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CASE REPORT

Extracranial and Temporal Lobe Abscesses Resulting from Cholesteatoma Associated with Congenital Aural Atresia

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Abstract

Objective of this to report an extremely rare case of cholesteatoma (CS) associated with congenital aural atresia (CAA), resulting in extracranial and temporal lobe abscesses.

A 14-year-old boy presented with severe ear pain, right temporal swelling, headache and high fever. Presentation of patient's data, CT imaging and surgical treatment were reviewed. On computerised tomography (CT) he was found to have CAA of the right ear and an extensive CS underlying an extracranial and a temporal lobe abscess. The brain abscess was evacuated through lateral craniotomy. The CS was subsequently removed by modified radical mastoidectomy. CS associated to CAA may remain undiagnosed in early childhood, resulting in life a threatening cerebral abscess formation. Timely detection of hearing loss with CT evaluation would allow for accurate diagnosis and may prevent complications.

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Introduction

Congenital aural atresia (absence of the external ear canal) is a defect almost always accompanied by a variable degree of abnormalities of both the external ear and the bones of the middle ear. In the literature several classifications and grading systems for this condition have been proposed.1 The incidence is estimated at 1 in 10,000 live births.[2] It is usually associated with microtia and it may also be accompanied by congenital CS.[3] An undiagnosed slow-growing CS could cause extensive damage to the middle ear and the surrounding tissues. Accurate diagnosis requires a high index of clinical suspicion and familiarity with the signs and symptoms of an expanding mass in the mastoid region. Imaging CT studies are diagnostic. Congenital aural stenosis (CAS) associated with cholesteatoma and otogenic extratemporal complication was already published,[4] but only one previous case of an extracranial complication of CAA has been published.[5] We describe a case of both extracranial and intracranial complications due to extensive CS developing in the context of CAA.

Case Report

A 14-year-old boy was admitted to our emergency unit with a two day history of right temporal swelling and fever (39˚C). His complaints included vertigo, severe headache and progressive right ear pain not responding to analgesic medication. On examination he appeared fully conscious (GCS 15/15). He was noted to have complete atresia of the right external ear canal and microtia associated to hearing loss on that side. At the age of [3], he had undergone reconstructive surgery of the right ear helix in another institution. Audiometry or CT had not been undertaken at the time. His malformed ear was covered by long hair and did not appear obvious. On admission, pure tone audiometry showed a moderate right sided mixed hearing loss. The air conduction threshold in the atretic ear was at 60 dB HL range with a compatible speech reception threshold. The bone conduction threshold on the same side was within normal limits. CT of the temporal bone

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and endocranial region confirmed complete atresia of the right external auditory canal. The middle ear of the involved side showed a poorly developed cavity and reduced mastoid cells (Figure 1). The small cavity of the right middle ear contained a soft tissue mass extending into the endocranial space. The findings were radiologically in keeping with CS. There was also an inflammatory process spreading laterally into the overlying connective tissue, forming a subcutaneous abscess (Figure 2). This also extended to the right temporal lobe forming an intracerebral abscess (Figure 3). The patient was commenced on a broad spectrum of antibiotics. The right temporal lobe abscess was evacuated through lateral craniotomy. Following improvement in patient's general condition a radical modified right mastoidectomy was performed. Within the small cavity of the middle ear occupied by suspected cholesteatoma, malformed middle ear ossicules were identified. The cholesteatoma was removed. A reconstruction of the middle ear ossicular chain and the ear drum was undertaken with a modification of the maleus-incus complex. On pathological examination diagnosis of cholesteatoma was confirmed. The postoperative course was uneventful. The five year follows up CT scan revealed no recurrent cholesteatoma or inflammation. The follow up audiometry indicated that the surgical middle ear conductive chain reconstruction did not significantly improve the hearing loss.

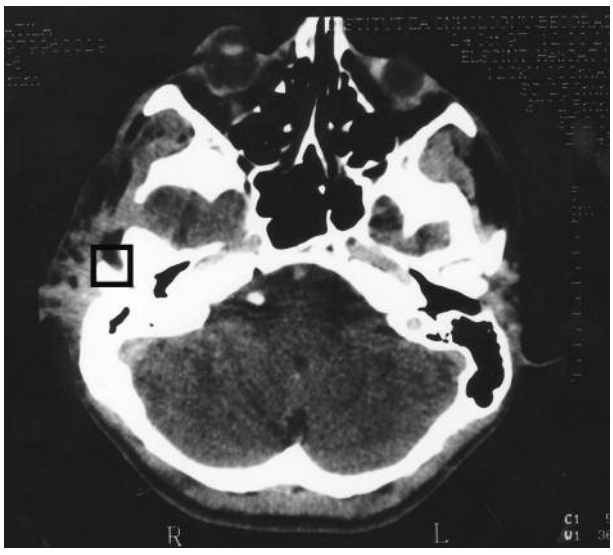

Figure 1. CT scan shows atretic ear (black window) of the right side

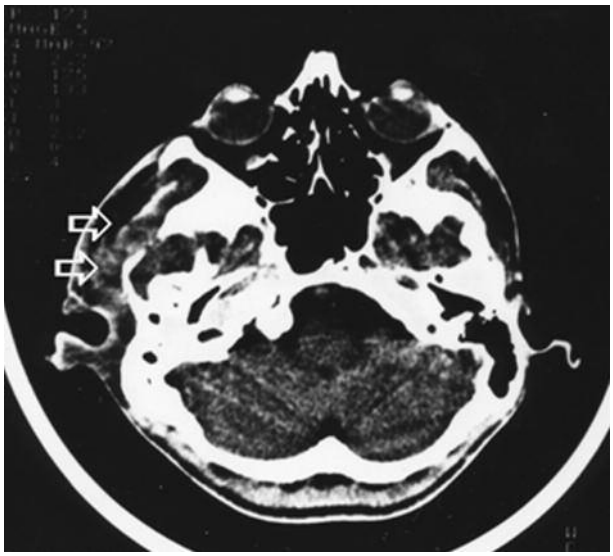

Figure 2. CT scan presents extension of cholesteatoma and inflammation with extracranial abscess in the region of temporal muscle (arrows)

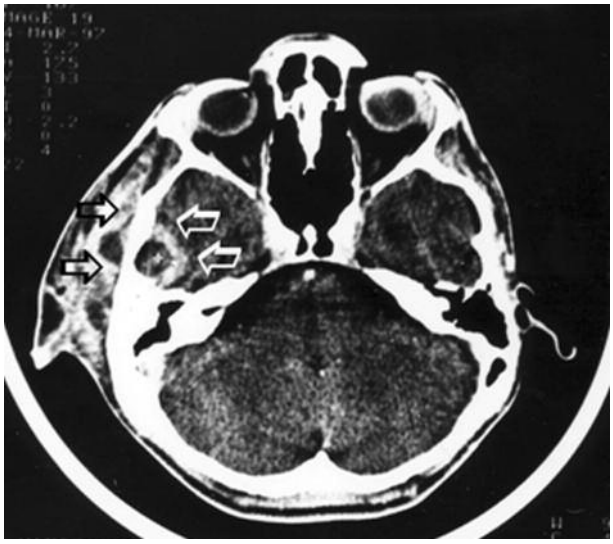

Figure 3. CT demonstrates abscess (star) in right temporal brain lobe (arrows))

Discussion

The dominating feature of CAA is absence of the external auditory canal. The severity of this deformity is variable. It generally depends on the point in time during intrauterine development at which the arrest of the evolving ear structures takes place.[6,7] In its most severe form, as in our case, the developmental arrest

occurs early during intrauterine growth and the ear canal is not formed at all. Patients affected by CAA are more often males. It is more commonly unilateral, more frequently involving the right ear.[8] Patients with severe cases of bilateral CAA require vigorous hearing rehabilitation with a bone conducting hearing aid. Adults with CAA usually presented to otologists with unilateral conduction hearing loss that was not corrected during childhood. If the auditory function in the unaffected ear is normal,[9] further evaluation of unilateral CAA in children is not necessary until the age of 5-6. At this time, a high resolution CT scan is recommended to assess the extent of the disease and consider surgical repair.[10] A CT scan with and without contrast would also serves as a reliable tool to detect CS. An earlier CT assessment would be required for patients with CAA who develop symptoms such as secretion from the ear or acute facial nerve palsy which may all be induced by an underlying CS.

