Case 16439



Ruptured congenital temporal bone meningocele through facial nerve canal with CSF otorrhoea

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Section: Head & neck imaging

Area of Interest: Ear / Nose / Throat Head and neck

Neuroradiology brain Imaging Technique: CT Imaging Technique: MR

Imaging Technique: MR-Functional imaging

Case Type: Clinical Cases

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Patient: 12 years, male

Clinical History:

A 12-year-old male patient attended the otorhinolaryngology outpatient clinic complaining of continuous left ear discharge. The patient reported a past history of recurrent attacks of bacterial meningitis. Clinical examination revealed normal auricle and no clinical facial nerve palsy. There was no past history of trauma, surgery or chronic medical problems.

Imaging Findings:

Computed tomography (CT) images in axial and coronal planes show abnormal bulbous enlargement of the left geniculate ganglion with a dilated labyrinthine segment of the facial nerve canal and osseous dehiscence of the bone that separates the geniculate ganglion from the middle ear cavity (Fig. 1 and 2).

Magnetic resonance imaging (MRI) examination using heavily T2-weighted in axial, coronal and sagittal oblique planes and contrast enhanced T1-weighted images in axial and sagittal sequences. MRI images show a dilated CSF-filled meningeal sac at the left geniculate ganglia region and dilated labyrinthine segment of the facial nerve canal (Fig. 3 and 4). There is downward displacement of cerebellar vermis by 11 mm below the basion opsithion line (Fig. 4), smooth diffuse pachymeningeal enhancement and distention of dural venous sinuses denoting intracranial hypotension.

Discussion:

Background

Cerebrospinal fluid (CSF) fistula means egression of CSF through an osteodural defect resulting in an abnormal communication between the subarachnoid space and a pneumatised structure in the skull base. CSF leakage from the ear is in the category of perilymphatic fistula. Perilymphatic fistulae may be congenital or acquired. Traumatic, non-traumatic, and spontaneous are the causes of acquired perilymphatic fistula [1].

Congenital temporal bone CSF fistulae are rare in occurrence and could be perilabyrinthine or translabyrinthine in origin depending on the route of CSF transmission. There are five potential routes of these fistulae: through the tegmen tympani, a large apical air cell, Hyrtl's fissure, the petromastoid canal and the facial nerve canal [2]. The rare perilabyrinthine group arises from bony defects close to but not including the labyrinth and usually has normal hearing initially. The commoner translabyrinthine group is nearly always accompanied by anacusis, severe labyrinthine dysplasia and a route which communicates between perilymph and the middle ear cavity as through the oval window [3].

Clinical Perspective

In patients with recurrent meningits, a careful search for abnormal pathway connecting CSF and middle ear cavityis a must.

Imaging Perspective

Bulbous enlargement of geniculate ganglion region on CT examination guiding us to ask for MRI examination, which confirms the nature of this dilatation to be CSF connecting to the internal auditory canal with normal morphological appearance of the cochlea, vestibule and semicircular canals. These imaging findings go with the diagnosis of perilabyrinthine meningocele through the facial nerve canal.

Outcome

Regarding the management of perilymphatic fistulae, surgery is avoided in those with only hearing loss as the symptoms are unilateral [4]. There have been cases of translabyrinthine fistulae repaired by filling the internal auditory meatus (IAM) with the muscle to stop CSF flowing during stapedectomy [5]. Surgical intervention for serious complication as CSF otorrhoea can be done with surgical repair of the leak with a subtotal petrosectomy or middle fossa craniotomy [5, 6]. In our case report, the patient's parent preferred a conservative watchful waiting line of treatment.

Teaching Points

The presence of continuous ear discharge with recurrent attacks of meningitis should direct attention to the possible risk of CSF fistula, which requires imaging by CT and MRI for determination of the site of origin of this CSF fistula. Early intervention protects the patient from the life-threatening bacterial meningitis.

Patient consent

Written informed patient consent for publication has been obtained.

Differential Diagnosis List: Ruptured perilabyrinthine congenital temporal bone meningocele, Eroding cholesteatoma, Geniculate ganglion schwannoma, X-linked stapes gusher

Final Diagnosis: Ruptured perilabyrinthine congenital temporal bone meningocele

References:

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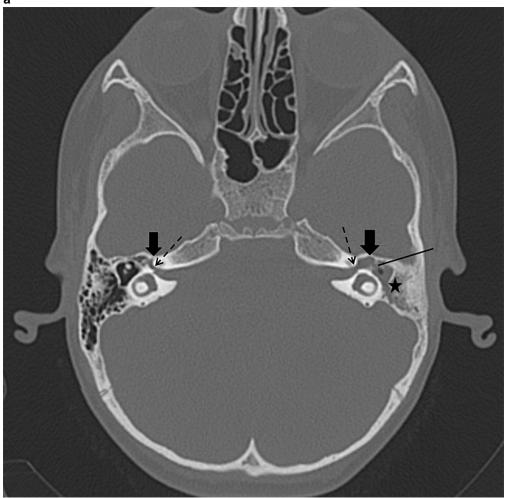
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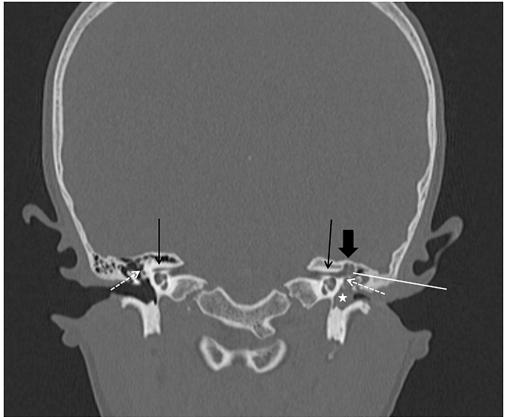


