Cerebrospinal Fluid Otorhinorrhea in Patients with Defects through the Lamina Cribrosa of the Internal Auditory Canal

Prashant G. Shetty, Manu M. Shroff, Milind V. Kirtane, and Swati S. Karmarkar

Summary: We describe three patients with bilateral cerebrospinal fluid (CSF) otorhinorrhea with unilateral progressive hearing loss in whom CT showed the defect to be located in the lamina cribrosa of the internal auditory canal. CT cisternography showed the CSF fistula in two of the three patients who had Mondini malformation, whereas the CSF fistula was obvious on the plain high-resolution temporal bone CT study in the third patient, who had a posttraumatic (nonsurgical) fracture of the lamina cribrosa. Fast spin-echo T2-weighted coronal MR cisternography also showed the site of leakage in the third patient. In the presence of an intact tympanic membrane, the CSF egressed to the nose via the eustachian tube in all three patients.

Index terms: Cerebrospinal fluid, leakage; Temporal bone, abnormalities and anomalies

Computed tomographic (CT) cisternography is useful for detecting the site and extent of bone defects in patients with cerebrospinal fluid (CSF) rhinorrhea (1). The most common site of a CSF fistula is the floor of the anterior cranial fossa, predominantly in the cribriform plate and/or the fovea ethmoidalis (roof of the ethmoidal sinus). Rarely, the defect is located in the roof of the sphenoidal sinus or on the tegmen tympani in the temporal bone. We describe two patients who had Mondini malformation in whom CT cisternography showed a CSF fistula at the lamina cribrosa (lateral wall of the internal auditory canal) and a third patient in whom plain high-resolution CT of the temporal bone and magnetic resonance (MR) cisternography suggested a definitive leak through the lamina cribrosa consequent to a nonsurgical fracture. In all three patients the CSF leaked through both nostrils in the presence of an intact tympanic membrane.

Materials and Methods

For the CT cisternography, 8 mL of nonionic iodinated contrast material (180 mg I/mL) was injected intrathecally via a lumbar puncture in two patients (cases 1 and 2). In all three patients, 3-mm coronal CT scans were obtained through the anterior cranial fossa, the roof of the sphenoidal sinus, and the temporal bones with the patient in the prone position; 1.5-mm contiguous coronal sections were obtained through the region of interest for greater bone detail. A bone algorithm was used for the same purpose. In one patient, coronal fast spin-echo T2-weighted MR imaging was performed, with parameters of 6000/90/1 (repetition time/echo time/excitations), a section thickness of 3 mm, a distance factor of 0.1, a field of view of 230 mm, and a matrix of 192 × 256.

Case Reports

Case 1

A 45-year-old man had recurring headaches, neck stiffness, intermittent leakage of clear fluid through both nostrils, and progressive, left-sided hearing loss. He had no history of surgery. Results of a CSF analysis were in keeping with the diagnosis of pyogenic meningitis.

A CT cisternographic study, which had been performed at an outside institution, showed a complete absence of the left lamina cribrosa. Contrast material was seen in the middle ear cavity, having leaked through a defect in the lamina cribrosa and from there to an enlarged vestibule through the oval window (Fig 1). The cochlea was represented by a saclike diverticulum from the vestibule without apical turns.

At surgery, a defect was identified at the posterior aspect of the oval window, with disruption of the footplate of the stapes. CSF was seen to be exiting from this defect. Since the patient had a complete sensorineural hearing loss on the left side, documented by audiography, and the internal auditory canal was in direct communication with the vestibule, the latter was filled with fat and covered with

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From the Departments of Imaging (P.G.S., M.M.S., S.S.K.) and Otorhinology (M.V.K.), PD Hinduja National Hospital, Bombay, India.

Address reprint requests to Prashant G. Shetty, MD, Department of Imaging, PD Hinduja National Hospital, Veer Savarkar Marg, Mahim, Bombay 400 016, India.

temporalis fascia and the middle ear cavity was filled with Gelfoam to plug the leak. The patient has not reported for follow-up since the surgery.

