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Case Report

A rare finding of bilateral facial canal meningoceles involving the tympanic segment in suspected idiopathic intracranial hypertension ☆

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ABSTRACT

Meningoceles are a common radiological feature found in cases of idiopathic intracranial hypertension (IIH). Rarely, they can affect the facial canal within the petrous temporal bone, leading to symptoms such as facial nerve palsy, hearing loss or meningitis. This is the first case report that describes bilateral facial canal meningoceles involving the tympanic segment of the canal. Prominent Meckel's caves were also seen on MRI, a feature commonly associated with IIH.

Introduction

Meningoceles are defined as protrusions of the lining of the brain and spinal cord tissue that can occur congenitally, spontaneously, or via iatrogenic means [1]. They are also a common radiological feature that can be seen in cases of idiopathic intracranial pressure (IIH), where increased cerebrospinal fluid (CSF) pressure can lead to meningeal herniations through bony defects in the base of the skull [2–5]. We report the first documented case of bilateral facial canal meningoceles involving the tympanic segment of the nerve in a patient with radiological features of IIH.

Case report

A 72-year-old man presented to a tertiary Otolaryngology clinic for investigation of long standing right sided hearing loss and tinnitus. He denied any history of otorrhea, headache, or facial nerve palsy; his past medical history included morbid obesity, as well as a range of cardiac and respiratory comorbidities. Examination, including facial nerve function and otoscopy, was unremarkable. Audiometry demonstrated a right sided moderate to profound mixed hearing loss and a left high frequency sensorineural hearing loss (SNHL) with normal (type A) tympanometry bilaterally.

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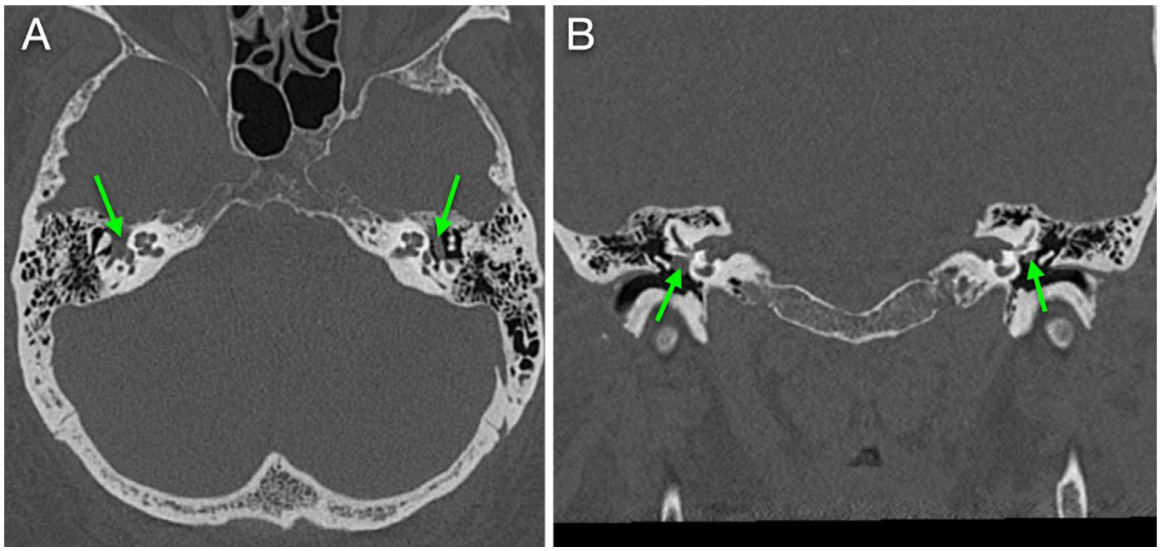

Fig. 1 – High resolution noncontrast computed tomography: (A) axial view of an abnormal soft tissue or fluid density involving the geniculate ganglia and (B) coronal view of the tympanic segment of the facial canal being involved.