In our case, microtia had been detected and was reconstructed for cosmetic reason. At the time, this had not been aided by additional imaging investigations. Further evaluation at the recommended age of 5-6 was also not carried out as it might have been assumed by the parents of the patient that the underlying problem had been corrected. The caring physicians also failed to indicate a necessity for later detailed clinical and radiological evaluation. It is thus likely that the opportunities had been missed to detect an asymptomatic CS. The clinical presentation of CS depends on the site of involvement. If in the middle ear, it will result in deafness during early childhood, as in our case. Early diagnosis and surgical management of CS would prevent potential late complications.[11] It would be advisable that patients with one sided atretic ear are checked up every 6 months. Alternatively, the process may remain undiscovered and result in life threatening complications as in our case.

Congenital CS associated with CAA is not uncommon.[12,13] While its prevalence in this context increases with the advancing age of the patients,[14] the associated extracranial otogenic complications are extremely rare.[15,16] However, congenital CS has a well recognised capacity for bone destruction. In our patient it resulted in an invasion and destruction of the tegmen plate of the middle ear, establishing an open communication between the cranial cavity and the middle ear. Brain abscesses may develop as a complication of bacterial middle ear infections but also as a result of insidiously growing subclinical CS which may reach a considerable size before producing symptoms.[17] CT is an examination of choice for the diagnosis of both CS and brain abscesses. In our case the diagnosis was based on the CT rather than clinical findings. The CT verified the anatomical proximity of the temporal lobe abscess to the underlying CS and confirmed the causative sequence.

The choice of management for intracerebral abscesses depends on the features of each individual case and the suspected causative agents. It usually comprises a combination of a course of antibiotics with surgical debridement and drainage.[18,19] In all cases early diagnosis is pivotal with the identification of the source of infection. This is particularly important for cerebral abscesses resulting from a direct communication between the cranial cavity and the surrounding anatomical structures. In such cases the surgical intervention is also aimed at re-establishing the normal anatomical barriers and removing the underlying primary pathological process such as the middle ear CS in our patient.

However, recommendations regarding surgical management of the otogenic abscess differ. Some surgeons advocate early craniotomy and excision of the abscess,[20] whereas others prefer combined approach with abscess drainage first followed by mastoidectomy in the same setting.[21] Others have advocated a radical mastoidectomy approach with evacuation of the abscess through the masoidectomy.[22]

The timing of both otologic and neurosurgical treatment are also controversial. Some authors recommend that intracranial surgery should be done first and otologic surgery should be scheduled several weeks later.[23] Sing and Maharaj[24] advocate that otologic surgery should not be delayed and should be carried out immediately after the neurosurgical procedure under the same anaesthesia. Hafidh et al.[25] also recommend that treatment of ear disease be performed at the same time as drainage of the brain abscess. By doing this, the necessity for a second operation can be avoided, and the infected ear can be eliminated as a source of intracranial sepsis, thus preventing seeding of organisms to the brain. In our case the modified radical mastoidectomy was

undertaken as a second operation after allowing the patient to recover from the life threatening complication.

Given the significant complications, resulting from cholesteatoma, the mastoidectomy is necessary to determine the underlying pathology and to control the disease process.[26]

In summary, multidisciplinary assessment of children with unilateral CAA is required before surgery is undertaken for cosmetic or other reasons. The multidisciplinary team should involve an ENT surgeon, plastic/reconstructive surgeon and an audiologist. A clear schedule of diagnostic interventions to include a timely application of CT should be defined. This should prevent late and life threatening complications such as intracranial infections as a result of undetected CS associated to CAA. Clinicians should be aware of this rare but serious complication of CS in the context of CAA.

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References

1. Jahrsdoerfer RA, Yeakley JW, Aguilar EA, et al. Grading system for the selection of patients with congenital aural atresia. Am J Otol 1992; 13:6-12.

