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Case report

Non-classical presentation of congenital cholesteatoma as cerebrospinal fluid rhinorrhea – Case report and systematic review of the literature

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ABSTRACT

Objective: Congenital cholesteatoma (CC) becomes clinically apparent as a cholesteatoma usually during childhood. Nontraumatic otogenic cerebrospinal fluid (CSF) rhinorrhea with an intact tympanic membrane is a very rare symptom.
Methods: The review of recent literature and case report of the 60-year old patient – a trumpeter presented with nontraumatic otogenic CSF rhinorrhea, intact tympanic membrane on microotoscopy, and besides colorless fluid in right nasal cavity, normal finding on nasal endoscopy examination.
Results: CSF rhinorrhea was caused by CC in the petrous bone apex. Early diagnosis was facilitated by computed tomography scanning. Complete cholesteatoma removal was accomplished using a middle fossa craniotomy and an open non-radical antromastoidectomy.
Conclusion: The diagnosis of otogenic cerebrospinal fluid (CSF) rhinorrhea is challenging and it can easily be misdiagnosed. Congenital cholesteatoma is a rare entity. We present a non-classical presentation of CC in an adult male, with a previously unreported symptom of CSF rhinorrhea. Symptomatic improvement occurred after surgical treatment of the disease.

1. Introduction

Petrosal cholesteatomas are rare lesions, which may be primary or acquired in the nature. Primary – congenital cholesteatoma (CC) are unusual. They may arise in different sites of the temporal bone: petrous process, middle ear and mastoid. CC originates from embryonary ectodermal inclusions but the pathogenesis is not clear and many different hypotheses regarding its origin have been suggested. It can

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present at any age from birth to early adulthood but become clinically apparent as a cholesteatoma usually during childhood. CC may be asymptomatic or manifest as a conductive hearing loss. Other presentations include otalgia, vertigo and facial palsy. Manifestation of cerebrospinal fluid (CSF) rhinorrhea has not previously been reported as a representing complaint. CC is diagnosed on the basis of a specific set of criteria: (i) cholesteatoma sac medial to intact tympanic membrane; (ii) a normal tympanic membrane; and (iii) no history of tympanic membrane perforation, otorrhoea, trauma or surgery [1]. Early diagnosis of CC is important to prevent extensive surgery and complications.

Cerebrospinal fluid (CSF) rhinorrhea is characterized by a leak through a communication between the subarachnoid space and the nasal cavity or paranasal sinuses through osteodural defects at the base of the skull. CSF leaks can be classified according to etiology, either as traumatic (accidental and iatrogenic) or nontraumatic. Nontraumatic CSF fistulas are further divided into high-pressure (associated with tumors or hydrocephalus) and normal-pressure leaks (associated with congenital anomalies) [2]. Spontaneous CSF fistula is idiopathic, separate entity from other nontraumatic causes, and accounts for 3–4% of leaks [3]. In addition, CSF rhinorrhea can occasionally occur from the temporal bone through the Eustachian tube into the nasal cavity, and is called otogenic cerebrospinal fluid rhinorrhea. In most of those cases CSF rhinorrhea is associated with CSF otorrhea, tympanic membrane perforation or middle ear diseases such as cholesteatomas which make the clinical diagnosis easier. The clinical diagnosis is not easy if the tympanic membrane is intact. In such cases computerized tomography and magnetic resonance imaging are useful [4].

We present a rare case of petrosal CC located in the petrous process and the mastoid without affection of the tympanic cavity with an atypical presentation in the form of nasal CSF leak.

