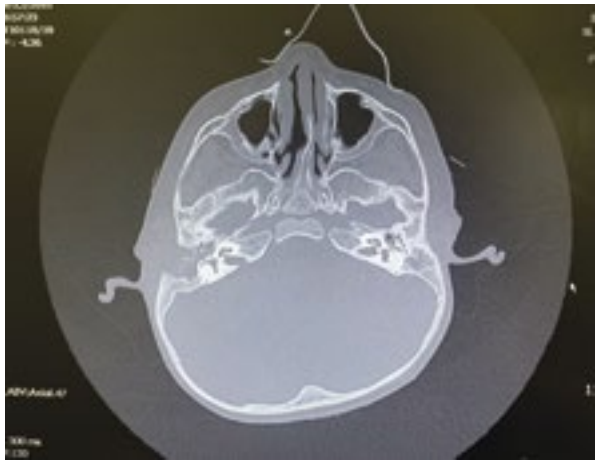

Fig. 1. Computed tomography findings (axial view) showed destruction of right mastoid air cell involving the external auditory canal and middle ear cavity

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that the pars tensa is intact, and a deep retraction pocket (cholesteatoma sac) extending to the attic, probably due to chronic negative pressure, is present. In contrast, in secondary acquired cholesteatoma, with the mixture of chronic middle ear infection, recurrent acute attacks and polyps, granulation tissues, and extensive retraction sacs that pathologically involve the pars tensa are observed[1].

In the case of our patient, there were no otological symptoms prior to his initial presentation of right facial nerve palsy, although his background history of cleft palate may raise some red flags as one of the risk factors conducive to the development of cholesteatoma.

An estimated 0.9–5.9% of children with a cleft palate develop primary acquired cholesteatoma[4]. Among them, it appears that grommet insertion does not alleviate the risk of cholesteatoma, with the incidence rate still ranging from 0 to 6.9% after insertion[5].

Cholesteatoma is known for its local aggressiveness, which may lead to catastrophic implications. The rate of extracranial complications, namely subperiosteal mastoid abscess, facial nerve palsy, mastoiditis, has been reported to be as high as 57% while 40% of cholesteatoma cases are associated with intracranial complications. The intracranial complications include meningitis, cerebral abscess, lateral sinus abscess, to name a few. About 10% of patients have multiple concomitant intracranial and extracranial complications[6]. Cholesteatoma in children has been found to be more aggressive compared to the adult population. The mastoid in children is more cellular and has better pneumatisation compared to that in adults', which is often found to be sclerotic. Due to this feature, the disease is said to be more extensive[7].

In our reported case, we did not suspect the patient's facial nerve palsy to be essentially a complication of cholesteatoma; however, the patient recovered well due to close follow-up and management including a surgical intervention. Surgical treatment remains the main treatment for cholesteatoma and its complications. Antibiotics are used as adjuvant supportive therapy and adjusted according to microbiological sensitivity. The main objectives are to eradicate the disease and restore a healthy aerated ear, preserve and improve the patient's hearing, and prevent residual and recurrent disease. A second-look surgery is sometimes needed in recurrent cases or if the disease was not entirely eradicated during the first intervention[8].

Based on a retrospective study done by Goh et al., up to 25% of patients had recurrent disease. All recurrent cases underwent revision surgeries; however, 15% of the patients still have ear discharges that need frequent aural toileting[9]. Poor cooperation from children in keeping the cavity clean and ineffective care was recognised as a cause of the incidence of recurrence[7].

CONCLUSION

Congenital cholesteatoma remains a challenge for otolaryngologists both in terms of diagnosis and management. However, high-risk cases of developing cholesteatoma should be closely monitored in order to achieve an early diagnosis and reduce morbidity caused by the disease.

Conflict of interest

The authors do not report any financial or personal affiliations to persons or organisations that could adversely affect the content of or claim to have rights to this publication.

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