Case 2

A 40-year-old man had intermittent leakage of clear fluid through both nostrils and profound right-sided hearing loss since childhood. During childhood, he had undergone a right-sided mastoidectomy for chronic purulent discharge of the right ear. He also had had unsuccessful surgery for a presumed defect in the floor of the anterior cranial fossa 2 years before the present admission. CT cisternography was performed, in which the temporal bone was studied in the absence of any demonstrable leak through the anterior cranial fossa and the roof of the sphenoidal sinus. A definite dehiscence was seen at the right lamina cribrosa, and leaked contrast material was visible within an enlarged vestibulocochlear complex (Fig 2A and B). The lateral semicircular canal was also enlarged. Contrast material was also identified within the right middle ear cavity and in the hypopharynx on the right side. At surgery, a 2 × 1-mm bone chip was seen in the right middle ear cavity, embedded within the right oval window, where the CSF was leaking out.

Case 3

A 10-year-old boy had had bilateral CSF rhinorrhea, more pronounced on the right, and complete right-sided hearing loss for about 3 months after a fall. A CT cisternographic study performed elsewhere did not reveal a CSF fistula on the floor of the anterior cranial fossa, on the roof of the sphenoidal sinus, or on the tegmen tympani. On nasal endoscopic examination, CSF was seen to egress from the right eustachian tube. Hence, a plain high-resolution coronal CT study of the temporal bones was performed on the basis of our experience with the previous two cases. Fig 3A shows a definite linear fracture through the right lamina cribrosa and the roof of the cochlear promontory. MR cisternography based on the coronal T2-weighted fast spin-echo sequence clearly shows a hyperintense column communicating between the basal CSF cistern and the middle ear cavity (Fig 3B). The right-sided mastoid air cells were also filled with fluid.

At surgery, the right-sided lamina cribrosa fracture was seen to extend up to the cochlear promontory, where CSF was seen to be leaking through the round window into the middle ear cavity. After filling the vestibule and cochlea with fat and covering it with temporalis fascia, the middle ear cavity was filled with absorbable gelatin sponge (Gelfoam) to plug the leak. There was no evidence of CSF leakage at follow-up 2 years later.

Discussion

CT cisternography is useful for detecting the site and extent of CSF fistulas (1). A CSF fistula is usually seen on the floor of the anterior cranial fossa, chiefly at the cribiform plate and the fovea ethmoidalis. Other sites include the roof of the sphenoidal sinus and the tegmen tympani (the roof of the middle ear cavity). The most common causes of CSF otorhinorrhea are closed head injury and surgery.

CSF otorhinorrhea can be spontaneous or traumatic (2). Spontaneous cases account for 4% of CSF leaks (1, 3), which have been de-
scribed through the Mondini malformation and through thin areas of the tegmen and in the wall of the middle cranial fossa into the sphenoidal sinus. Tumors and infections can cause skull base erosions and secondary CSF leaks.

In patients with CSF otorhinorrhea it is important to locate the CSF fistula and surgically plug it, as it can be the source of recurrent pyogenic meningitis (3), which has been reported in 9% to 50% of cases; however, current investigators have put this figure at 4% (2, 3). Surgical repair of the dura is required to prevent this complication and to plug the CSF leak. In all three of our patients we found the source of the CSF fistula to be the lamina cribrosa of the internal auditory canal. The lamina cribrosa is not a common site for spontaneous or posttraumatic (nonsurgical) CSF fistula; such a location has been associated with Mondini deformity, in which there is bony and dural dehiscence along with cochlear and vestibular abnormalities (4). The internal auditory canal can also be a source of CSF leakage after surgery for vestibular schwannoma (5). Routes of postoperative CSF leakage into the middle ear cavity have been reported through the lamina cribrosa, though the posterior dural plate into retrolabyrinthine air cells posterior to the internal auditory canal, and into the lateral mastoid air cells (6).

The lateral wall of the internal auditory canal is a thin bony plate known as the lamina cribrosa, through which passes the nerve fibers of the seventh and eighth cranial nerves and blood vessels. The lamina cribrosa has a medial bony projection called the falciform crest. It has a dural and arachnoid covering medially and forms a barrier between the inner ear and the subarachnoid space (7). The vestibule is located immediately lateral to the lamina cribrosa.