2. Case report

A 60-year-old man, a trumpeter, was referred to our center for evaluation due to the 5 months history of unilateral leak of colorless fluid through the right nostril, varying with postural changes. Episodes of rhinorrhea were continuing for few seconds. On admission, the patient did not manifest focal neurological signs, except for profound hearing loss in the right ear associated with recurrent otitis media with effusion in childhood. The patient revealed no history of tympanic membrane perforation, otorrhoea, trauma or surgery. His blood pressure had always been good controlled. The ENT examination revealed colorless fluid in right nasal cavity which was taken for biochemical examination, and the intact but opaque tympanic membrane. Pure tone audiometry recorded over 90 dB hearing loss (Pure Tone Average 500–1000–2000 Hz) that corresponded with the profound degree of hearing loss in the right ear and normal hearing in the left ear (≤25 dB of hearing loss). The biochemical result of the fluid samples raised suspicion of CSF. On the nasal fiberoscopy the origin of a CSF leak was not found. The head and paranasal sinuses CT scan did not show pathological changes and bone dehiscence. Magnetic resonance imaging (MRI) revealed a cyst of the hypophysis dimension of 3 mm and intermediate signal intensity on T1-images, high signal intensity on T2-weigted images, and, on postgadolinium images, a thin rim of enhancement representing cholesteatoma with adjacent granulation tissue in the right petrous process (data based on the radiologist's description, no scans available). Due to the MRI result and patient's history of profound hearing loss in the right ear, a CT scan of the temporal bone was performed. It showed opacification of the antrum, mastoid air cells and tympanic cavity by CSF on the right side, as well as dehiscence of the internal auditory canal wall and the fistula on the superior semicircular canal. In addition, the image also revealed partial destruction of air cells in the petrous process and the mastoid through low radiological density pathological formation (6 HU – Hounsfield Units) (Fig. 1).

During hospitalization the patient developed fever – 38.5 °C. For this reason, lumbar puncture was performed excluding meningitis and the antibiotic therapy was implemented (Biotraxon, Dalacin). Given the bony dehiscence in the internal auditory canal wall, pathological changes in the temporal bone, and the risk of infection, surgical treatment was recommended.

3. Operation

The patient underwent initially right middle fossa craniotomy in order to revise the anterior wall of the temporal bone pyramid where the bony dehiscence and meningocele were expected. This approach excluded both bony dehiscence and meningocele as a origin of the CSF leak. Subsequently, an open non-radical antromastoidectomy on the right side was

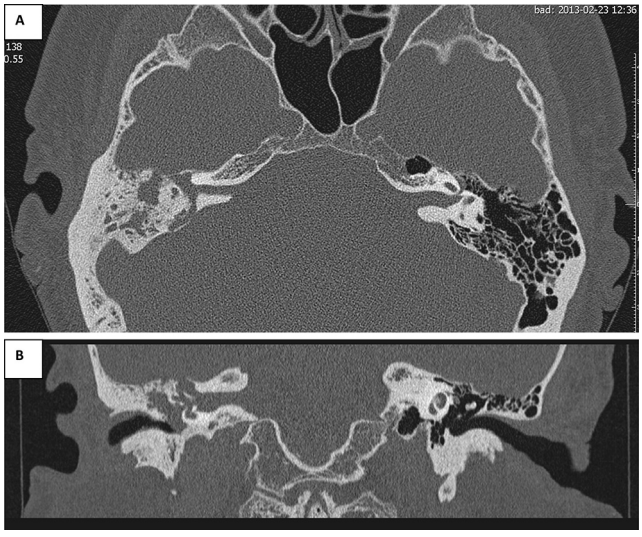

Fig. 1 – Preoperative axial (A) and coronal (B) CT scans reveal opacification of the antrum, mastoid air cells and tympanic cavity by CSF on the right side, as well as dehiscence of the internal auditory canal wall. The image also reveal partial destruction of air cells in the petrous process and the mastoid through low radiological density pathological formation (6 HU – Hounsfield Units).

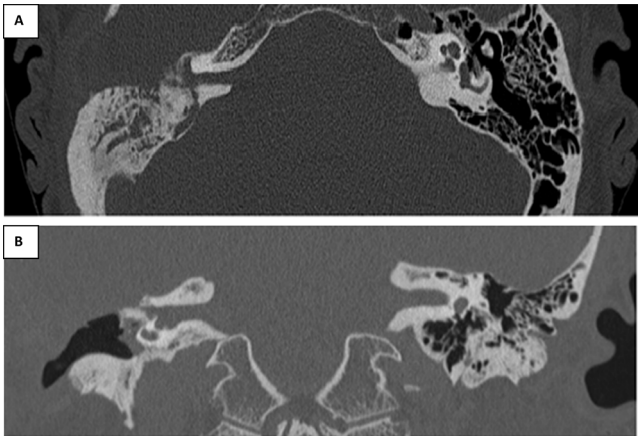

Fig. 2 – Postoperative axial (A) and coronal (B) CT scans reveal on the right side wide surgical defect of mastoid process and tympanic cavity completely filled with the soft tissue mass representing obliteration material.

