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Case report

Delayed cochlear implantation in post-meningitic deafness and hereditary complement C2 deficiency

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ABSTRACT

We report the case of an adolescent with post-verbal severe/profound sensorineural hearing loss, occurring as a consequence of two bouts of pneumococcal meningitis at 12 and 32 months of age. A possible immunodeficiency was investigated, revealing hereditary complement C2 deficiency (C2D). Given the insufficient benefit from high-power hearing aids, the boy received a cochlear implant (CI) at age 12. Despite the long interval of partial hearing deprivation and the post-meningitic etiology, improvement in open-set speech perception and quality of life were observed. The C2D did not favour post-operative infections nor meningitis recurrence. The risks and benefit of CI in this peculiar clinical circumstance are discussed after reviewing the literature.

1. Introduction

Lack of complement factors, such as hereditary C2 deficiency (C2D), affect the classical pathway of complement activation. It is associated with increased susceptibility to invasive infections by encapsulated bacteria, such as Streptococcus Pneumoniae and Haemophilus Influenzae type B [1]. In particular, the risk of meningitis with possible subsequent sensorineural hearing loss (SNHL) is increased [2–8].

The CI itself has been also associated with an increased risk of pneumococcal meningitis, mainly in patients with inner ear malformations [9]. However, the CI may be the only means to recover the auditory function in post-meningitic patients with severe to profound sensorineural hearing loss (SNHL). The peri-operative infective risks in case of C2D should be carefully balanced in each patient against the speech perception benefits offered by the CI.

We report the case of a boy affected by C2D for whom the CI was initially contraindicated for the increased risk of recurrent meningitis, and who successfully underwent it without complications, many years after the diagnosis of hearing loss.

2. Case report

The patient was born in 1997 with normal hearing. The first episode of pneumococcal meningitis occurred at 12 months and lasted 15 days. One month later, bilateral severe SNHL was identified by behavioural audiometry, absence of otoacoustic emissions and stapedial reflexes, and electrophysiological tests (auditory brainstem responses with clicks and tone bursts). The baby showed a hearing threshold of about 80 dB HL. Better and more reliable behavioural responses were obtained from the right ear. High-power hearing aids (HA) were immediately supplied and verbo-acoustic training was started.

Speech perception outcomes with the hearing aids were sufficiently satisfactory. The behavioural pure-tone threshold average (PTA) reached 50 dB HL and the gain in free-field with HA was 32.5 dB HL; the boy slowly developed language skills, although not comparable to his age peer. At 24 months, the Production of Infant Scale Evaluation [10], the Meaningful use of speech scale [11] and the Infant-toddler Meaningful Auditory Integration Scales [12] scores were 100% and MacArthur Communicative Development

List of abbreviations: C2D, hereditary C2 deficiency; CI, cochlear implant; HA, hearing aids; SNHL, sensorineural hearing loss; CSF, cerebrospinal fluid.

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Table 1
Results from basal serum immunological analyses at 3 years of age.

	Value	Normal range
Total IgG (mg/dL)	934	462–1710
IgG1 (mg/dL)	714	310–1050
IgG2 (mg/dL)	67	41–245
IgG3 (mg/dL)	26	9–69
IgA (mg/dL)	71	27–173
IgM (mg/dL)	161	62–257
Lymphocytes, $CD3^+$ (%)	71	56–86
Lymphocytes, $CD4^+$ (%)	54	32–64
Lymphocytes, $CD8^+$ (%)	20	13–37

Inventory [13] was at 75th percentile.

A second episode of pneumococcal meningo-encephalitis occurred at 32 months of age; a negative cerebrospinal fluid sample was obtained at an early stage of the disease (on the 5th day from onset), after prompt intravenous antibiotic treatment. No further clinical neurological deficits due to menigitis were observed.

A search for possible risk factors for recurrent meningitis was then implemented (Table 1).

Having excluded other etiopathogenetic causes of recurrent meningitis, a diagnosis of C2D was finally established at 3 years of age, by performing a total haemolytic activity assay (CH50) that was greatly reduced: 125 U/ml, vs. normal serum values of 1000 ± 200 U/ml.

Genetic tests showed homozygosity for a deletion of 28 base pairs in the C2 gene, located on chromosome 6 at 6p21.3, responsible for the C2D type I [14]. Given the clinical characteristics of the disease, adequate prophylaxes and vaccinations were performed.