In the absence of a demonstrable leak in the floor of the anterior cranial fossa, in the roof of the sphenoidal sinus, or in the tegmen tympani, it is important to examine the lamina cribrosa for a CSF fistula. This is particularly so when a patient has bilateral CSF rhinorrhea and ipsilateral progressive hearing loss, as in our series.

Mondini malformations with dilatation of the cochlear sac and undersegmentation of the cochlea are known to present with perilymphatic fistula as a result of abnormal communication of CSF spaces and the labyrinth via the fundus of the internal auditory canal. They may be found in association with Klippel-Feil, Pendred, and DiGeorge syndromes, or they may occur as an isolated abnormality (8), as in cases 1 and 2 in this series. Familial occurrence has been reported. The malformation may be unilateral or bilateral, and may present as a congenital hearing loss, which is usually progressive through childhood. It may also occur in association with multiple episodes of meningitis.

Schuknecht (8) suggested that the lamina cribrosa of the internal auditory canal is the most likely site of CSF leakage. Other potential sites of CSF leakage that are rarely found in Mondini malformation are the cochlear aqueduct, the petromastoid canal, and the facial canal (9).

Cases 1 and 2 are examples of Mondini malformation with dehiscence of the dura and the lamina cribrosa. The vestibulocochlear complex was dilated in both the cases. The basal turn of the cochlea itself appeared dilated along with the lateral semicircular canals. The enlarged vestibulocochlear complex and the lat-
eral semicircular canals may be dysplastic as part of the congenital structural abnormalities, possibly augmented by chronic CSF pulsations extending through the dehiscent lamina cribrosa.

Although CT cisternography is the best way to document a CSF fistula in patients with CSF otorhinorrhea and unilateral hearing loss, plain coronal high-resolution CT through the temporal bone may be useful to exclude the possibility of a defect in the lamina cribrosa. High-resolution CT offers superb bone detail and can give indirect evidence of a CSF fistula, such as a bone fracture or defect. Additional findings that can help identify a CSF leak on plain high-resolution CT scans include opacification of the middle ear cavity with fluid, the presence of a fracture involving the vestibulocochlear complex in cases of trauma, and congenital enlargement of the vestibulocochlear complex. Claims for the accuracy of fistula localization on plain high-resolution CT scans range from 22% to 100%, with the greatest accuracy reported in the anterior cranial fossa (3, 10). A noninvasive method for confirming this finding is a fast spin-echo T2-weighted MR sequence through the region (11), as in case 3 in our series. CT cisternography, however, is the standard of reference and should be used until alternative noninvasive methods of diagnosing CSF fistula have been proved in larger series.

Retrospectively, it was thought that plain high-resolution CT study of the temporal bones coupled with coronal MR cisternography of this region with the use of a fast spin-echo T2-weighted sequence would have noninvasively shown the site of the CSF fistula in the first two patients. Fluid in the middle ear cavity and mastoid air cells can also be seen in patients with inflammatory disease, which can be confused with CSF otorhinorrhea. However, a history of CSF leakage together with a bone defect at the lamina cribrosa seen on high-resolution CT scans should easily differentiate these entities. The three patients in this series had CSF otorhinorrhea in the presence of an intact tympanic membrane. Obviously, CSF otorhinorrhea can also be seen in the presence of a perforated tympanic membrane. MR cisternography has been reported to be highly sensitive in detecting inactive leaks from other sites (11).

To conclude, lamina cribrosa is a rare site of spontaneous or posttraumatic (nonsurgical) CSF fistula. Patients may present with bilateral CSF otorhinorrhea and progressive hearing loss, and the temporal bone should be suspected as the source of leakage in the absence of a CSF fistula at the usual known sites. Although CT cisternography shows the CSF fistula definitively, a plain high-resolution CT coronal temporal bone study with MR cisternography may show the defect and the leak noninvasively, particularly in patients with bilateral CSF otorhinorrhea associated with unilateral hearing loss. Ancillary findings include the presence of fluid in the middle ear cavity and mastoid air cells as well as deformity or fracture of the vestibulocochlear bony labyrinth. A history of temporal bone surgery should suggest the possibility of a CSF fistula at this site.

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