Diagnosis: Mondini dysplasia

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The CT scan of the right petrous bone shows a widening of the basal turn of the cochlea, vestibule and the anterior limb of the lateral semicircular canal (Figure 1A), a short internal auditory canal and a wide opening between the internal auditory canal and cochlea (Figure 1B). The MRI of the temporal bones (Figure 2A) shows a fluid filled right middle ear cavity and fluid in the cochlea, apparently connected at the oval window, and a well-demonstrated saccular deformity of the basal turn of the cochlea. The left inner ear is normal (Figure 2B). The radiological findings are consistent with the diagnosis of Mondini dysplasia with incomplete expression.

Discussion

Recurrent meningitis secondary to congenital inner ear anomaly is a rare clinical entity.¹ Certain congenital dysplasias of the inner ear provide a potential communication between the subarachnoid space and the middle ear cavity, which might result in cerebrospinal fluid leak manifesting as CSF otorrhea. This provides a portal of access for various organisms, both those that are known to cause meningitis and those that are unusual invaders of the CSF. This can result in recurrent meningitis if not corrected.²

Four major patterns of congenital inner ear malformation have been described: the Michel, Mondini, Scheibe and Alexander dysplasias. Mondini dysplasia, which was first described in 1791 by Carlo Mondini, is a developmental defect of the otic capsule, resulting from arrested development during the seventh week of fetal life.³ It is characterized by bony and membranous anomalies of the inner ear exhibiting a wide range of morphological and functional abnormalities. Typically the cochlea is short and flat with only 1 to 1.5 turns instead of the usual 2.5 to 2.75. The vestibule is large. The endolymphatic sac is bulbous with an immature auditory and vestibular end organ and nerves. Partial expression of the disorder may occur with little or no loss of auditory or vestibular function, while severe forms show no hearing or vestibular response.⁴ Deafness may first become manifest in childhood or early adult life.⁵ This malformation may occur in isolation or in association with malformations of the middle ear or other organs. It has been reported with syndromes such as Klippel-Feil, Pendred, DiGeorge, Wildervanck, Fountain, Johanson-Blizzard and Kabuki make-up syndrome. Also, it has been reported to occur with trisomies 13, 18 and 21. Intrauterine exposure to chemicals such as thalidomide or infectious agents such as rubella has also been implicated. Familial nonsyndromic cases have also been described.^{3,6,7} Patients with Mondini dysplasia are at increased risk of recurrent meningitis or perilymphatic fistula. The leak could be secondary to an enlarged cochlear aqueduct or an abnormal connection between the internal auditory canal and membranous labyrinth. The cerebrospinal fluid enters into the middle ear either through a trans-labyrinthine route, a defect in the stapes footplate near

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Figure 1a. CT scan of the right petrous bone (top left); **Figure 1b.** CT scan of the right petrous bone, (top right); **Figure 2a:** MRI of the temporal bones (bottom left); **Figure 2b:** MRI of the temporal bones (bottom right).

or via the oval window or less commonly the round window, leading to CSF otorrhea. The fistula is either spontaneous or as a result of head injury.⁸ In our patient a history of mild head trauma was reported two month prior to presentation, which may explain the timing of onset of his recurrent meningitis. The CSF leak was not sufficiently profuse to be noted clinically and CT and MRI failed to demonstrate the CSF leak. By surgically exploring the right middle ear, the presence of a cerebrospinal fluid leak around the oval window was confirmed. In Mondini dysplasia recurrent meningitis may be the initial presenting feature,

especially if the defect is unilateral and the hearing loss is unrecognized.

Management should be tailored to the specific clinical presentation. For patients presenting with meningitis, surgical exploration is recommended and various techniques have been used. Surgical management will depend on the functional status of the dysplastic ear. If functional hearing is preserved the leak can be patched with fascia and fibrin glue. When the ear is nonfunctional, the stapes may be removed and the vestibule packed with fascia, muscle or fat, and the oval window corked with bone.³ In the

asymptomatic patient, most otolaryngologists will maintain a watchful wait policy. Middle ear infections should be treated aggressively to minimize the risk of meningitis.⁸ Hearing rehabilitation may be achieved with conventional amplification. However, cochlear implantation with aural rehabilitation may augment acquisition of language and enhance education in pa-

tients with profound sensorineural hearing loss who did not benefit from conventional amplification.⁹

Recurrent meningitis or meningitis in a child with deafness should alert the clinician to the possibility of the presence of congenital anomaly of the petrous temporal bone and CSF fistula. In such cases complete otologic evaluation should be undertaken.

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