CASE STUDY

Atypical Etiology of Rhinorrhea: Spontaneous Bilateral Temporal Encephalocele*†

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Abstract  Spontaneous herniation of brain parenchyma through a dural and osseous defect in the temporal bone is a rare entity and a bilateral form is even more infrequent. It usually presents as an intermittent but persistent otorrhea. Manifestation as nose cerebrospinal fluid (CSF) leak is very uncommon. Our objective is presenting this unusual case report of a spontaneous bilateral encephalocele with a bilateral tegmen tympani defect.
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Rinolicuorraquia de etiología atípica: encefalocele espontáneo temporal bilateral

Resumen  La herniación espontánea de tejido cerebral a través de un defecto óseo y dural a nivel temporal es una rara entidad, siendo todavía más infrecuente que esta circunstancia se produzca de forma bilateral. La presentación suele ser en forma de otorrea intermitente y persistente en el tiempo. La manifestación como rinolicuorraquia es muy poco habitual. El objetivo es presentar este inusual caso de un encefalocele espontáneo bilateral por defecto bilateral del tegmen tympani.
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Introduction

Skull base brain encephalocele is a rare finding, with an incidence of 1/35 000.1 In most cases, it is associated with other conditions such as trauma, infection, inflammation, tumours, radiation therapy or prior surgery.1–3 Spontaneous cases are even more rare, accounting for less than 9%,4 with the unilateral form being the most common.

The most common clinical presentation takes the form of otorrhea, often being confused with serous otitis media,1,3,5 and up to 20% of cases have a history of meningitis.6

We present a rare case of bilateral spontaneous encephalocele with an atypical presentation in the form of rhinorrhea or nasal CSF leak.

Clinical Case

We present the case of a 50-year old male who had recently suffered meningitis and was referred to our centre for
evaluation due to right rhinorrhea with various months of evolution. The ENT examination found no abnormalities in the tympanum. Nasal fibroscopy found a CSF leak by the Eustachian tube in the right Rosenmüller fossa. Isotope cisternography revealed signs of nasal fistula without signs of fistula in the ears. A bone CT scan of the skull base and ears (Fig. 1A and B) showed occupation of the antrum, mastoid air cells and tympanic cavity by CSF on the right side, as well as anterosuperior dehiscence of the tegmen tympani, with an image indicative of encephalocele. In addition, the image also revealed clear dehiscence of the tegmen tympani on the left side, with an image indicative of encephalocele which prolapsed into the middle ear.

The study was completed with an MRI scan which confirmed the diagnosis and ruled out the existence of cerebritis or abscess (Fig. 2A and B).

Bilateral involvement was considered, but was finally discarded in favour of reconstruction of the right side, as this appeared to be the origin of the fistula.

An extradural subtemporal approach was performed through temporal craniotomy, retracting the temporal lobe in order to visualise the bone dehiscence and encephalocele. Devitalised tissue was found and subsequently coagulated and removed. The reconstruction process employed artificial dura mater (DuraGen®) and autologous fascia temporalis, and involved sealing with bioglu and placing an osteoinductive agent (Copios®) for the bone defect.

External lumbar drainage with antibiotic coverage was maintained for 4 days.

After 24 months, the patient remained asymptomatic, with regular neuroimaging controls of the left encephalocele being performed.

**Discussion**

The existence of spontaneous meningoeceles in the middle fossa due to dehiscence of the tegmen tympani is rare and their aetiology is unknown. Some autopsy studies report a relatively high rate, between 18% and 21%. Some authors postulate that this dehiscence has a congenital origin, due to a lack of chondral ossification, but common presentation in middle-aged patients would be difficult to explain. Another, perhaps more widely accepted theory, attributes this defect to the presence of arachnoid granulations which would erode the bone after decades by the action of intracranial pressure and pulsatility, causing a thinning of the dura mater and ultimately causing meningoeencephalocele.

Clinically, otological manifestations are usually predominant. The most common is the existence of an "effusion" or otorrhea in the middle ear with
Conductive hearing loss (often confused with serous otitis media\textsuperscript{2-7}) and showing CSF leak after performing a myringotomy. Rhinorrhea is rare.\textsuperscript{11} The determination of b2-transferrin can confirm the diagnosis.\textsuperscript{1-5} Other symptoms may include vertigo syndrome, facial paralysis and even epilepsy in encephaloceles. Approximately 20%-40% of cases have a history of recurrent meningitis.\textsuperscript{4,14}

Diagnosis and surgical planning require high-definition axial and coronal CT scans of the temporal bone,\textsuperscript{1,4-6,11} with 3D reconstructions also being useful. Performing T1 and T2 MRI scans is recommended as this helps to identify the existence of a meningo/encephalocele in greater detail, and can also be useful in the differential diagnosis of cholesteatoma, granulation tissue, cholesterol granuloma and meningoencephalocele.\textsuperscript{4,7} Cisternography has become relegated, since it provides scarce anatomical information, as it only confirms the presence of a fistula.\textsuperscript{5}

The approach pathway should enable adequate exposure of the area and facilitate reconstruction. The most commonly used methods are transmastoid extradural, temporal craniotomy or a combination of both.\textsuperscript{4,11}

The transmastoid pathway is less invasive, enables access to the posterior and middle fossae and is very useful in tegmen mastoideum defects,\textsuperscript{3,7,11} although it may be insufficient for large defects of the tegmen tympani. Its main disadvantage is the risk of damage to the ossicular chain, thus some authors reserve this approach for cases with no useful hearing.\textsuperscript{6}

Temporal craniotomy provides an excellent surgical field and enables ideal dural reconstruction and bone repair.\textsuperscript{4,5} However, it is more aggressive and may not be feasible in older patients. It is the procedure of choice in large defects or cele\textsuperscript{4} and also facilitates direct dural closure, preventing damage to the ossicular chain and preserving hearing.\textsuperscript{6}

In conclusion, treatment should be individualised according to the characteristics of each patient, location and size of the defect and cele.\textsuperscript{4,5,11}

Regarding the surgical technique, the hernial content is considered to be devitalised and not functional,\textsuperscript{4,15} so its extradural excision is recommended. This should be done by coagulating the neck,\textsuperscript{7} except in large cele where it would be better to reintroduce it (Fig. 3A and B). If feasible, direct suture is the best option for the repair of the dural mater.\textsuperscript{4} It is also possible to employ dural plasties or autologous tissue such as free or pedicled fascia temporalis and pericranium.\textsuperscript{1,4-7,15} A bioglue can also be associated. Some authors use pedicled temporalis muscle flaps as a last barrier. Both autologous bone and hydroxyapatite cements are valid for bone reconstruction of the tegmen tympani.\textsuperscript{11,16,17}

Finally, there is no agreement on the use of external lumbar drainage. While there is no evidence in this respect, it may be useful in order to decrease CSF pressure,\textsuperscript{1,4,11} with a consequent reduction in size of the cele, and for injecting fluorescein. It should be maintained for 3-5 days postoperatively.

Conflict of Interests

The authors have no conflicts of interest to declare.

References