performed and an extensive CSF leak was observed. After the identification of the tympanic segment of the facial nerve, the cholesteatoma was found in the anterosuperior part of the temporal bone pyramid. Complete cholesteatoma removal was performed step by step allowing preservation of the facial nerve continuity. Due to the extensive CSF leak and the CC limited to the anterosuperior part of the bony pyramid further drilling of the mastoid process was ceased. The bone defect, as a source of CSF leak, was identified at the posterior wall of the petrous pyramid, at the lateral wall of the internal auditory canal. A temporal fascia graft, fibrin glue, and collagen patch TachoSil treated the fistula and CSF leak. Eradication of all accessible air cell tracts and mucosa in the petrous pyramid, obliteration of the eustachian tubal orifice, closure of the external auditory canal, and obliteration of the middle ear and mastoid clefts were essential in this procedure. Bony defects were repaired through the use of an acrylic mass. A lumbar drain was implemented to the subarachnoid space and removed after seven days. The patient has been followed for the next 6 months without rhinorrhea recurrence. Early after the surgery the right facial nerve paralysis House–Brackmann grade VI (H–B) [5] was observed and it recovered to H–B grade II after 6 months of rehabilitation. The deafness of the right ear remained. The follow-up 13 months after the surgery included nasal endoscopy, otoscopy, CT and MRI scan (Figs. 2 and 3). No CSF leak, meningocele or residual cholesteatoma were observed.

4. Discussion

CC develops from a congenital epithelial cell crest in the temporal bone and may present at any age from infancy to adulthood with a mean age of 16.7 years [6]. CC is usually asymptomatic in the initial stages of growth and microotoscopy cannot reliably predict the extent of disease; therefore, radiological evaluation with CT scan should be considered [7]. This case confirms that CC may be manifested non-classically in an older age, on an uncommon location without the appearance of a white mass behind the tympanic membrane and otogenic CSF rhinorrhea. The diagnosis of CSF rhinorrhea in such case is challenging.

CC is a very rare pathology that may cause severe functional damage and may be unilateral or bilateral. It is known to invade frequently the labyrinth and Fallopian canal causing facial nerve paralysis and congenital hearing loss, which is usually progressive through the childhood. The most common presenting symptom, hearing loss in the affected ear, occurred in 60% of reported patients [1,6,8–14]. However, as measured by pure tone audiometry at presentation, 95% had a significant hearing loss [15,16]. Rates for facial nerve weakness preoperatively varies in series from 20 to 64% [16–18]; vertigo/imbalance affected 30–61% of patients before surgery [19]. Tinnitus, otalgia, and headaches were less common manifestations. It may also occur in association with multiple episodes of meningitis [20,21]. From a careful review of the literature, however, we did not find case report of CSF rhinorrhea caused by CC (Table 1).

The most common cause of CSF rhinorrhea is a fistula is the floor of the anterior cranial fossa, predominantly in the cribriform plate and/or the fovea ethmoidalis (roof of the ethmoidal sinus). Rarely, the defect is located in the roof of the sphenoid sinus or on the tegmen tympani in the temporal bone (the roof of the middle ear cavity). The enclosed Table 2 shows literature review of otogenic rhinorrhea. There is no available current data on otogenic CSF rhinorrhea in the course of CC. Based on the review of the literature, we observed that otorhinorrhea resulted mainly from the closed head injury, surgery (especially operation for tumor of the eighth nerve or destruction of the apex of the pyramid) [22–27] and the thinness of the tegmen because of well-pneumatized mastoid air cells and the weakness of the dura after chronic inflammatory changes [28–30]. Tumors and infections can cause skull base erosions and secondary CSF leaks. Spontaneous cases of otogenic CSF account for 4% of CSF leaks [31,32], which have been described through the Mondini malformation [22] and through thin areas of the tegmen and in the wall of the middle cranial fossa into the sphenoid sinus. In those cases CSF leak occurred because CSF seeped through the auditory tube into the nasopharynx and the nasal cavity or through the tympanic membrane into the external acoustic meatus.