2. Schuknecht HF. Congenital aural atresia. Laryngoscope 1989; 99:908-17.

3. De La Cruz A, Linthicum Jr. FH, Luxford WM. Congenital atresia of external ear. Laryngoscope 1985; 95:421-7.

4. Yildrim N, Şahan M, Akkaya A. Case report: Primary cholesteatoma within the stenotic ear canal: report of two cases, one with extra-temporal complications. Int Adv Otol 2009; 5:2.

5. Zalzal GH. Acute mastoiditis complicated by sigmoid sinus thrombosis in congenital aural atresia. Int J Pediatr Otorhinolaryngol 1987; 14:31-9.

6. Yeakley JW, Jahrsdoerfer RA. CT evaluation of congenital aural atresia: What the radiologist and surgeon need to know. J Comput Assist Tomogr 1996; 20:724-31.

7. Nishimura Y, Kumoi T, Sano S. Cholesteatoma auris congenital arising from microtia. Ann Plast Surg 1985; 14:296-300.

8. Doyle KJ, Ray RM. The otolaryngologist role in management of hearing loss in infancy and childhood. Ment Retard Dev Disabil Res Rev 2003; 9:94-102.

9. Declau F, Cremers C, Van de Heyning P. Diagnosis and management strategies in congenital atresia of the external auditory canal. Study group on otological malformations and hearing impairment. Br J Audiol 1999; 33:313-27.

10. Tasar M, Yetiser S, Yildirim D, Bozlar U, Tasar MA, Saglam M, et al. Preoperative evaluation of the congenital aural atresia on computed tomography. An analysis of the severity of the middle ear and mastoid. Eur J Radiol 2007; 62:97-105.

11. Savic D, Djeric D. Congenital malformations of the ear associated with secretory otitis media. Rev Laryngol 1989; 110:479-80.

12. Miyamoto RT, Fairchild TH, Daugherty HS. Primary cholesteatoma in the congenitally aretic ear. Am J Otol 1984; 5:283-5.

13. Caughey RJ, Jahrsdoerfer RA, Kesser BW. Congenital cholesteatoma in case of congenital aural atresia. Otol Neurotol 2006; 27:934-6.

14. Cole RR, Jahrsdoefer RA. The risk of cholesteatoma in a case of congenital aural atresia. Laryngoscope 1990; 100:576-8.

15. 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.

16. Pawelczyk M, Czarski J. Congenital cholesteatoma of the middle ear in a child with aural atresia complicated by mastoid abscess. Otolaryngol Pol 1998; 52:209-13.

17. Yamane H, Takayama M, Sunami K, Tochino R, Morinaka M. Disregard of cholesteatoma in congenital aural stenosis. Acta Otolaryngol 2007; 127:221-4.

18. Kurien MS, Job A, Mathew J, et al. Otogenic intracranial abscess. Arch Otolaryngol Head Neck Surg 1998; 124:1353-6.

19. Nalbone VP, Kuruvilla A, Gacek RR. Otogenic brain abscess: the Syracuse experience. Ear Noe Throat J 1992; 71:238-42.

20. Kulali A, Ozatik N, Topcu I. Otogenic intracranial abcesses. Acta Neurochir 1990; 107:140-6.

21. Murthy PSN, Sukumar R, Hazarika P, et al. Otogenic brain abscess in childhood. Int J Pediatr Otolaryngol 1991; 22:9-17.

22. Sennaroglu L., Sozeri B. Otogenic brain abscess: review of 41 cases. Otolaryngol Head Neck Surg 2000; 123:751-5.

23. Kangsanarak J, Navacharoen N, Fooanant S, et al. Intracranial complications of suppurative otitis media: 13 years, experience. Am J Otolaryngol 1995; 16:104-9.

24. Singh B, Maharaj TJ. Radical mastoidectomy: its place in otitisc intracranial complications. J Laryngol Otol 1993; 107:1113-8.

25. Hafidh M, Koegh I, Walsh Mc Conn R, Walsh M, Rawluk D. Otogenic intracranial complications. A 7-year retrospective review. Am J Otolaryngol 2006; 27:390-395.

26. Wanna G, Dharamsi L, Moss J, Bennet M, Thompson R, Haynes M. Contemporary management of intracranial complications of otitis media. Otol Neurotol. 2010; 31:11-117.