Meningoencephalocele presenting as a mass at the external auditory meatus

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Endaural meningoencephalocele or brain herniation into the middle ear and mastoid is an entity that is usually encountered in relation to chronic otitis media (COM) and multiple ear surgery.[1] Only 139 cases have been reported in the literature in 40 years up to1989,[1] and some more in recent years.[2-4] The pathology frequently presents with a mass that is localized to the middle ear and mastoid. Extension to the external ear is very rare and has been reported in few cases only.[2,4] We report a case of a patient with an endaural meningoencephalocele that presented as a mass visible at the external auditory meatus. The report aims to highlight some of the features related to external ear meningoencephalocele and to remind clinicians to include meningoencephalocele in the differential diagnosis of external auditory canal lesions.

A 22-year-old male presented with an 18 months history of a painless mass in his right ear. The patient had a longstanding history of COM for which he had mastoidectomy elsewhere 2 years ago. He also gave a history of a few self-limiting episodes of watery discharge from the right ear but no bleeding or meningitis. On examination, the patient was deaf in his right ear and a mass was visible at the external auditory meatus. Computed tomography (CT) showed evidence of bone erosion in the tegmen tympani. Magnetic resonance imaging (MRI) (**Figure 1**), showed a mass that was partly solid partly cystic directly extending through the tegmen defect. The solid part was non-enhancing, isointense, in continuity with the temporal lobe and interrupting the dural line. The cystic part was containing CSF and reaching the external auditory meatus. The mass was therefore diagnosed to be a meningoencephalocele. The lesion was managed by a right middle fossa craniotomy. An intradural approach was preferred as it allowed a better visualization of the prolapsed brain and the bony defect. The encephalocele part of the prolapse was excised and the petrous bone defect, which measured 2-2 cm, was exposed. The membranous part, the meningocele, which was located in the external canal, was excised via the external meatus. The defect was then repaired by covering it with a piece of bone fashioned to size, fascia lata and tissue glue. An external ventricular CSF drainage was also set up and maintained for 3 days. The patient made a good postoperative recovery and was discharged from hospital 7 days later. Outpatient assessment 4 weeks after surgery revealed no evidence of the mass and the external auditory canal was lined with granulation tissue.

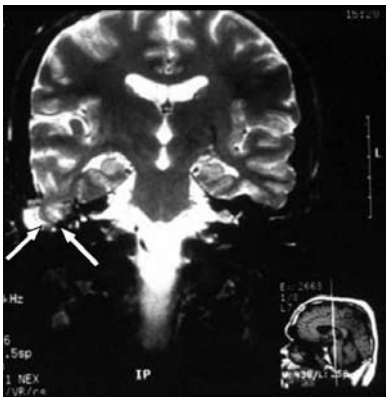

Figure 1 - Coronal MRI, T2-weighted image showing the petrous defect and the endaural meningoencephalocele (double arrows).

Follow up MRI at 3 months postoperatively showed no evidence of the meningoencephalocele. The patient remained well at 2 years follow up.

Endaural meningoencephalocele is usually encountered in relation to cholesteatoma, temporal bone trauma, iatrogenic injuries, congenital cranial base defects, neoplasia, irradiation and idiopathic causes.[3] Their occurrence as a consequence of COM is reported to be as low as 1% and as high as 26.4%.[3,5] Such discrepancy is related to the reporting of a high percentage of neglected COM cases in some series and the inclusion of pure dural dehiscence without a meningoencephalocele, which is a more frequent finding, in others.[4,5] In addition, the surgical procedure rate in reported COM series also varies. The latter is a relevant finding, considering that 59-77% of meningoencephaloceles are due to iatrogenic injury and are found in revision surgery.[5] However, the chronic suppurative disease itself can cause the bony erosion and the dural injury that lead to the development of the meningoencephalocele in COM.[5] Once the dura is injured in the presence of a tegmen defect, the arachnoid herniates first through the defect into the middle ear cavity, which can be associated with CSF leakage. Secondarily, the temporal lobe herniates through the arachnoid and dural defect obstructing the CSF leak.[4] The extent of the arachnoid and brain herniation that determines the size of the meningoencephalocele is related not only to the size of the bony and dural defects but also to the middle ear anatomy, the CSF hydrodynamics and the local intracranial pressure. The latter can be elevated as a result of local edema in relation to a regional cerebritis or due to an abortive attempt at brain abscess formation.[4] In addition, localized arachnoiditis may increase CSF flow towards the defect contributing to the herniation. In the presence of a perforated tympanic membrane an expanding meningoencephalocele is likely to follow the path of low resistance to the external auditory canal presenting as a mass at the external auditory meatus.

On reviewing recent middle ear meningoencephalocele articles,[1-5] it can be deduced that the presentation of the meningoencephalocele as a mass in the external auditory canal is extremely rare as it was limited to 8 cases only.[2,4] It is not even clear if these lesions were visible at the external auditory meatus, as in our case. In addition, it can be observed that middle ear meningoencephalocele is a diagnosis that usually found incidentally or detected on the follow up CT scan imaging.[3] The lesion is usually clinically silent beneath the more common chronic middle ear disease signs and symptoms such as hearing loss, otorrhea, tinnitus, vertigo, perforated tympanic membrane and residual cholesteatoma.[2,3] Rarely, they can be associated with significant neurological complications such as facial weakness, seizures, brain abscess, meningitis[2] and CSF leak.[4] Combined imaging with CT and MRI facilitates the diagnosis of endaural meningoencephalocele. Computed tomography provides details of the bony anatomy that will alert the surgeon to the tegmen defect and the soft tissue going through it. MRI provides improved soft tissue resolution over CT and allows the surgeon to differentiate between recurrent disease in the mastoid and brain herniation.[3] Therefore as a general principle clinicians should suspect a brain herniation in all patients with bony defect of the tegmen and a soft mass in the mastoid in continuity with the brain tissue. Under such circumstances an MRI is imperative in order to diagnose the brain herniation. Repair of an endaural meningoencephalocele is usually accomplished either from above by way of the middle cranial fossa approach or from below by way of the transmastoid approach using autologous fascia, cartilage, or bone.[3,4] Small defects (<1 cm) can be repaired from below while large defects, like our case, should be managed by a craniotomy and a middle fossa approach. However, if there is evidence of residual cholesteatoma or the brain hernia is very large and difficult to remove completely from above then a combined transmastoid-middle cranial fossa approach should be used.[4] The standard principles of chronic ear surgery emphasizing a safe, dry ear over hearing restoration should be adhered during the management of the patients.[3] According to a number of series of endaural meningoencephalocele that were reported in the literature, the rate of using craniotomy for defect repair was 26%,[5] 42%,[3] 61%,[2] 100%,[4] a reflection of the significant variation in the size of the defects in the various studies. The outcome of patients with an endaural meningoencephalocele managed in the setting of revision chronic ear surgery usually correlates with the outcome of extensive chronic ear disease in general. The repair procedure itself is usually associated with low complication rate and no reported recurrence rate.[2-4] Our patient had presented with a middle ear meningencephalocele that had expanded to reach the external auditory meatus. We believe that the latter occurred because of certain local features such as; large tegmen defect, large dural dehiscence most likely iatrogenic, ruptured tympanic membrane and alteration in the local CSF hydrodynamics that might have been related to localized arachnoiditis. We conclude that meningencephalocele should be included in the differential diagnosis of a lesion in the external auditory canal and that the pathology is likely to be associated an increase the risk of meningitis and CSF leak due to exposure indicating the need for an expedited repair.

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