We describe for the first time the patient who had a CSF fistula at the lateral wall of the internal auditory canal caused by CC, manifesting as a nasal CSF leak with an intact but opaque tympanic membrane. The lateral wall of the internal auditory canal is a thin bony plate known as the lamina cribrosa, through which pass the nerve fibers of the seventh and eighth cranial nerves and blood vessels. The lamina cribrosa has a dural and arachnoid covering medially and forms a barrier between the inner ear and the subarachnoid space [33]. The lamina cribrosa is not a common site for CSF fistula; such location has been associated with Mondini deformity, in which there is bony and dural dehiscence along with cochlear and vestibular abnormalities [34]. The internal auditory canal can also be a source of CSF leakage after surgery for vestibular schwannoma [22]. Postoperative CSF leakage into the middle ear cavity has been reported through the

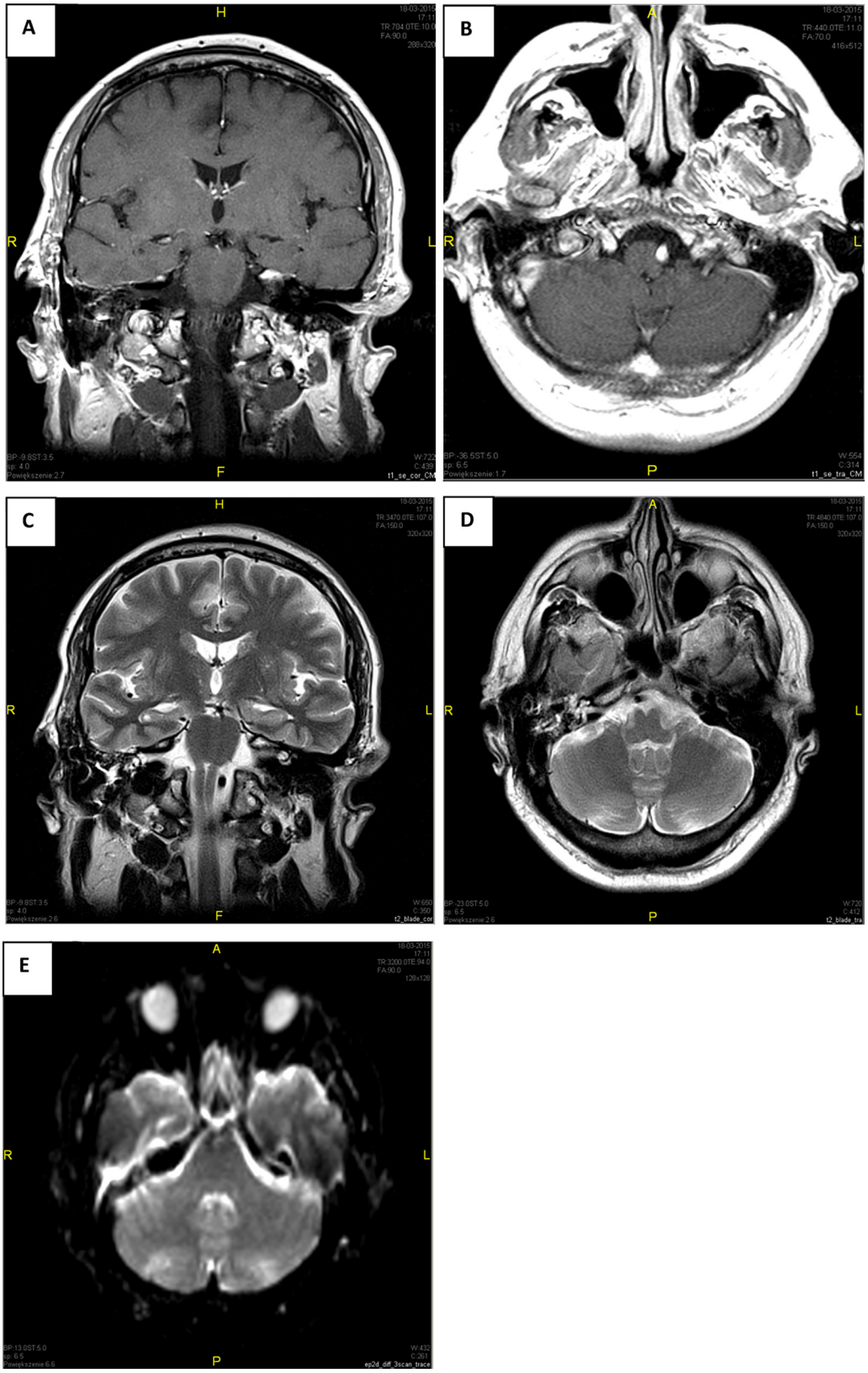

Fig. 3 – Postoperative coronal and axial T1 postgadolinium (A, B) and T2-weigted (C, D) MRI scans and diffusion-weighted imaging (DWI) (E) demonstrate high signal intensity areas within the mastoid and tympanic cavity representing scarring and the obliteration material. Internal auditory canal, facial and vestibular nerves are intact.

Table 1 – Congenital cholesteatoma – systematic review of literature.

Author	Number of cases	Patient age	Main symptoms
Weber and Adkins [1]	3	1, 3, 14 years old	A mild conductive hearing loss on audiometry, symptoms on otoscopy examination
Kojima et al. [6]	48	Aged ranged: 2–62 years old 30 cases ≤15 years old 18 cases >15 years old	Hearing loss
El Bitar et al. [7]	35	children	Symptoms on otoscopy examination, hearing loss
Kojima et al. [9]	63	Aged ranged: 2–52 years old 46 cases ≤15 years old 17 cases >15 years old	Hearing loss
Okano et al. [10]	34	Aged ranged: 2–55 years old	Hearing loss
Friedberg [11]	78	Children and adults	Hearing loss
Sanna and Zini [12]	11	9 children 2 adults	Hearing loss
Takagi et al. [13]	71	Children and adults	Hearing loss
Yamatodani et al. [14]	26	Children	Hearing loss
Popli and Popli [20]	1	Child 6 years old	Otalgia, conductive deafness
Ye et al. [21]	9	Children and adults	Facial paralysis, hearing loss, tinnitus, vertigo

lamina cribrosa, though the posterior dural plate into retrolabyrinthin air cells posterior to the internal auditory canal, and into the lateral mastoid air cells [21].

