Cerebrospinal fluid otorhinorrhea in a child: Case Report
Magdy Samir, Hossam Rabie
Department of ENT, Head & Neck Surgery, Ain Shams University, Cairo, Egypt

Abstract
Leakage of cerebrospinal fluid (CSF) through the ear is a rare but potentially life-threatening situation that requires rapid intervention. Even though a leakage of CSF is occurring through otologic structures, actual leakage of the fluid from the ear is not always present, but the fluid flows down the Eustachian tube and manifests as a clear rhinorrhea. The presence of an abnormal communication of the sterile subarachnoid space with the flora of the sinonasal tract places the patient at great risk for meningitis. Indeed, an episode of meningitis may be the presenting problem for a person with an otologic CSF leak. Diagnosis of such cases needs special attention, high index of suspicion, careful examination and relevant investigations to find the site of leakage.

This report presents a thinking flow chart for management of a child presented with recurrent attacks of meningitis and clear fluid leaking from the nose on crawling in which all investigations revealed intact anterior skull base.

Key words: CSF Rhinorrhea, Otorrhea, Congenital CSF leak, recurrent meningitis.

Introduction:
Leakage of cerebrospinal fluid (CSF) through the ear structures is a rare occurrence (1). It is more common in children than adults, and represents the greatest challenge to the clinician because the source of the leakage is not readily obvious (2).

The chief presenting symptom one would expect when a CSF leakage through the ear is present is a clear watery drainage from the ear. This, however, is not always the case and does not occur unless the eardrum or canal is in some way violated. If this is not the case, the leakage may be evident as a clear watery nasal discharge or as recurrent meningitis. This discharge may be positional or intermittent in nature and may only become apparent during straining or leaning forward (3).

Case presentation
Twenty months old female child was referred from pediatrics department with recurrent attacks of meningitis usually following upper respiratory tract infections. Mother noted hearing loss since birth. Recently, her mother reported clear fluid leaking from the right side of the nose on crawling. Hearing tests revealed bilateral profound hearing loss. Fluid sample was collected and chemical analysis for b2 transferrin was positive, consistent with CSF. Computed tomography of the paranasal sinuses (CT PNS) was normal.

Figure 1: CT Temporal bone: Bilateral Mondini anomaly, opacity of right ME space

Because of this normal CT PNS, hearing loss, and relevant history of CSF leak, we were directed for radiological assessment of the ear. CT scan of the temporal bone revealed bilateral Mondini dysplasia, opacification of right middle ear by fluid, and bilateral wide internal auditory canals (figure 1). MRI showed fluid in middle ear and mastoid in T2 imaging (figure 2). Right middle ear exploration was done through a trans-canal approach. Leakage was found coming through the stapes footplate. On attempt of platinectomy, a perilymph gusher was encountered. A time was allowed for the gusher to end, then a good sealing was achieved by an hour glass fat graft, fibrin-glue and fascia overlay grafting. The patient was followed for two months postoperatively with no recurrence of leak or meningitis.
Discussion
Spontaneous CSF otorhinorrhea is usually due to congenital defects in the otic capsule. Mondini deformity often present with a patent lateral aspect of the internal auditory canal, allowing direct movement of CSF into the inner ear. A defect at the annular ring of the stapes footplate may also result in drainage of CSF into the middle ear. Similar presentation may be observed in a patient with a widely patent cochlear or vestibular aqueduct, abnormal patency of the petromastoid canal or patent Hyrtl fissure (4). Meningitis is often the presenting symptom. Hearing loss is another presentation, which is typically sensorineural, suggesting the associated abnormality of the inner ear (5).

Localization of an otogenic spinal fluid leakage is usually accomplished using imaging studies, a high-resolution CT scan with axial and coronal sections. Unless an otologic source is certain, the scan should cover all 3 cranial fossae. Localization of leakage site with CT scan may be enhanced with the use of intrathecal contrast (1). MRI is a critical adjunct when a defect is found in the bony plate of the tegmen or the posterior fossa to evaluate the presence of meningoencephalocele (6).

Spontaneous leaks in children with otic capsule defects, such as Mondini deformity, can often be repaired by a transcanal approach, as the site of leakage is commonly through the footplate. However, if the exact site of the leakage is not known or suggested on imaging studies in most cases of spontaneous leakage, a transmastoid approach is preferred. In rare cases, the exact site of leakage is not found, and diffuse leak is observed from multiple mastoid air-cell tracts. In this situation, the mastoid may need to be obliterated with fat. Obliteration of the middle ear and Eustachian tube may also be required, especially if the leakage is not limited to the mastoid (5). In even big defects in the floor of the middle fossa, the problem is best addressed with a combined middle fossa/transmastoid approach (7). Continuous lumbar spinal fluid drainage is an important adjunct to surgical repair of otogenic CSF leakage (8).

Congenital CSF leaks usually presents in different ways, and need thorough clinical assessment. Complete otologic and neurologic examination as well as audiologic evaluation is a mandate. To define the site of leak, imaging of the three fossae is essential. Surgical treatment is usually the gold standard management, but rehabilitation of other associated sensory or anatomical defects should not be overlooked.

References: