

Case Report

Adult-Onset Labyrinthine Fistula as a Sign of Undiagnosed Bilateral Congenital Cholesteatoma

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Received 7 August 2012; Accepted 12 April 2013

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Abstract Objective. Bilateral congenital cholesteatoma is a very rare condition. We describe the first case of an adult patient presenting with bilateral congenital cholesteatoma with labyrinthine fistula. **Case report.** We present a 34-year-old woman of bilateral congenital cholesteatoma with labyrinthine fistula, inducing left-sided mixed conductive-sensorineural hearing loss and vertigo, subsequently accompanied by right-sided conductive hearing loss. Otoscopic examination and computed tomography scan revealed bilateral congenital cholesteatoma with a fistula in the left lateral semicircular canal. Tympanomastoidectomy was performed, and both congenital cholesteatoma were completely removed, and membranous fistula of the left lateral semicircular canal was covered with a graft. **Conclusion.** Congenital cholesteatoma is usually detected in childhood not but in adulthood. With delay in diagnosis, the risk of cholesteatoma complications increases. Greater attention should be paid to complications such as labyrinthine fistula resulting from long-term exposure to congenital cholesteatoma in adults.

Keywords congenital cholesteatoma; labyrinthine fistula; sensorineural hearing loss

1. Introduction

Bilateral congenital cholesteatoma is a very rare condition. Since Peron et al. reported the first case of bilateral congenital cholesteatoma, several cases have been reported [11]. Bilateral congenital cholesteatoma has been reported in patients aged 2 to 23 years [10]. House reported that the incidence of congenital cholesteatoma is 3.7% [5], and incidence of bilateral congenital cholesteatoma has been estimated to be less than 3% of all congenital cholesteatomas [4]. Although both acquired and congenital cholesteatoma can lead to several complications, e.g., ossicular erosion, facial palsy, intracranial infection, and labyrinthine fistula, these complications tend to present less often in congenital cholesteatoma than in acquired cholesteatoma [7]. There are no reports of labyrinthine fistula resulting from bilateral congenital cholesteatoma in the international literature. We describe the first adult case of bilateral congenital cholesteatoma with lateral semicircular canal fistula.

2. Case presentation

A 34-year-old woman presented with a 10-day history of left-sided hearing loss and a 3-day history of vertigo. She had no history of otorrhea or ear surgery. The left ear membrane was slightly dull, while the right ear membrane was intact. Pure-tone audiometry showed left-sided mixed conductive-sensorineural hearing loss of 63 dB (average of 0.5, 1, and 2 kHz) (Figure 1(a)). She had gaze nystagmus toward the right. A computed tomography (CT) scan revealed areas of low soft tissue density in the superior quadrant, the epitympanum, and the mastoid of the left middle ear cavity. On the left side, all ossicles of the malleus, incus, and stapes were eroded and, the lateral semicircular canal showed a fistula (Figures 2(a) and 2(b)). On the right side, CT scan showed an area of slight soft tissue density in the epitympanum and the mesotympanum with intact ossicles. Hydrocortisone sodium succinate (500 mg for 2 days, 300 mg for 2 days, and 100 mg for 2 days) was intravenously administered for 6 days to prevent acute inner ear damage. Nevertheless, the threshold of

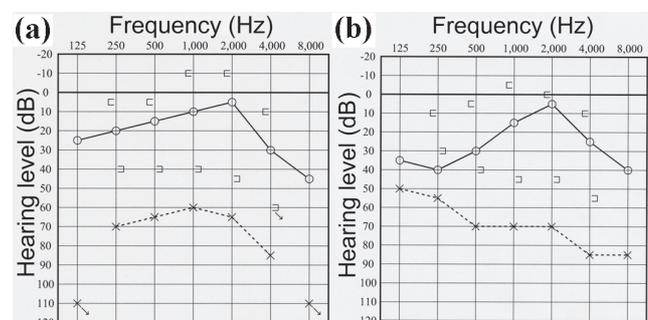


Figure 1: Pure-tone audiometry showing mixed conductive-sensorineural hearing loss (a) on the left side at the first episode of hearing loss and vertigo, and conductive hearing loss (b) on the right side at the second episode.

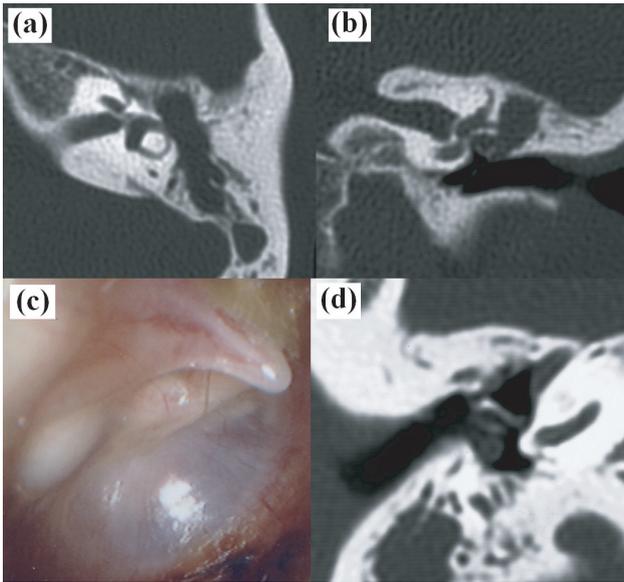


Figure 2: CT scan of the left temporal bone reveals an area of soft density in the epitympanum and lateral semicircular canal fistula in the axial (a) and coronal (b) views. A white mass was observed through the posterior superior quadrant of the right tympanic membrane (c). A round area of soft density was displayed behind the tympanic membrane on the right axial view of CT scan (d).

pure-tone audiometry was elevated to 87 dB. Six days later, left exploratory tympanomastoidectomy was performed. The cholesteatoma matrix extended into the epitympanum and the mastoid, eroding the malleus, incus, and stapes; and the left lateral semicircular canal showed a membranous fistula. The whole cholesteatoma, malleus head, and incus were carefully removed, including the matrix covering the lateral semicircular canal fistula. We did not suction the perilymph leaking out of the fistula. The fistula was covered with a fragment of the perichondrium and the temporalis fascia graft. Tympanoplasty was performed. The bone-conductive hearing level did not deteriorate and remained almost unchanged after the surgery. Six weeks after the surgery, the air-conductive hearing level deteriorated, showing conductive hearing loss of 17 dB on the right side (Figure 1(b)). Around the same time, a white mass could be observed through the posterior superior quadrant of the right tympanic membrane (Figure 2(c)). Re-examination of the CT scan revealed a small round mass behind the posterior superior quadrant of the left tympanic membrane (Figure 2(d)). Because cholesteatoma on the right side was suspected, exploratory tympanoplasty was performed. A small white cholesteatoma was adhered to the incus long process and the incudostapedial joint. Therefore, tympanomastoidectomy (canal wall up method) was performed to remove the cholesteatoma matrix, including

the incus. We performed type III modified tympanoplasty laying cartilage as a columella between the stapes head and the tympanic membrane. Bilateral congenital cholesteatoma was diagnosed, according to the criteria for congenital cholesteatoma described by Levenson et al. [6].

3. Discussion

In 1965, Derlaki and Clemis defined congenital cholesteatoma of the middle ear as a white mass detected behind an intact tympanic membrane with normal pars tensa and pars flaccida in a patient with no history of otorrhea, tympanic membrane perforation, or otologic surgery [3]. Levenson et al. revised the criteria in 1986 to include patients with previous episodes of acute otitis media [6]. The etiology of congenital cholesteatoma is controversial; and various theories exist, such as implantation [4,5], invagination [14], metaplasia [15], and epidermoid formation [8]. However, etiology is still unclear. The most common site of congenital cholesteatoma is the anterior superior quadrant of the mesotympanum, followed by the posterior superior quadrant [9]. The most common sites of acquired cholesteatoma are the epitympanum and the posterior superior quadrant of tympanum.

To date, only a few studies have reported labyrinthine fistula in congenital cholesteatoma. The reported frequency of labyrinthine fistula in congenital cholesteatoma (0% to 5%) [5,7,12,13] is less than the frequency of acquired cholesteatoma (7%) [1]. However, Darrouzet et al. reported a relatively higher rate (8.8%) of labyrinthine fistula in congenital cholesteatoma [2]. They attributed this to differences between health care systems in Europe and North America: the mean age at diagnosis is greater in Europe than in North America. Our case presented fistula of the lateral semicircular canal. We postulate that the risk of labyrinthine fistula is related to various factors, such as age at diagnosis, location, and infection. First, with delay in diagnosis, the cholesteatoma stage becomes more advanced. Long-term exposure to cholesteatoma matrix increases the probability of destruction of the labyrinth. Second, the most common site of congenital cholesteatoma, the anterior superior quadrant, is farther from the lateral semicircular canal and oval window than the usual site of acquired cholesteatoma, i.e., the posterior superior quadrant. Third, infection accelerates the destructive feature of the cholesteatoma matrix. Generally, congenital cholesteatomas are free of infection, because the cholesteatoma matrix is separated from the external ear canal. These factors will determine the destructiveness of the cholesteatoma. Our patient had two disadvantages: long-term exposure to the cholesteatoma and anatomical proximity to the lateral semicircular canal.

Congenital cholesteatoma is usually detected in childhood during medical screening. However, it is difficult to

detect if otoscopy findings are normal, and there are no complications, such as hearing loss or facial palsy. In such cases, diagnosis of congenital cholesteatoma is delayed, possibly until adulthood, as in the case presented here. Even in adults, congenital cholesteatoma, including bilateral congenital cholesteatoma, may not be detected. Especially in cases of adult congenital cholesteatoma, attention should be paid to potential complications, including labyrinthine fistula, because of long-term exposure to congenital cholesteatoma.

4. Conclusion

We present the first case of bilateral congenital cholesteatoma with lateral semicircular canal fistula; this case was especially unusual because of adult onset. With delay in diagnosis, the risk of complications increases. Greater attention should be paid to potential complications, such as labyrinthine fistula, in cases of adult congenital cholesteatoma because of long-term exposure to congenital cholesteatoma.

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