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Original article

Rapidly progressive bilateral postmeningitic deafness in children: Diagnosis and management

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ABSTRACT

Objectives: To determine the diagnostic approach to severe or profound bilateral postmeningitic deafness and to propose management guidelines.
Material and methods: A retrospective review of five patients (two adolescents and three infants) with rapidly progressive severe bilateral deafness following an episode of meningitis managed between 2004 and 2010.
Results: The two adolescents presented *Neisseria meningitidis* meningitis and the three infants presented *Streptococcus pneumoniae* meningitis. Acquired bilateral deafness was diagnosed by audiometry an average of 68.8 days (range: 9–210) after the episode of meningitis. Behavioural audiological testing, adapted to age and state of health, was performed in all patients. Deafness was confirmed by Auditory Brainstem Response tests. All five patients were assessed by computed tomography (CT) and magnetic resonance imaging (MRI) within ten days. T2-weighted MRI sequences showed endolymph changes in four patients. CT scan demonstrated ossification in only one patient. Bilateral cochlear implant was performed in all patients, with complete electrode array insertion for eight implants and partial insertion for two implants (20 and 21 out of 22 electrodes inserted). Good results were obtained with cochlear implants in four cases.
Conclusions: Bilateral deafness can occur immediately or several months after bacterial meningitis, regardless of the micro-organism responsible, justifying screening by behavioural audiological testing adapted to age for two years following bacterial meningitis. Auditory Brainstem Response testing can confirm audiometric findings. When severe or profound bilateral deafness is observed, MRI must be performed urgently to detect endolymph inflammation or ossification. Early bilateral cochlear implantation is recommended in the presence of ossification.

1. Introduction

Bacterial meningitis is the most common cause of acquired hearing loss in children [1]. The risk of hearing loss has been estimated to be 22% in *Streptococcus pneumoniae* meningitis and 8% in *Neisseria meningitidis* meningitis [2]. According to data of the Epibac network, the infectious disease surveillance laboratory network, the incidence of acute bacterial meningitis in 2006 was 2.23/100,000 in metropolitan France with *S. pneumoniae* meningitis in 59% of cases and *N. meningitidis* meningitis in 25% of cases. Our centre is situated in Seine-Maritime, a department with an incidence of bacterial meningitis higher than the national average (3.26/100,000) which, according to some authors, could be related to an increased incidence of *N. meningitidis* meningitis [3].

Haemophilus influenzae vaccination has decreased the incidence of *Haemophilus influenzae* meningitis in France [4] and *S. pneumoniae* vaccination has decreased the incidence of severe *S. pneumoniae* infections in the United States [5]. However, a French epidemiological study failed to demonstrate any reduction of the number of cases of *S. pneumoniae* meningitis following *S. pneumoniae* vaccination in children [6]. In France, *S. pneumoniae* vaccination is recommended in children under the age of two years, but is not mandatory.

Bacterial meningitis can cause deafness due to peripheral or central hearing loss. Bacterial meningitis causes lesions via immune, inflammatory, and ischaemic reactions, or by cerebral oedema. Ossification of the organ of Corti is the most serious complication after bacterial meningitis. The organ of Corti can be damaged by inflammation with three successive stages: acute stage, fibrosis then ossification. Ossification after bacterial meningitis is reported in as many as 80% of cases [7]. The ossification process obliterates endolymph and perilymph spaces with more marked damage of the basal turn of the cochlea than the apex [8]. Ossification varies

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according to the time since onset of meningitis and its capacity for extension and can make cochlear implant electrode array placement difficult or even impossible. When ossification is observed after bacterial meningitis, there is therefore a race against time to establish the diagnosis of severe or profound bilateral deafness in order to rapidly propose a cochlear implant.

This retrospective study reviewed the medical charts of five children with postmeningitic deafness requiring cochlear implants during the year following the infection and proposes management when bilateral deafness is suspected following an episode of bacterial meningitis.

2. Material and methods

We retrospectively reviewed the charts of all paediatric patients in whom severe or bilateral profound deafness following bacterial meningitis was identified between 2004 and 2010. Relevant data of the clinical history, the diagnostic procedure, preoperative imaging data, time interval between diagnosis of bilateral deafness and cochlear implant, surgical findings with evaluation of fibrosis or ossification, depth of electrode array insertion, number of adjustments and their characteristics, and hearing performance were recorded for each case.

Hearing performance was evaluated on the basis of the last available results of pure-tone and speech audiometry for adolescents 1 and 2 and the APCEI scale for infants 3 to 5 [9]. The APCEI scale can be used to evaluate communication capacities according to 5 items summarized by the APCEI acronym word: Acceptance of the implant, Perception of language, Comprehension, oral Expression, and speech Intelligibility in children. Each item is scored from 0 to 5 with a maximum score of 25. The results depend on the child's age and performances with the implant.