Cholesteatoma of the petrous bone represents a complex challenge for the otologist both from a diagnostic and a therapeutic view. Its slow, silent growth, in particular in the congenital form, may cause in many cases a delay in its diagnosis, which is only clear when the damage is considerable and clinically involves structures important for quality of life (facial nerve, acoustic nerve). Sometimes it involves all the anatomical structures within the temporal bone with the risk of damaging structures vital for life (internal carotid artery, jugular vein, sigmoid sinus, dura).

Some characteristics of clinical presentation can supply important clues and confirmed diagnosis should be obtained according to these clues and suitable imaging studies before meningitis develops. Different surgical techniques should be adopted to treat the otorhinorrhea according to different leakage etiologies, and good results can be obtained. While posttraumatic leaks often heal spontaneously, the nontraumatic variant persist and surgical repair is mandatory to seal these CSF leakages. There are two main surgical approaches, the middle fossa craniotomy and the subtotal petrosectomy with removal of the bony labyrinth to reach petrous lesions [35]. In our case, though, an open non-radical antromastoidectomy was sufficient to remove the CC in total. A multilayered obliteration technique in which autologous and artificial materials are combined is considered as a modality with the highest rate of success.

Table 2 – Otogenic rhinorrhea – systematic review of literature.

Author	Number of otogenic rhinorrhea No. of cases
Shetty et al. [22]	3
Kronenberg et al. [23]	4
Sataloff et al. [24]	5
Konovalov and Blagoveshchenskaia [25]	3
Yi et al. [26]	7
Tyagi et al. [27]	4

5. Conclusions

The diagnosis of otogenic cerebrospinal fluid rhinorrhea is challenging and it can be easily misdiagnosed. Congenital cholesteatoma is a rare entity. We present a non-classical presentation of CC in an older man, with a previously unreported symptom of CSF rhinorrhea. Symptomatic improvement occured after surgical treatment of the disease.

Conflict of interest

The authors declare that they have no conflict of interest.

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None declared.

Ethics

The work described in this article has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans; Uniform Requirements for manuscripts submitted to Biomedical journals.

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