Congenital cerebrospinal fluid fistula with a Mondini type dysplasia. Report of a case

**Hector Aguirre Mariscal** MD,* **Norma Torres Macedo** MD,** **Enrique Segovia Forero** MD,*** **Yolanda Sevilla Delgado** MD,**** **Maria del Carmen Medrano Tinoco** MD

**Abstract**

Congenital malformations of the petrous portion of the temporal bone may produce a fistulous communication between the intracranial subarachnoid space and the perilymphatic spaces, which added to the alterations of the otic capsule, can produce a leaking of cerebrospinal fluid (CSF) into the middle ear. These malformations can facilitate the entrance of pathogenic microorganisms and produce recurrent meningitis. We present the case of a six year-old male with three bouts of meningoencephalitis of otic origin. He had a history of right ear CSF otorrhea, hearing loss and frontal headache for the last two years. Physical examination: Centrally perforated right tympanic membrane, draining clear liquid. A cytological analysis, auditive studies and a cisternotomography were performed which showed a congenital malformation of the inner ear compatible with a Mondini type dysplasia, as well as the presence CSF leak.

**Key words:** Dysplasia, Mondini, CSF leakage, meningoencephalitis.

There are four major patterns in congenital malformations of the ear. The Michel type aplasia is a severe malformation in which there is a total lack of development of the inner ear. Scheibe’s dysplasia is the most common malformation characterized by abnormal development of the membranous labyrinth. Alexander’s dysplasia is the least severe of all, it consists of cochlear duct aplasia affecting the basal turn of the cochlea. Mondini type dysplasia is characterized by incomplete development of the membranous and osseous labyrinth. It can be suspected in patients with the clinical triad of deep sensorineural uni or bilateral hypoacusia, cochleovestibular alterations and recurrent meningitis. It can be associated with CSF leakage. 2-4,8-9 (Figure 1)

**Case report**

A six year-old male with a history of three episodes of meningoencephalitis of otolitic origin was seen in the Pediatric Otorhinolaryngology Department at the Children’s Hospital of the XXI Century National Medical Center with a history of two years right CSF otorrhea, right hypoacusia.
and frontal cephalgia for ten months. Physical examination revealed a patient with stable vital signs, no fever and without meningeal involvement. There was a central perforation of the right tympanic membrane, through which there was abundant leakage of clear fluid. The rest of the examination was non-contributory. A cytochemical study of the fluid leaking from the ear indicated it was CSF. Audiological studies showed left normal audition and deep hypoacusia of sensorineural type in the right ear. A cisternotomy, showed structural changes of the inner ear with an altered cochlea: there was no evidence of turns; it appeared as an open cavity towards the internal auditory canal; at the vestibular level a cystic shaped common cavity was found. The presence of contrast material in the cavity of the right middle ear, which leaked from the posterior fossa through the internal auditory canal, crossing the internal ear and leaving through the oval window was found, consistent with evidence of a perilymphatic fistula. (Figures 2, 3)

A surgical occlusion of the fistula was performed. The patient had a successful course. There was no recurrence of the fistula and no episodes of meningocencephalitis in the last four years.

Figure 1. Mondini type dysplasia may be associated with CSF leak.

Figure 2. CAT. Contrast material in the right middle ear cavity.

Discussion

The original description of the Mondini type dysplasia made by Carlo Mondini in 1971, was of a cochlea with one and a half turns. The apical turn was absent and ended in a cavity which replaced the last turn of the cochlea. The oval and round windows, the semicircular canals, and their openings were normal. The vestibule was lengthened. The internal auditory canal and the cochlear duct were not described. The incomplete embryological development of the inner ear between the 6th and 8th weeks, results in the cochlea displaying only the basal turn and a dilated vestibule; the internal auditory duct opens into the labyrinth. This is associated with the risk of a spontaneous fistula of cerebrospinal fluid and episodes of recurrent meningitis, as in the case of a Mondini type dysplasia.

Some congenital malformations of the inner ear are characterized by a close association between the perilymphatic and the subarachnoid spaces. 1,5

There must be two abnormalities for this to take place:

1) At the middle portion of the labyrinth: the CSF can flow from the subarachnoid space within the perilymphatic space, through an internal auditory canal fistulous communication.

Figure 3. CAT. The cochlea appears as a cavity opening into the internal auditory canal.
2) At the lateral portion of the labyrinth: the CSF can reach the middle ear through a defect of the otic capsule. The most common location of an otic capsule defect is at the oval window, the Eustachian tube, the promontory, the Fallopian canal and the hypotympanum. This results in a communication between the subarachnoid space and the middle ear, thus facilitating recurrent meningitis.1,2,5,10

Embryologically, in the Mondini type dysplasia the development is arrested in the early fetal stages, between the 6th and 8th weeks, resulting in the labyrinth forebody forming an amorphous cavity or a primitive cochlea.3,4

Characteristically, the cochlea is shortened by one to one and a half turns instead of two and a half turns. Only the basal turn is fully developed; the superior turns form a common cavity. The modiolus and the spiral bony lamina are hypoplastic and the cochlear duct is dilated and shortened. Corti’s organ may be absent, mainly in the superior portion of the cochlea, and the number of cells in the spiral ganglion is diminished. The endolympathic duct, vestibule and semicircular canals may be lengthened, and in extreme cases, the vestibule has a cystic appearance.1,2,11

A Mondini type dysplasia should be suspected in children presenting with the following triad: deep bilateral or unilateral sensorineural hearing loss, cochleovestibular alterations, episodes of recurrent meningitis, and at times spontaneous CSF otorrhea.1,2,13

Diagnosis is confirmed with audiological and radiological studies. The comparative CT of both ears shows the structural alterations of the inner ear in the affected site. The small size and spiral nature of the organ makes it difficult to obtain imaging of the cochlea, even with the two-dimensional thin sections of the most advanced high-resolution computed tomography. However the natural plane for CT, namely axial or base projection, gives the best demonstration of the individual coils of the cochlea. Coronal sections formerly used with polytomography cut the coils obliquely and are not satisfactory, even though the central bony spiral is well shown as a small curl. The state of the membranous cochlea or the cochlea endorgan cannot be assessed by imaging at present. Magnetic resonance confirms that the cochlea is filled with fluid rather than with fibrous tissue. For all practical purposes, therefore, the radiological assessment is confined to the state of the bony cochlea.6,7

Early diagnosis and surgical treatment are essential to minimize the morbidity and mortality in these patients.

REFERENCES