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Case report

An 11-year-old girl with recurrent bacterial meningitis due to liquorrhea caused by bone malformation of the skull base

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Abstract

The patient is a healthy 11-year-old girl with no history of trauma or hearing impairment. She developed pneumococcal meningitis three times, at ages 7, 10, and 11. Intracranial examination revealed, pore expansion and cerebrospinal fluid leakage in the right internal acoustic foramen, which were attributed to a bone malformation of the base of the skull. A procedure was performed to close the cerebrospinal fluid leakage; no relapse has been observed thus far. Previous case reports indicate that repetitive bacterial meningitis is often caused by internal ear malformation, trauma, tumors, or surgical operation. This case suggests the possibility that underlying disorders may not be apparent in cases of repetitive bacterial meningitis and, more proactive investigations are required to prevent further recurrence of meningitis.

Keywords: Recurrent bacterial meningitis; Liquorrhea; Congenital malformation; Child

1. Introduction

A national survey was conducted in 2008 to examine pediatric bacterial meningitis in Japan. This survey revealed the number of cases was 1.6 per 1000 annual pediatric hospitalizations and was approximately 1% in school-age children [1]. Drummond et al. [2] reported that only 1.3% of children with meningitis admitted to a large tertiary unit over an 11-year period had experienced bacterial meningitis on at least one previous occasion. In a literature review encompassing 363 cases of recurrent bacterial meningitis occurring between 1988 and 2007, Tebruegge [3] reported that 214 of these cases were related to anatomical defects, whereas 132 included immuno deficiencies [3]. Congenital inner ear malformations accounted for 15% of the cases with recurrent bacterial meningitis (55 of 363 cases) [3]. Other congenital anatomical causes have been reported, including heterotopic brain tissue, epidermoid cysts, and neural tube defects [4–6]. Here, we report a Japanese school-age girl who showed relapse of pneumococcal bacterial meningitis due to cerebrospinal fluid (CSF) leakage caused by bone malformation of the base of the skull.

2. Case report

An 11-year-old girl was admitted to Yamaguchi University Hospital to examine the cause of her recurrent bacterial meningitis. She developed *Streptococcus pneumoniae* bacterial meningitis and was treated at another hospital at ages 7 and 10. In two episodes, she

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experienced right middle ear inflammation. Since then, she has demonstrated normal neurological and intellectual development without hearing impairment or trauma. She did not receive vaccines for *Haemophilus influenzae* type b or *S. pneumoniae*.

When she was admitted to our hospital, her general condition was good. We observed no dermal sinus or middle ear inflammation, and her neurological examination was normal. A blood test for complement and immune globulin also showed normal findings. The initial CSF pressure was low at 0 mmHg. A head CT scan revealed cystic expansion at the right jugular vein hole, bone bulging in Meckel's cavity, and internal acoustic pore expansion (Fig. 1). Brain MRI revealed an abnormal signal in the same areas as CT, and in the mastoid cells (Fig. 2). Abnormal accumulation through the middle cranial fossa to the right parapharyngeal space was confirmed by radioisotope cisternography (Fig. 3a).

We detected expansions in porous structures, including the right Meckel's cavity, jugular foramen, and internal acoustic pore. This finding led us to assume that her condition was congenital, as she had no trauma history. We suspected CSF leakages were present in the right internal acoustic foramen to the middle ear and from the Eustachian tube to the nasal cavity. Thus, bacterial invasion into CSF through the permeable meninges was the source of infection. At the two remaining sites (inside the right Meckel's cavity and the right jugular foramen to the right jugular vein hole), the CSF was confined in the closed cavity and did not appear to communicate with regions outside the skull.

Her third episode of meningitis occurred when she was 11 years old. *S. pneumoniae* was detected in the CSF culture. Subsequently, she underwent an operation to close the CSF leakage. The petrous bone was drilled until the tympanic cavity was exposed, with careful preservation of facial canal. The tympanic cavity was packed by fat tissue taken from the abdominal wall in order for permanent discommunication to the nasal cavity. The fat was also packed in the right Meckel's cave, the mastoid air call and the extradural space made by the drilling. We did not observe the internal acoustic foramen. Continuous CSF drainage through a spinal subarachnoid catheter to reduce CSF pulse pressure was done for 10 days. We did not detect any CSF leakage on postoperative radioisotope cisternography (Fig. 3b). She has not experienced any further episodes of meningitis for more than 1.5 year after the operation. Severe conductive hearing impairment of the right ear was developed. Balance function and facial nerve function were intact.

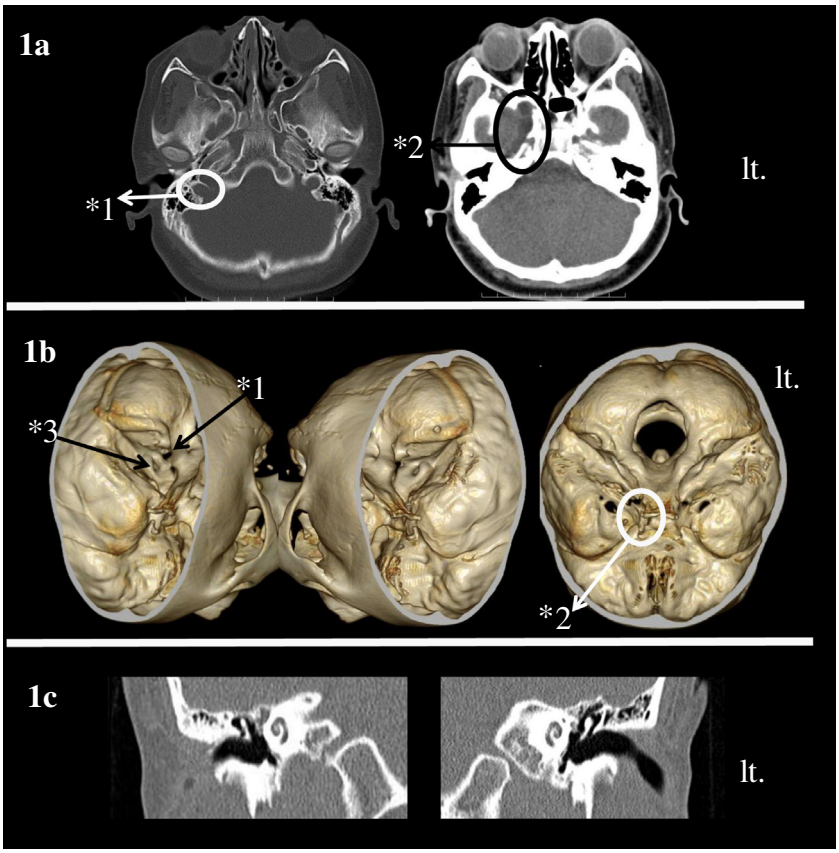

Fig. 1. Head and Ear CT. (1a) Head CT scan: Cystic expansion of the right jugular vein hole (∗1) and Meckel's cavity expansion was observed (∗2). (1b) Head 3D-CT: Expansions of the right jugular foramen (∗1), internal acoustic pore (∗3), and Meckel's cavity (∗2) were observed. (1c) Ear CT scan: No obvious internal ear malformation was observed.