Recurrent Bacterial Meningitis in a Child with Hearing Impairment, Mondini Dysplasia: A Case Report

Behdad Gharib¹, Sara Esmaeili², Golnaz Shariati¹, Narges Mazloomi Nobandegani¹, and Mehrzad Mehdizadeh²

¹ Department of Pediatrics, Children's Medical Center, Tehran University of Medical Sciences, Tehran, Iran  
² Department of Radiology, Children's Medical Center, Tehran University of Medical Sciences, Tehran, Iran

Received: 16 Mar. 2012; Received in revised form: 13 Jun. 2012; Accepted: 10 Dec. 2012

Abstract- Recurrent bacterial meningitis is not a common disease and makes physicians seek underlying predisposing factors which can result from anatomic anomalies or immunodeficiency. In this paper we present a boy with recurrent bacterial meningitis with the history of trauma and sensorineural hearing loss. Mondini dysplasia was demonstrated with computed homographic scans (CT-Scan) of temporal bones.

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Keywords: Hearing impairment; Mondini Dysplasia; Recurrent bacterial meningitis

Introduction

Recurrent bacterial meningitis is an uncommon disease of childhood (1). It is different from relapse or recrudescence of meningitis without full recovery period, which means incomplete resolution of signs, symptoms and laboratory findings (2,3). It can emerge from anatomic anomalies or immunodeficiency (1). Mondini dysplasia often remains undiagnosed and it should be concerned in children with hearing deficit and/or recurrent bacterial meningitis (2). Only radiological examinations can reveal the underlying predisposing factor (4).

Case Report

A 4.5 year-old boy was brought to the emergency department with the history of a two-day fever. He had also vomited the night before.

During examination, he looked ill with inflamed tonsils and no exudate, ear canals were packed with wax and tympanic membrane could not be examined, heart beats were regular with no murmurs. The abdomen was non-tender and there was no hepatic or splenic enlargement, auscultation of lung was normal. He had no rash, lymphadenopathy, diarrhea or cough.

When the emergency doctor tried to examine the meningeal signs, the patient flexed his neck easily several times as if he was familiar with this maneuver. He did not have any meningeal signs. The temperature was 39°C. Blood pressure was 105/70 mmHg, the pulse rate was 113 beats per minute, and the respiratory rate was 34 breaths per minute. He was born with congenital deafness, diagnosed when he was 6 months old.

His father showed a letter from a district hospital stating that the child had been hospitalized 6 times in the last 3 years because of meningitis and referred the patient for evaluating the patient for immune deficiencies as the cause of recurrent meningitis. The last hospitalization was one month ago where he was discharged with negative cerebrospinal fluid (CSF) culture and in good condition. He has never had any positive CSF or blood culture but obvious meningism signs, fever, vomiting and pleocytosis in CSF; he was admitted in the infectious disease unit and was administered ceftriaxone, vancomycin and antipyretic after initial blood test when lumbar puncture were performed.

The results of initial laboratory tests were as follows: WBC: 13220 per μl, neutrophil: 52.6%, lymphocyte: 26.9%, monocyte: 10%, eosinophil: 0.2%, basophil: 0.3%, RBC: 5.05 × 10⁶ per μl, Hb: 12.2 g/dl, MCV: 73.5 fl, Hct: 37.1%, MCH: 24.2 pg, Platelet count: 603 × 10³ per μl, ESR: 26 mm/h, blood glucose: 61 mg/dl, BUN: 9 mg/dl, Creatinine: 0.6 mg/dl, Ca²⁺: 9 mg/dl, Na⁺: 141 meq/l, K⁺: 4.5 meq/l.
Mondini dysplasia

Figure 1. CT cisternography and CT of petrous bone with sagittal reconstruction. Bilateral abnormal inner ear structures is visible with Mondini appearance. Note the presence of cisternal contrast in the right inner and middle ear in favor of associated Gaucher's ear that could be cause of recurrent meningitis.

A cerebrospinal fluid puncture and other laboratory test results were as follows: glucose: 20 mg/dl, protein: 240 mg/dl, direct smear for bacteria was negative for bacteria, white blood cells: 3200/μl (PMN: 85%, lymphocyte: 15%), RBC: 0, CSF culture: negative, C-reactive protein: +3. Urine analysis was unremarkable and urine culture was negative. Immunologic studies revealed normal results and were as follows: NBT: 100%, C3: 197 mg/dl (86-166), C4: 26 mg/dl (13-32), IgM: 320 mg/dl (43-196), IgA: 189 mg/dl (25-154), IgE: 3.3 mg/dl (1.07-68.9), IgG: 1077 mg/dl (643-1236).

In consultation with neurosurgery unit, a brain cisternography was obtained and it showed bilateral abnormality of inner ear structure with Mondini appearance (Figure 1). The presence of cisternal contrast in the right inner and middle ear was in favor of associated Gaucher's ear that could be due to recurrent meningitis, surgical repair and sealing in the right side. The patient had undergone a head trauma 2.5 years ago. His mother noted that since then he has had rhinorrhea from right nostril whenever he bends; however, the first episode of meningitis happened prior to the head trauma.

The patient was also examined through consultation with otolaryngology unit. It was revealed that the patient has had a cerebrospinal fluid otorrhea which presents itself as rhinorrhea, because of Gaucher's ear without any confirmed relation to the mentioned head trauma. The patient referred to otolaryngology unit and surgical procedure was performed.

Discussion

Having a patient with recurrent meningitis referred from a district medical center for being evaluated for immunodeficiency, mislead the low experienced doctors in our unit to approach the patient through laboratory tests to rule out immunodeficiency. Mondini dysplasia is a developmental anomaly of the middle ear, cochlear and vestibule and an abnormal connection between subarachnoid and the middle ear may result in recurrent bacterial meningitis and is commonly associated with hearing deficits (2). Bacterial meningitis is the most common problem which happens in 25% to 50% of patients with unrepaired fistulae and 70% of trauma patients in the first week following trauma and 10% of cases with the leak of CSF (4). Cerebrospinal fluid leakage may result from abnormal connection between middle ear and inner ear, which can be considered as route for recurrent meningitis (5). An important clue for assessment of ear-related recurrent meningitis is hearing deficit, which might be undiagnosed in early childhood (1).

In our case report, we faced a boy with the history of impaired hearing and recurrent bacterial meningitis and CT-scan revealed Mondini malformation (Figure 1). Mondini dysplasia is a kind of congenital malformation of the osseous labyrinth which can be diagnosed by radiographic examination (5). The ability of CT-scan in evaluation of temporal bone, makes it the best choice for examination (5) and it needs less than one minute immobilization (6). So, it should be considered from the beginning of evaluating the patient with recurrent meningitis. Although Mondini dysplasia is a sort of rare anomalies, but in the patients, with recurrent bacterial meningitis, must be included as a possible diagnosis.

References