Initial high resolution noncontrast Computed Tomography (CT) of the petrous temporal bones demonstrated abnormal fluid or soft tissue density filling the geniculate ganglia (Fig. 1A) and the tympanic segment of the facial canal with evidence of impingement on the middle ear ossicles on the right side (Fig. 1B)

Subsequent Magnetic Resonance Imaging (MRI) with contrast of the petrous temporal bone revealed bilateral T2 hyperintense signal expansion of the facial canal involving the labyrinthine, geniculate and proximal tympanic segments (Fig. 2A and B); these areas were nonenhancing with no evidence of diffusion restriction. Axial T2 views also

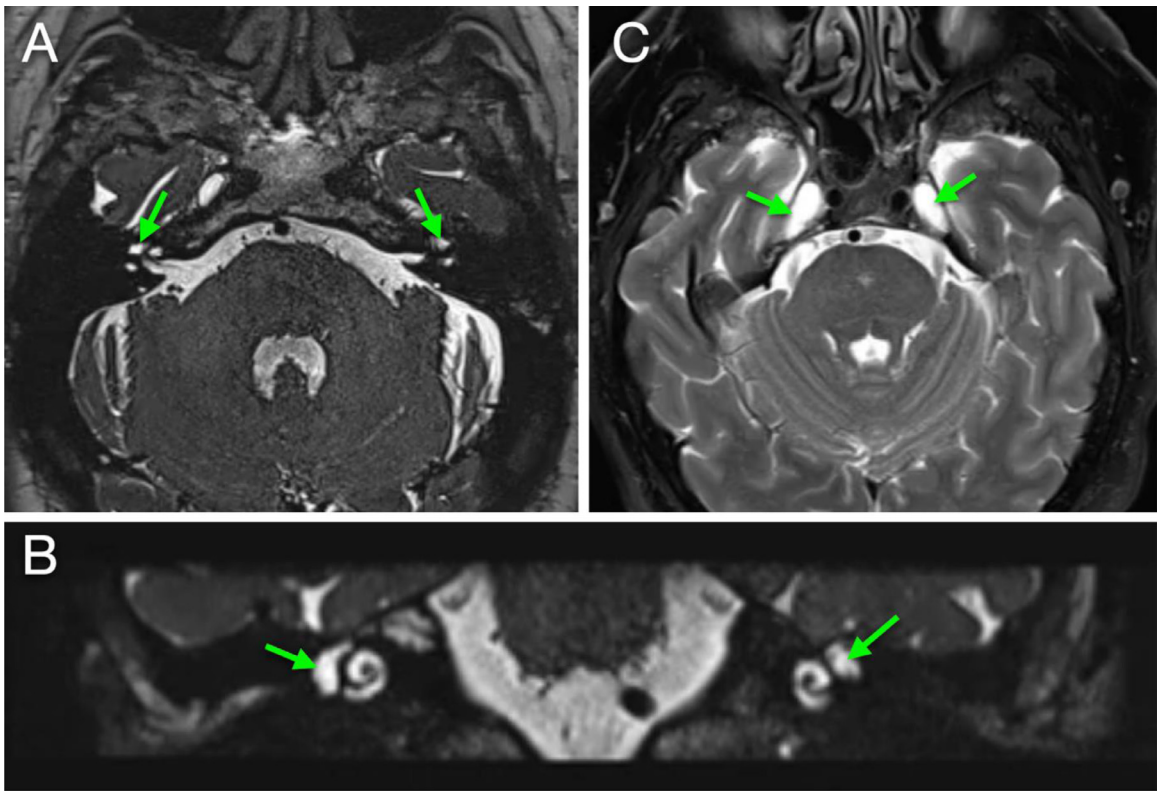

Fig. 2 – Proton density weighted magnetic resonance images demonstrating: (A) T2 SPACE axial view of bilateral fluid signal along the labyrinthine, geniculate and proximal tympanic segment of the facial canal bilaterally (B) T2 SPACE coronal view of bilateral fluid signal involving the geniculate ganglion and (C) T2 axial view of prominent Meckel's cave.

demonstrated distended Meckel's caves bilaterally in the middle cranial fossa (Fig. 2C). There were no other retrocochlear or intracanalicular lesions found to explain the patient's sensorineural hearing loss, and no other features of raised intracranial pressure (ICP).

