

Informe de un caso interesante

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 MD¹

Resumen

Las malformaciones congénitas de la porción petrosa del hueso temporal pueden producir comunicaciones entre el espacio intracraneal subaracnoideo y el espacio perilinfático, lo cual aunado a alteraciones de la cápsula ótica, puede producir una fistula de líquido cefalorraquídeo (LCR) hacia el oído medio. Estas malformaciones pueden facilitar la entrada de microorganismos patógenos y producir meningitis recurrente. Se presenta el caso de un niño de seis años, con tres episodios de meningitis recurrente. El paciente tenía el antecedente de otorraquia derecha de dos años de evolución, pérdida audiológica y cefalea frontal. A la exploración física se encontró una perforación central de la membrana timpánica derecha por la cual salía líquido claro. Un análisis citoquímico del líquido, un estudio audiológico y una cisternotomografía, evidenciaron la presencia de una malformación congénita del oído interno compatible con una displasia tipo Mondini, así como una fistula de LCR.

Palabras clave: Displasia, Mondini, fistula de LCR, meningoencefalitis.

Introduction

Congenital malformations in the petrous portion of the temporal bone can produce fistulous communications between the intracranial subarachnoid space and the middle ear. These abnormalities can result in leakage of cerebrospinal fluid (CSF) with otorrhea, and invasion of pathogenic organisms provoking recurrent meningitis.¹

* Chief of the Otorrhinolaringology Service. Hospital de Pediatría Centro Médico Nacional Siglo XXI, I.M.S.S.

** Physician of the Otorrhinolaringology Service. Instituto Nacional de Pediatría

*** Otorrhinolaringologist. Hospital Ángeles del Pedregal

**** Physician of the Otorrhinolaringology Service. Hospital de Pediatría Centro Médico Nacional Siglo XXI, I.M.S.S.

¹ Chief of the Otorrhinolaringology Service. Instituto Nacional de Pediatría

Correspondence: Norma Torres Macedo MD. Servicio de Otorrinolaringología. Instituto Nacional de Pediatría. Insurgentes Sur 3700-C. Col. Insurgentes Cuicuilco. México 04530 D.F.
Recibido: agosto, 2003. Aceptado: octubre, 2003.

La versión completa de este artículo también está disponible en internet: www.revistasmedicasmexicanas.com.mx

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 meningo-encephalitis 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 cytochemical 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 otolithic 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 cephalaea 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 timpanic 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 cisternotomography, 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 meningoencephalitis in the last four years.

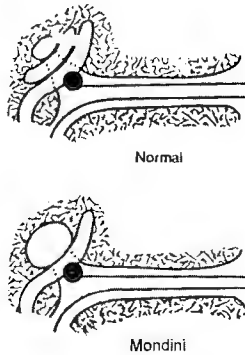


Figure 1. Mondini type dysplasia may be associated with CSF leak.

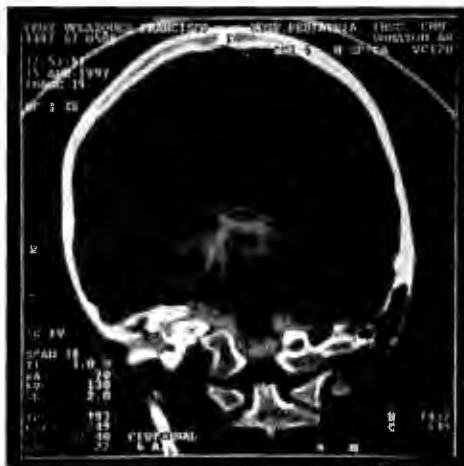


Figure 2. CAT. Contrast material in the right middle ear cavity.



Figure 3. CAT. The cochlea appears as a cavity opening into the internal auditory canal.

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 internal ear between the 6th and 8th weeks, results in the cochlea displaying only the basal turn and a dilated vestibule; the internal auditive 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.

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 forms a common cavity. The modiolo 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 endolymphatic 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.^{12,13}

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

1. Phelps DF, Props D, Sellars S, Evans J, Michaels L, Path F. Congenital cerebrospinal fluid fistula through the inner ear and meningitis. *J Laryngol Otol* June 1999;107:492-5.
2. Ohlms AL, Edwards M, Mason E, Igarashia M, Alford B, Smith R. Recurrent meningitis and Mondini dysplasia. *Arch Otolaryngol Head Neck Surg* 1990;116:608-12.
3. Shuknecht HF. Mondini dysplasia: a clinical and pathological study. *Ann Otol Rhinol Laryngol Suppl* 1980;89(Suppl 5):1-23.
4. Phelps DP. Ear dysplasia after Mondini. *J Laryngol Otol* June 1994;108:461-5.
5. Desjardins R, Guergverian AJ, Dube J, Dechamps N, Lavertu P. Meningitis and congenital fistula of the internal ear. *J Otolaryngol* 1982;11:97-100.
6. Phelps DP. The basal turn of the cochlea. *Brit J Radiol* 1992;65:370-4.
7. Dahm M, Seldon L, Pyman B, Laszing R, Lehnhardt E, Clark G. Three-dimensional reconstruction of the cochlea and temporal bone. *Adv Otorhinolaryngol* 1993;48:17-22.
8. Sennarogly L, Saatci I. A new classification for cochleovestibular malformations. *Laryngoscope* 2002;112(12):2230-41.
9. Komune S, Enatsu K, Morimitusu T. Recurrent meningitis due to spontaneous cerebrospinal fluid otorrhea. A case report. *Int J Pediatr Otorhinolaryngol* 1998;11(3):257-64.
10. Page E, Eby T. Meningitis after cochlear implantation in Mondini malformation. *Otolaryngol Head Neck Surg* 1997;116:104-6.
11. Phelps PD. Cochlear implants for congenital deformities. *J Laryngol Otol* Nov 1992;106:967-70.
12. Lovaib D, Francois M, Coderc E. Pneumococcal meningitis revealing dysplasia of the bony labyrinth in an infant. *Arch Pediatr* Mar 1996;3(3):254-7.
13. Elverland H, Mair L. Recurrent meningitis, congenital anacusis and Mondini anomaly. *Act Otolaryngol* 1983;95(1-2):147-51.