CASE REPORT

An unusual case of dysphasia and seizures

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Accepted 24 July 2014

SUMMARY

Defects in the tegmen tympani can have serious consequences such as intracranial infection and seizures, however, they can be difficult to diagnose due to their subtlety on cross-sectional imaging and the non-specific way in which they can present. Repair of such defects can be undertaken via a middle fossa or transmastoid approach; both, however, usually necessitate a general anaesthetic and, furthermore, middle cranial fossa surgery carries the risk of epilepsy while transmastoid repair can result in the loss of hearing. We present a novel method of repair in a patient who would otherwise have been managed conservatively.

BACKGROUND

The tegmen tympani is the part of the temporal bone that makes up the roof of the attic of the middle ear. Tegmen defects may lead to a temporal lobe encephalocele and can present as a conductive deafness, with cerebrospinal fluid (CSF) otorrhoea or rhinorrhoea, pulsatile tinnitus,¹ seizures or intracranial infection, or can be found as an incidental finding on cross-sectional imaging. Defects are either congenital, secondary to trauma, secondary to infection (ie, chronic otitis media), iatrogenic or idiopathic. Sanna *et al*² have shown tegmen defects with encephalocele with an aetiology of spontaneous 24.8%, chronic otitis media 21.8%, iatrogenic 45.9% and post-traumatic 7.5%. Locating a defect with cross-sectional imaging is often difficult, with abnormalities often being very subtle. High-resolution CT (HRCT) will, however, pick up most bony defects with MRI distinguishing an encephalocele from middle ear disease.³

Repair of tegmen defects can be undertaken via a middle fossa craniotomy, a transmastoid approach or through a combined approach.³ The repair is best undertaken as a multilayer closure using auto- and allogeneic materials.⁴ The choice is usually based on the location of the defect, presence of concurrent infection and operative fitness. Transmastoid surgery may result in the loss of hearing as the middle ear is usually obliterated as part of the repair; less of the defect is exposed during the procedure and the success rate is, therefore, lower than that of a middle cranial fossa approach.³

CASE PRESENTATION

A 60-year-old obese woman was admitted to the medical assessment unit following a grand mal seizure with a 24 h history of expressive and receptive dysphasia, headache and confusion. When assessed, she was noted to be restless and confused and a further grand mal seizure was witnessed;

Figure 1 Axial non-contrast CT scan demonstrating a left temporal lobe hypodensity.

neurological examination revealed only up-going plantars.

This presentation was preceded by a similar episode 7 years previously, during which she was assessed by the medical and ear, nose and throat (ENT) department teams and was investigated by CT scan of the head, which failed to demonstrate any pathology. She had multiple comorbidities, including chronic obstructive pulmonary disease, hypertension and a previous intensive care unit admission for renal failure and sepsis secondary to cellulosic leg ulcers; despite this she lived independently with her husband; she gave no prior otological history.

INVESTIGATIONS

Routine bloods showed a raised white cell count of $14.1 \times 10^9/L$ with neutrophils of $12.4 \times 10^9/L$, arterial blood gas demonstrated hypoxia and chest X-ray was unremarkable.

Figure 2 High-resolution CT of left temporal bone in coronal section.

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To cite: Williams MR, Easson A, Khwaja S, *et al*. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2014-204988

Figure 3 High-resolution CT of left temporal bone in sagittal section.

She underwent a CT scan of the head, which was reported as showing a small area of recent haemorrhage with surrounding oedema in the left temporoparietal region (figure 1).

She continued to have grand mal seizures that responded slowly to intravenous antibiotics and antiepileptics, she was left with a slowly improving dysphasia.

A subsequent MRI of the head was reported as showing an area of cerebritis associated with a low-grade mastoiditis, rather than the initial CT findings suggestive of infarct. A lumbar puncture 3 days following was suspicious for encephalitis and therefore acyclovir was started. ENT evaluation revealed a dull left tympanic membrane and conductive deafness, normal cranial nerve examination and on-going dysphasia.

A CT of the temporal bones showed subtotal opacification of the mastoid air cells, mastoid antrum and middle ear cavity on the left hand side, in keeping with severe mastoiditis. There was evidence of thinning and erosion of the tegmen tympani (figures 2 and 3). The incus, malleus and scutum appeared intact.

This was at odds with examination in the clinic revealing a normal tympanic membrane, leading to uncertainty as to the cause of the middle ear opacification. It was decided to explore the middle ear following a repeat MRI to exclude a vascular tumour.

DIFFERENTIAL DIAGNOSIS

Initially the patient was managed for meningitis complicated by seizures.

Figure 4 Preoperative pure tone audiometry.

TREATMENT

An elective tympanotomy found a pink polypoidal mass in the middle ear, which gave a gush of clear fluid on biopsy. Histology proved this to be brain tissue, leading to a diagnosis of left temporal lobe herniation through a tegmen tympani defect (temporal bone encephalocele).

Clinical follow-up showed the tympanic membrane had healed successfully but the patient had developed CSF rhinorrhoea. A neurosurgical review confirmed an on-going CSF leak, which would normally be repaired via craniotomy. Owing to the patient's comorbidities, it was felt that such an approach would be too risky, and a decision was made to attempt repair under local anaesthesia using a transmastoid approach.

During the elective procedure the lateral attic wall was burred away and a 1 cm defect in the tegmen tympani was identified. The herniated temporal lobe was reduced into the middle cranial fossa. Conchal cartilage was inserted via the bony defect and covered with bone chips; this was covered with free and pedicled temporalis fascia.

OUTCOME AND FOLLOW-UP

The procedure has been successful with no complications or recurrence of symptoms. Postoperative audiometry has shown an improvement in the patient's hearing (figures 4 and 5).

DISCUSSION

We feel this case highlights several important points relating to the difficulty in diagnosing a tegmen defect. Our patient gave no prior otological history, did not have any symptoms at presentation to suggest middle ear disease and only on otological examination was the evidence of a middle ear effusion found. The middle ear is an important source of intracranial pathology and the benefit of otological examination as a routine part of neurological examination in this case is clear. In this patient an earlier ENT review might have brought to light an inflamed tympanic membrane suggesting the diagnosis earlier.

The importance of appropriate imaging is also clear, as the initial CT scan failed to diagnose the tegmen defect, only MRI and subsequent HRCT of the temporal bones highlighted middle ear pathology, the HRCT revealing the defect to the tegmen tympani. Had this follow-up imaging not been performed the diagnosis might never have been obtained, resulting in potentially fatal consequences.

Figure 5 Postoperative pure tone audiometry.

The gold standard for tegmen repair is considered to be a middle fossa approach, owing to its success rate and durability. However, the incidence of epilepsy following a middle cranial fossa surgery has been noted to be as high as 22%⁵ and many patients are not considered suitable for a craniotomy due to their

comorbidities. As in our case, such patients undergo transmastoid repair. In our case we have demonstrated it is possible to undertake such a repair without the need for general anaesthesia, thus lowering the risk for such patients, although we accept that this requires a cooperative patient and a confident surgeon.

Learning points

- An otological history and examination should always be considered relevant to intracranial lesions, particularly those presenting in the temporal lobe.
- Patients with a tegmen defect may be asymptomatic or present with symptoms easily attributable to other causes.
- Defects on the tegmen are only likely to be seen on specialised imaging (ie, high-resolution CT of the temporal bones).
- Repair of a tegmen defect under local anaesthesia may be considered in those who are not fit for a craniotomy or a general anaesthetic.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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