Further audiological evaluation (on February 2000) showed a worsening of the bilateral severe SNHL, especially in the left ear; and the positive effects of the rehabilitation with HA started to weaken. Tympanograms were type A bilaterally, stapedial reflexes and otoacoustic emissions were absent. Auditory brainstem responses (ABR) could not be elicited at the highest levels of stimulation. The behavioural PTA at low and middle frequencies in free-field with HA was no better than 80 dB HL; the toddler did not react to loud sounds of higher frequencies. Word recognition scores with age-appropriate material in a closed set (pictorial aid) at signal to noise (SNR) +10 dB dropped to 25%.

A CI was offered at 3 years of age, but both the parents and the Surgeon were concerned about the risk of a new episode of meningitis. The parents asked for another consultation, where both the other Surgeon and Anesthesiologists denied the CI, based on a supposed increased peri-operative infectious complication risk. From an immunological viewpoint, the risk of meningitis after CI was considered too unpredictable on repeated visits at the age of 5 and 7 years at different institutions. HA use and oral rehabilitation were continued but, for the next 5 years, the family discontinued any medical control since they felt discouraged and hopeless.

When the boy was 12 years old, he was again referred to our institution by his Family Pediatrician for an audiological evaluation because his relational and communicative performances were inadequate for his age and the parents reported a worsening of school performances: he was unable to follow the lessons unless the teachers were facing him o writing; the performance scores on many topics (mathematics, physics, Italian and English language, history and geography), were insufficient. A severe lack of oral language production and multiple phonetic difficulties were assessed. He tended to discontinue the use of his hearing aids during the day, owing to subjective insufficient benefit. Actually his audiogram showed hearing residuals on the low frequencies

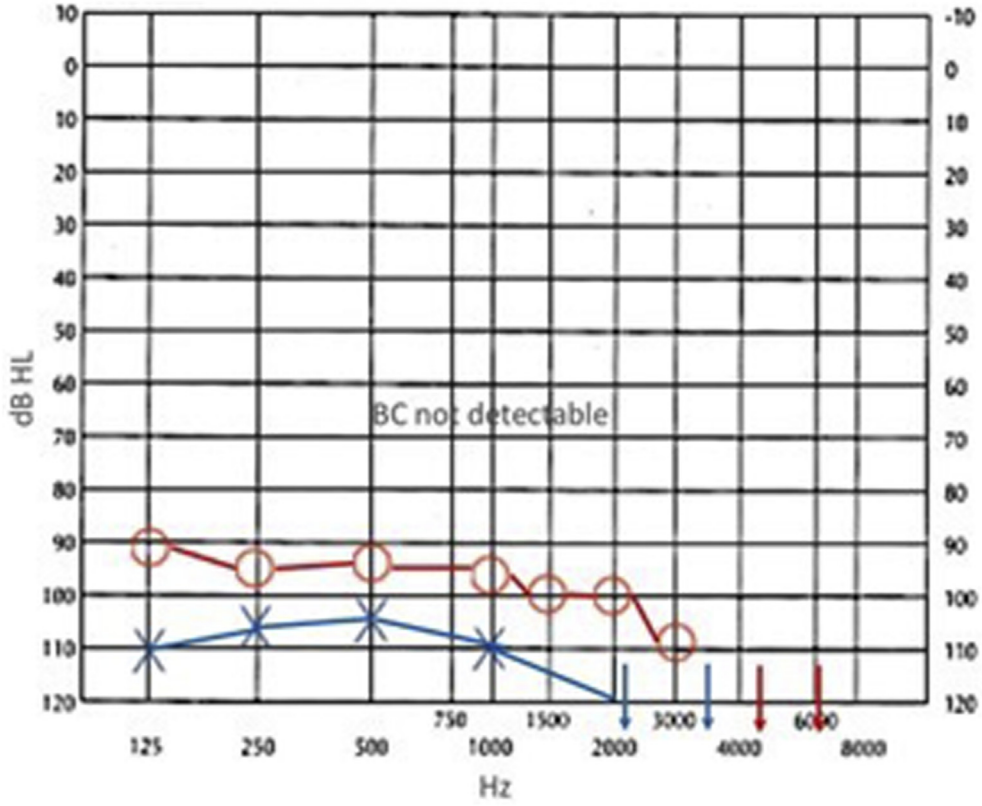

Fig. 1. Audiogram.