

Case Report

Utility of magnetic resonance cisternography with intrathecal gadolinium in detection of cerebrospinal fluid fistula associated with Mondini dysplasia in a patient with recurrent meningitis: Case report and literature review

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Abstract

Background: The intrathecal contrast-enhanced magnetic resonance cisternography (MRC) is a diagnostic method that has been proven effective in selected patients with various disorders of the cerebrospinal system, including the detection of cerebrospinal fluid (CSF) leaks. The Mondini dysplasia is a malformation of the inner ear characterized by an incomplete cochlear development. The cerebrospinal fistula associated with Mondini dysplasia usually occurs in the first 5–10 years.

Case Description: The case of a 34-year-old woman with CSF rhinorrhea and recurrent meningitis associated with CSF fistula into the right inner ear, which was detected by MRC with intrathecal gadolinium, is presented. The computed tomography (CT) cisternography failed to detect the exact location of the leak. The right Mondini dysplasia was identified on CT of the temporal bone. A subtotal right-sided petrosectomy and fistula closure into the bony labyrinth were performed. After the procedure the patient no longer presented meningitis or CSF leak.

Conclusions: The radiological identification of the site of CSF leak through sensitive imaging studies such as MRC with intrathecal gadolinium is crucial for surgical approach.

Key Words: Intrathecal gadolinium, Mondini dysplasia, magnetic resonance cisternography, recurrent meningitis

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INTRODUCTION

Cisternography with intrathecal gadolinium is defined as the magnetic resonance imaging (MRI) of the cerebrospinal system and associated structures after intrathecal administration of contrast material into the subarachnoid space.^[2] It is a technique that has been shown to be effective in selected cases of various disorders of the cerebrospinal system.^[7] It allows evaluation of dynamic cerebrospinal fluid (CSF) flow,

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so that, unlike the cisternography without contrast, it provides not only morphological but also physiological information.^[3,4] Several studies have reported the safety of intrathecal injection of gadolinium; in a recent series that included 95 patients who underwent contrast-enhanced magnetic resonance cisternography (CE-MRC), no major complications were reported, and headache was the most commonly reported minor periprocedural complication.^[27,33]

In CE-MRC, visualization of leakage in T1 of hyperintense contrast images with fat suppression is sufficient for the diagnosis of CSF leaks.^[6,30]

The precise preoperative leakage site identification is crucial for surgical approach and increases the success rate of surgery while reducing the risk of complications. The primary reason for the repair of CSF leaks is that the overall risk of meningitis in patients with persistent CSF rhinorrhea is 19%.^[13]

Mondini dysplasia is a congenital malformation of the inner ear most commonly associated with CSF fistula in the temporal bone in children.^[31] In fact, this anomaly was described in 1971 by Carlo Mondini after dissecting the inner ear of an 8-year-old deaf child and it is the result of the discontinuation of the development of the labyrinth during the 7th week of fetal life.^[21] Typically, the cochlear capsule is flattened with underdevelopment of the bone structure in the apical portion of the cochlea, and this reduces the number of cochlear turns. The cerebrospinal fistula associated with Mondini dysplasia usually occurs in the first 5–10 years. However, in this case the inner ear fistula associated with Mondini dysplasia, detected by CE-MRC and temporal bone computed tomography (CT), was identified in a 34-year-old woman.

CASE REPORT

A 34-year-old female, right handed, with a 2-year history of “intermittent right unilateral hyaline rhinorrhea,” was admitted to our hospital with headaches, vomiting, and consciousness deterioration. A diagnosis of acute meningitis caused by *Streptococcus pneumoniae* was made, and specific antibiotic treatment was started with excellent results. After the total recovery a CT cisternography (CTC) was performed and the study concluded that the leak came from the cribriform plate to the right ethmoid sinus. An endoscopic exploration of the nasal cavity with fluorescein, in an attempt to closure the leak, revealed no defect in the ethmoidal roof, but leakage from the right Eustachian tube was detected. In that scenario, a second review of CTC suspected a defect at the floor of the middle fossa coming into the oval window. So, a right temporal craniotomy was performed with detachment of some adhesions of dura to the floor

with cribriform-like structures and was covered with temporal fascia. However, the patient continued with CSF rhinorrhea and intermittent retronasal discharge as well as symptoms of intracranial hypotension, so it was decided to expand the diagnostic approach.

A CE-MRC was done. The technique has been well described in the literature.^[1,5,8] It basically consists of the acquisition of isotropic T1-weighted, FS spin-echo images of the brain. The patient was then transferred to the recovery room. A lumbar puncture was performed and 0.5 ml of gadopentetate dimeglumine (Magnevist; Bayer Schering Pharma, Berlin, Germany) mixed with 2 ml of saline were injected into the subarachnoid space. The patient was left in the prone position for 2 h and then new sequences were repeated with the protocol used before.

The study showed that the contrast completely filled up the inner ear and went into the middle ear and Eustachian tube; in subsequent acquisitions, it was possible to observe a path that connected to the inner ear with the middle ear located near the oval window [Figure 1].