3. Results

The study group, limited to five individuals, was composed of 2 adolescents aged 16 and 17 years and 3 infants under the age of 1 year (Table 1). The micro-organism responsible for meningitis was *N. meningitidis* (serogroup B) in the two adolescents, and *S. pneumoniae* in the three infants. Only one infant (patient 4) had been vaccinated against *S. pneumoniae* (Prévenar®).

Hearing loss was identified during hospitalisation for meningitis in three patients (patients 1, 2, and 4) with a brief time to diagnosis (Table 1).

The parents of patient 3 observed a change of behaviour on the child's return home after the episode of meningitis. The diagnosis of deafness was confirmed by audiometry and ABR 92 days after the onset of meningitis.

Patient 5 developed neurological disorders following meningitis, making behavioural audiometry difficult to perform. Audiometry was repeated by various operators in this patient: mild hearing loss, mainly affecting high frequencies, was diagnosed immediately after the episode of meningitis. These findings were confirmed by ABR. This patient's behavioural disorders subsequently deteriorated and profound bilateral deafness was diagnosed 210 days after meningitis by audiometry and ABR.

All patients were assessed by CT and MR imaging of the petrous temporal bones within 10 days after the diagnosis of bilateral deafness in order to detect signs of ossification or fibrosis of the cochlea (Figs. 1 and 2). When no signs of active ossification were detected, MRI was repeated 1 month later (patient 2). For patient 5, two early MRI examinations, performed during intensive care management of acute meningitis, did not demonstrate any endolymph changes. A CT scan performed 211 days after the onset of meningitis subsequently visualized left cochlear ossification. Radiological and hearing changes progressed over time in this patient.

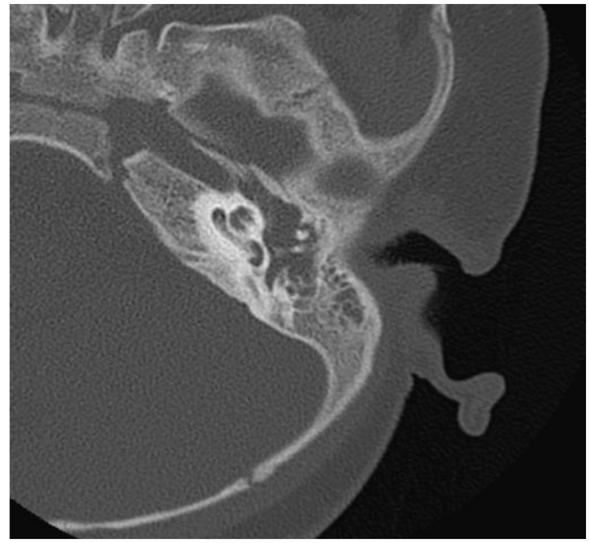

Fig. 1. CT scan, axial section of the left petrous temporal bone in patient 5: partial ossification of the basal turn of the cochlea.

All patients could be treated by cochlear implant. Patients 1 and 4 were operated very rapidly, as imaging demonstrated cochlear fibrosis and their general infectious and neurological state did not contraindicate early implantation.

Electrode array insertion was complete for 8 out of 10 implants and incomplete for the other 2 implants: 20 and 21 of the 22 electrodes were inserted, respectively. A classical electrode array cochlear implant and a straight electrode array cochlear implant were available for each procedure. Only patient 5 was implanted with a straight electrode array on one side and a classical electrode array on the other side.

In two out of four cases, the preoperative CT scan was normal, while MRI demonstrated endolymph changes. The surgeon encountered difficulties in all patients with abnormal MRI findings: absence of endolymph, cochleostomy difficult to perform due to early ossification or difficult electrode array insertion.

No unusual features in terms of the frequency and number of implant adjustments were observed for any of the five patients. Initial activation was performed sequentially with intervals

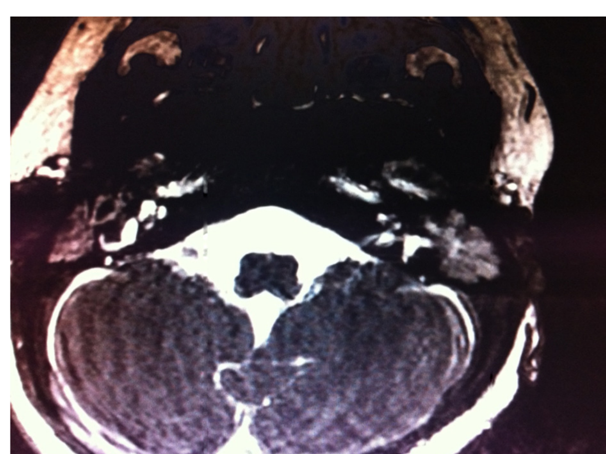

Fig. 2. MRI, T2-weighted sequence, axial section of the petrous temporal bones of patient 5: partial extinction of the T2 hyperintensity of the endolymph of the left cochlea suggesting cochlear ossification or fibrosis.