The patient was subsequently referred to the Neuro-ophthalmology team for further investigation of IIH. His hearing loss was managed conservatively with hearing aids given there were no other imminent complications associated with his meningoceles such as a CSF leak.

Discussion

IIH refers to a syndrome caused by raised ICP without any identifiable cause and is diagnosed using the modified Dandy criteria [4,6]. MRI is now frequently used as an adjunct to identifying key features commonly seen in IIH patients; these can include an empty sella, flattening of the posterior globes, optic nerve changes, cerebellar tonsillar herniation, transverse venous sinus stenosis, meningoceles and a dilated Meckel's cave [1,2,4,5,7]. Many of these findings can be used to suggest IIH in situations where major diagnostic criteria are not fulfilled.

In the context of IIH, meningocele formation is thought to result from increased CSF pressure and bony remodeling leading to outpockets of dura through defects in the base of skull [7]. The incidence is estimated to be around 11% when compared to controls and can develop anywhere along the skull base, although most commonly involve the petrous apex [1,4,8].

Clinical features of meningocele formation are variable depending on their location. Oftentimes, they are incidental until they rupture, resulting in a CSF leak. In those circumstances, patients may present with clear otorrhea or rhinorrhea, meningitis, headache, diplopia, pulsatile tinnitus or visual disturbances [1,9]. It can also cause a conductive hearing loss if the meningocele causes a mass effect on the middle ear ossicles, as seen in our case.

Meningoceles in the facial canal are particularly rare and have only been reported in three previous studies; these meningoceles were all unilateral and located exclusively in the geniculate ganglion [10–12]. Brackmann et al. 2007[10] reported a case of an enlarged right fallopian canal at the geniculate ganglion in a young child who presented with facial nerve palsy and progressive asymmetrical right sided high frequency sensorineural hearing loss. Similarly, Mong et al. 2009 [11] reported two patients who each presented with unilateral meningoceles involving the geniculate ganglion. Both presented critically unwell and proceeded to have surgery for repair of the CSF leak. Dey et al. 2019 [12] also reported on a right sided geniculate fossa meningocele which was thought to result from a defect between the fallopian canal and middle cranial floor.

Bilateral facial canal meningoceles in the context of IIH have only been described once in the existing literature. Kabuli et al. 2019 [13] reported a similar case to ours who presented with bilateral SNHL and other radiological features of IIH including dilated optic nerve sheaths, dilated sella turcica and dilated Meckel's caves. MRI revealed bilaterally dilated geniculate fossa with high T2 signal suspicious for facial canal meningoceles. Our study is thus the second to report on the presence of bilateral facial canal meningoceles in a patient with features suggestive of IIH. It is, however, the first where the meningocele involves the tympanic segment of the canal, which is a much rarer finding compared to the classic location at the geniculate ganglion.

A combination of CT and contrast MRI is often needed to confirm diagnosis of facial canal meningoceles. Initial CT demonstrates abnormal density along the canal however it is not sufficient to differentiate between fluid or soft tissue [14,15]. At this point, differential diagnoses could include a fluid filled meningocele, or a tumor such as a facial nerve schwannoma.

Contrast MRI often shows a T2 hyperintense signal with lack of diffusion restriction in the affected segment of the facial canal, consistent with fluid accumulation [2,7,15]. The lack of enhancement further supports the diagnosis of a CSF filled meningocele, versus a schwannoma which would show enhancement on MRI. These findings, in conjunction with a dilated Meckel's cave increase suspicion for IIH [2–4,7].

Treatment of meningocele formation does not always necessitate surgical treatment; in extreme cases where there is rupture of the meninges with subsequent CSF leak, surgical repair is often required and may involve repair of the tegmen defect either via a transmastoid or middle cranial fossa approach [11,12]. In other cases where the patient's symptoms are mild, such as in our case, conservative management is often preferred given the high risks associated with surgery.

Summary

This is the first reported case of bilateral facial nerve meningoceles that involve not only the geniculate ganglion but also the tympanic segment of the canal. The patient has radiological evidence of IIH, which is postulated to be the mechanism behind the meningocele formation.

Patient consent

We confirm that written, informed consent was obtained for publication of this case from the patient being discussed. The consent form has been signed by the patient and if required can be submitted to the Journal as evidence.

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