Description: Axial CT image of the temporal bone shows distended labyrinthine segment of the facial nerve canal on the left side in comparison to the right side (dashed arrows), bulbous dilatation of left geniculate ganglion in comparison to normal right one (thick black arrows), focal dehiscence in the wall separating geniculate ganglion from the middle ear cavity (black line) and opacification of the left middle ear cavity as well as mastoid air cells (black star). **Origin:** Department of Radiology, Zagazig university hospital, El Sharkia, Egypt, 2018

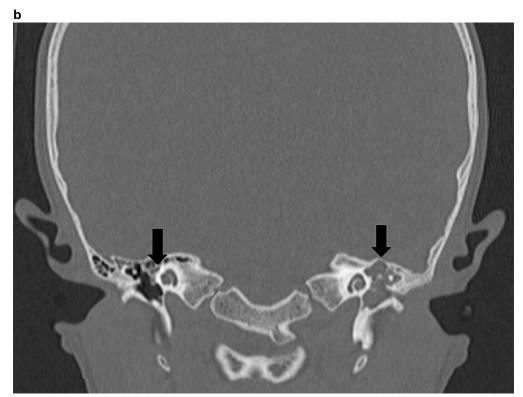


Description: Axial CT image of the temporal bone shows normal-appearing tympanic segment of facial nerve canals on both sides (back arrows). **Origin:** Department of Radiology, Zagazig university hospital, El Sharkia, Egypt, 2018

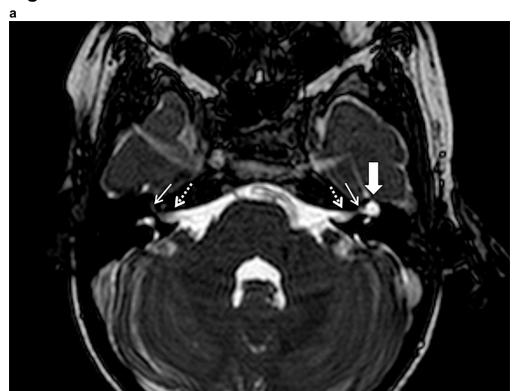
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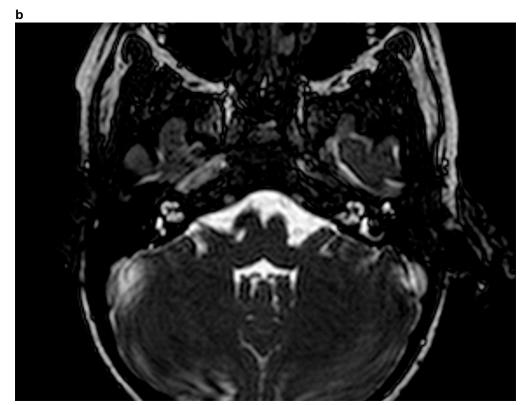
Description: Coronal CT image of temporal bone reveals a slightly distended labyrinthine segment of the facial nerve canal on the left side (thin black arrows), dilated left geniculate ganglion (thick black arrow) with focal dehiscence of the related lateral bony wall (white line), normal-appearing tympanic segments on both sides (dashed white arrows) and opacified middle ear cavity (white star). **Origin:** Department of Radiology, Zagazig university hospital, El Sharkia, Egypt, 2018



Description: Coronal CT image of temporal bone reveals bulbous dilatation of geniculate ganglion region on the left side (thick black arrows). The intact roof of the left geniculate ganglion region separating it from the middle cranial fossa. **Origin:** Department of Radiology, Zagazig university hospital, El Sharkia, Egypt, 2018

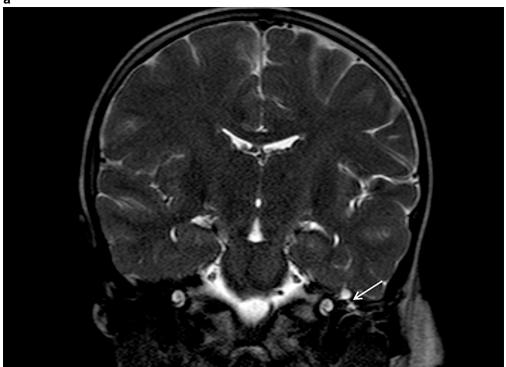


Description: Axial VISTA sequence shows a dilated labyrinthine segment of the left facial nerve canal in comparison to the normal right side (thin white arrows), an average calibre of the internal auditory canal (dotted arrows) and CSF-like signal intensity meningocele at the site of the left geniculate ganglion (thick white arrow). **Origin:** Department of Radiology, Zagazig university hospital, El Sharkia, Egypt, 2018



Description: Axial VISTA sequence shows normal cochlear turns and signal void internal bony modiolus on both sides. **Origin:** Department of Radiology, Zagazig university hospital, El Sharkia, Egypt, 2018

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Description: Coronal VISTA sequence shows left-sided CSF-filled meningocele superior and lateral to the normal-appearing cochlea and fluid is seen tracking from meningocele and communicating with the middle ear cavity (white arrow). **Origin:** Department of Radiology, Zagazig university hospital, El Sharkia, Egypt, 2018



Description: Sagittal contrast-enhanced T1WI reveals abnormal downward descend of cerebellar tonsil below basion opsithion line by 11 mm (white line). **Origin:** Department of Radiology, Zagazig university hospital, El Sharkia, Egypt, 2018