After detecting the CSF leak from the inner ear, a CT scan of the temporal bones was performed and the typical Mondini deformity of the right cochlear apex was identified, formed by cystic enlargement of the vestibular aqueduct [Figure 2].

A right subtotal petrosectomy was performed detecting the fistula in the right bony labyrinth, which was closed with fat and fascia from the temporal region. There have been no new episodes of meningitis or CSF leak up-to-date.

DISCUSSION

Currently, the most widely accepted method for evaluation of patients with suspected CSF leak is the combination of high-resolution CT with contrast cisternography. However, though CT images of thin sections are useful in visualizing bone defects, they may be limited in patients with multiple fractures or

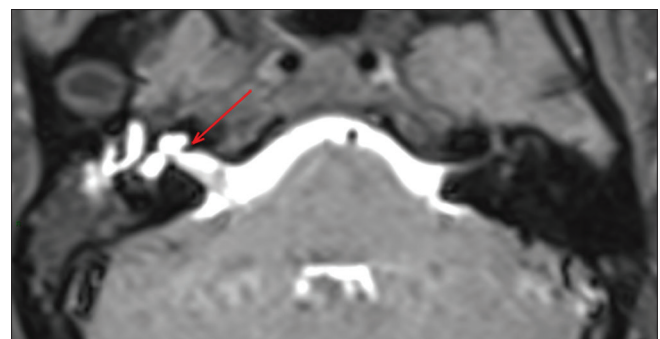


Figure 1: CE-MRC Axial view. Fistula in the Right Bony Labyrinth (Arrow)

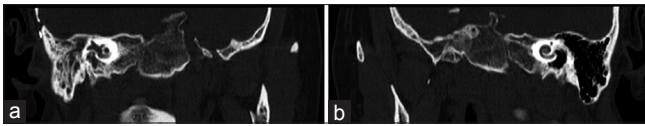


Figure 2: CT. Temporal bone, coronal view. (a) Right Temporal bone with Mondini malformation. (b) Left temporal bone with no abnormalities

intermittent CSF leak.^[5] Additionally, the bone defect may be congenital, and leakage may come from another location. The ability of CT to demonstrate the leak site depends on indirect findings such as lines of fracture at the base of the skull, bone defects, mucosal abnormalities, presence of fluid in the sinuses, pneumocephalus, and meningoencephalocele.^[26,30] The multislice CT has a sensitivity and specificity of 84–95% and 57–100%, respectively for detection of leak sites.^[5] The sensitivity of radionuclide cisternography scan for diagnosis of CSF leak is about 40%. The routine use of CT, alone or in conjunction with CTC or radionuclide cisternography, significantly increases radiation exposure.^[5,26,30] Both techniques are limited in detecting low-flow fistulas or “hair-like” communications.^[23]

On the contrary, with nonenhanced MRC, the leak is characterized by a hyperintense path between the subarachnoid space and sinu-nasal space.^[15] In addition, secondary changes in the brain parenchyma and associated encephalocele suggestive of CSF leak are not visible.^[5] However, these findings are not specific; sinusitis-mastoiditis, viscous secretions, and secondary artifacts of bone–air interface can condition increased rates of false-positive diagnoses (42%).^[23] Even when some authors have suggested that the combination of high-resolution CT and nonenhanced MRC with T2-weighted sequences is accurate in 96%,^[24] both tests are also limited in showing hair-like defects and intermittent or inactive leaks.

The CE-MRC with intrathecal gadolinium is a minimally invasive procedure compared to other cisternographic and ventriculographic tests (radionuclide or MRI ventriculography) with the advantages of using MRI images, and, unlike the cisternography without contrast, provides not only morphological but also physiological information.^[3,4] Postcontrast images must be obtained at the second hour after intrathecal administration of gadolinium.^[5] Additional postcontrast image acquisition is recommended if the anterior or medial cranial fossa do not show contrast. Valsalva maneuver is useful just before acquiring the postcontrast images to locate the leak site.^[7]

Although intrathecal injection of gadolinium-DTPA (gadopentetate dimeglumine) is not approved by the Food and Drug Administration, preliminary clinical studies have reported the success of this method in locating CSF fistulas for patients with rhinorrhea.^[10,17]

Intrathecal gadolinium-enhanced MRC was first performed in laboratory animals in 1985 by Di Chiro *et al.*^[14] A pilot study with 11 adult patients showed the relative safety and feasibility of low-dose intrathecal gadolinium administration, with a dose 30–50 times smaller than doses causing toxic changes in laboratory animals. No patient manifested gross behavioral changes, neurologic alterations, or seizure activity.^[32]

Although “gadolinium encephalopathy” has been reported in patients with renal impairment,^[9] the neurotoxic effect does not present at doses lower than 3.3 $\mu\text{mol/g}$ brain parenchyma^[22] and usually a 0.5 ml dose, equivalent to a dose of 0.17 $\mu\text{mol/g}$ brain parenchyma, is used. In rats, intracisternal gadolinium produces myoclonus, ataxia, and tremor at concentrations of 2.5–15 mol/g of brain.^[19]

Also gadolinium-DTPA is the safest and most recommended contrast in human studies.^[2-5,23] In a multicenter study that included 95 patients who underwent CE-MRC, all patients were hospitalized for an observation period of 24 h postprocedure and followed up with neurologic examinations for 6–12 months; afterward, no major complications were reported, and headache was the most reported minor periprocedural complication.^[27]

In a study conducted in 2008 by Aydin *et al.*, a sensitivity of 84% for locating CSF leaks was reported for CE-MRC after evaluating 44 patients with rhinorrhea, 43 of whom results were confirmed during surgery. No patient had adverse reactions related to the study (only headache which resolved with analgesic use in 24% of patients) or neurological symptoms during an average follow-up of 4 years.^[11] In the same way, Arbeláez *et al.* (2007) identified a CSF fistula in 22 of 24 patients with clinically suspected nasoethmoidal CSF fistulas, by showing leakage of contrast medium from the anterior cranial fossa into the ethmoidal, frontal, or sphenoidal air cells, through CE-MRC. The site of leakage was confirmed surgically in 14 of these patients and was repaired with dural grafting. Mild-to-moderate (in most instances mild) and self-limited postprocedural headache was found in eight patients.^[8]

The CE-MRC also allows to identify spinal leaks in patients with spontaneous intracranial hypotension, as shown by Vanopdenbosch *et al.*, in a prospective observational cohort study, who found a spinal epidural leak mainly at the cervicodorsal junction in 9 of 14 patients with spontaneous intracranial hypotension,^[29] besides the identification of cranial CSF leak in six of eight patients with liquorrhea and three of five patients with recurrent bacterial meningitis.

In cases of patients with CSF fistula outside the skull, Chazen *et al.* found a leak at CE-MRC in 9 of 24 patients (38%) but in only 3 of 24 (13%) patients with CTC ($P = 0.011$). CT myelography did not

identify any leaks that were not found on CE-MRC.^[12] These are similar to findings in a recent study by Akbar *et al.*,^[1] which detected a leak on MR myelography in 5 of 24 patients (21%) with negative CTC findings. No adverse reactions were observed.

In the case of our patient, MRC with intrathecal contrast helped identify the origin of the CSF fistula without any adverse events associated with the study, and this helped, in turn, through a detailed analysis of the anatomy of the inner ear via a high-resolution scan of the temporal bone, in the detection of the Mondini dysplasia associated fistula.

Mondini deformity refers to the absence of apical modiolus and interscalar septum, resulting in an incomplete segmentation of the cochlea and enlarged vestibular aqueduct and dilated vestibule.^[21] It means that true Mondini dysplasia has a cochlea with normal basal turn and an interscalar septum deficient at the distal and medial turn.^[28] In cases of Mondini dysplasia, the cochlea characteristically has 1–1.5 turns less than the 2.5–2.75 turns in normal individuals. This may be associated with hypoplastic modiolus, dilated vestibule, and spacious semicircular canals. The auditory and vestibular nerves may be immature or normal, and this is attributed to the different clinical degrees of auditory damage or vestibular dysfunction.^[20] Also, these deformities may result in a connection between the subarachnoid space and the ear, which may in turn cause recurrent episodes of meningitis. The most common route of CSF leak in cochlear dysplasia is a modiolus fistula connecting to the subarachnoid space via the internal auditory canal and the membranous labyrinth.^[16] The CSF enters the middle ear via a translabyrinthine route through a defect in the vicinity of the stapes footplate or via the oval window. The fistula may be spontaneous or a result of head trauma.^[20]

The fistula associated with Mondini dysplasia usually presents with rhinorrhea if the tympanic membrane is intact, as was the case of our patient.

In our patient, despite having a congenital malformation of the inner ear, Mondini dysplasia, curiously, the meningoencephalic infections did not begin until 10 years of age. The literature reports that about two-thirds of cases with congenital fistulas begin with these recurrent infections before 18 months of age.^[18]

Managing Mondini dysplasia depends on the clinical presentation. In the case of a fistula tract, after locating your site, closure of the defect as soon as possible is indicated. Adipose tissue was used in our case, to fill the vestibule which was reinforced with a temporal fascia implant, and the middle ear was obliterated with abdominal adipose tissue. Although it is reported that recurrence after surgical repair occurs in 30–60% of the

patients,^[25] our patient has not presented meningitis or CSF leak up-to-date after 3 years of follow up.

CONCLUSION

The radiological identification of the site of CSF leak through sensitive imaging studies such as MRC with intrathecal gadolinium is crucial for surgical approach. Also, the successful repair of CSF leaks is important to prevent damage and repeated episodes of meningitis. The Mondini dysplasia should be included in the differential diagnosis of a patient with recurrent bacterial meningitis.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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