Case report

Transnasal transpterygoid resection of meningoencephalocoele with abolition of seizures

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SUMMARY

Meningoencephalocoeles are herniations of intracranial contents through skull base defects. Patients may present with a plethora of symptoms ranging from cerebrospinal fluid (CSF) rhinorrhoea to epileptic convulsions, or indeed may be asymptomatic. We present a case of a 24-year-old man suffering from new onset, drug-resistant tonic-clonic seizures. Imaging studies revealed neural tissue and meninges protruding through the pterygoid portion of the sphenoid bone, into the anteromedial aspect of the middle cranial fossa. An image-guided endoscopic transnasal transpterygoid approach was carried out as a joint otolaryngological and neurosurgical procedure, resulting in the patient being seizure-free for over 12 months postoperatively. There is a paucity of literature supporting such an endoscopic approach to treat epilepsy secondary to a meningoencephalocoele. We illustrate that this is a safe and minimally invasive treatment option which ultimately rendered the patient free of all anticonvulsants.

BACKGROUND

Skull base meningoencephalocoeles refer to intracranial contents which have herniated through defects in the skull base and comprise both meninges and neural tissue.¹ The aetiology of these osseodural defects vary and may be divided into congenital, acquired or spontaneous.² It has been postulated that congenital meningoencephalocoeles are caused either by failure of neural tube closure or by incomplete fusion of precursor sphenoid wings to the basisphenoid and presphenoid, leading to the formation of the lateral craniopharyngeal canal of Sternberg.^{3,4} On the other hand, the leading causes of acquired meningoencephalocoeles are mainly traumatic, neoplastic, infectious or metabolic destruction of skull base.^{5,6} Patients may be entirely asymptomatic or indeed present with cerebrospinal fluid (CSF) rhinorrhoea, or features of intracranial involvement including meningitis, meningoencephalitis or brain abscess. Should the lesion herniate into the sphenoid sinus, there is a significant risk of developing temporal lobe epilepsy which may be refractory to medical treatment, thought to be in part due to traction on the herniated medial temporal lobe.^{3,7-9} Traditional approaches have centred on open craniotomies, until Bolger⁶ first described the endonasal transpterygoid approach to access the lateral recess of the sphenoid sinus. However, to date, very few cases have ever been documented of successful treatment of

meningoencephalocoeles presenting with epileptic seizures.³ The present paper describes one such case of successful endoscopic treatment of a meningoencephalocoele within the pterygopalatine fossa that had presented with grand mal seizures.

CASE PRESENTATION

An otherwise fit and well 24-year-old man was referred to our tertiary referral skull base unit via neurology in December 2017. Over the preceding 6 months, he had been experiencing twice monthly new onset tonic-clonic seizures and was on treatment with levetiracetam, with limited success. He had no history of head trauma or other relevant medical history whatsoever.

INVESTIGATIONS

An initial EEG revealed phase-reversing irregular epileptiform sharp focal waves emanating from the right anterior and middle temporal region, supporting an underlying partial onset seizure disorder of right temporal origin. As part of the work-up, an MRI scan identified a fluid-filled expansile lesion in the pterygoid portion of the right sphenoid bone. Neurological examination was otherwise unremarkable, and there was no evidence of a CSF leak. While often recommended for temporal lobe epilepsy, fluorodeoxyglucose-positron emission tomography (FDG PET) scanning was not recommended by two separate multidisciplinary neuroradiology meetings, comprising neurosurgeons, neurologists, neuroradiologists and histopathologists. Further advice from a dedicated multidisciplinary skull base unit was sought. Second, the patient was referred for consideration of video EEG, but due to the infrequency of the seizures, this was not deemed appropriate.

DIFFERENTIAL DIAGNOSIS

Due to the fluid-filled nature of the lesion on the MRI study, it was postulated as being either a mucocoele or a meningo(encephalo)coele. Further discussion at the Skull Base Multidisciplinary Team Meeting suggested brain tissue involvement that was causing the new onset epileptic activity due to traction on the temporal lobe. The final diagnosis could only be made intraoperatively but no additional pathology was identified as a cause for the epilepsy.

TREATMENT

A review of the patient's CT (figure 1) and MRI imaging (figure 2) at the Skull Base Multidisciplinary

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Figure 1 Coronal plane, non-contrast CT scan demonstrates the right-sided bony defect and meningoencephalocele.

Team Meeting identified an abnormality in the right pterygoid base. It was felt that the lesion would be eminently accessible via an endoscopic transnasal approach.

An image-guided endoscopic transnasal transpterygoid approach was carried out as a joint otolaryngological and neurosurgical procedure (figure 3). An initial partial middle turbinectomy was performed along with an uncinectomy and wide middle meatal antrostomy. A sphenoethmoidectomy was completed and a right-sided nasoseptal flap was raised (figure 4). The bony posterior wall of the maxillary antrum was gently removed and the periosteum of the pterygopalatine fossa incised—immediate bleeding from the internal maxillary artery which was compressed against the anterior wall of the fossa was encountered and controlled with diathermy. Just behind this, there was a fluid-filled space expanding into a bony cavity and effectively obliterating the right pterygopalatine fossa. The vidian and V2 maxillary nerve becoming the infraorbital nerve were identified. The branches of maxillary nerve were identified and mobilised inferolaterally to provide access to the cavity which was identified with meninges and part of temporal lobe herniating through the defect, just lateral to the maxillary nerve. Aberrant brain tissue retracted into the cranial fossa as it was diathermied, leaving a dural defect of 0.5–1 cm and a slightly larger bony defect. Next, the medial wall of the cavity was removed, opening it widely into the sphenoid sinus, which was facilitated by a posterior septectomy to allow access via the

Figure 2 Coronal plane, T2-weighted, MRI scan to demonstrate the meningoencephalocele.

Figure 3 Image-guided endoscopic transnasal transpterygoid approach of the meningoencephalocele in the right middle cranial fossa.

contralateral nares and nasal cavity. The vidian nerve and V2 nerves were preserved. Closure of the dural skull base defect was carried out with a collagen dural matrix (Duragen) inlay graft, and an onlay nasoseptal flap. Surgicel was applied to the edges of the flap followed by Duraseal mist. A 12F Foley catheter was then used to support the repair site. A lumbar drain was not deemed necessary.

OUTCOME AND FOLLOW-UP

The patient's initial postoperative course was uneventful with no evidence of a CSF leak. A repeat MRI was carried out at 5 months postsurgery and showed a satisfactory reconstruction with no residual meningoencephalocele. Of particular note is that to this date, 18 months from his surgery, he remains seizure-free. A repeat EEG, 12 months after his initial EEG, revealed no specific underlying focal, paroxysmal or epileptiform features of

Figure 4 Endoscopic approach of the right sphenoid wing, post middle turbinate partial excision.

note—and was within normal limits for his age. After review by the neurologists in January 2019, as he was completely seizure-free with a normal EEG, he was deemed to no longer be epileptic and all anti-seizure medications were weaned off.

DISCUSSION

Skull base meningoencephalocoeles mainly concern congenital defects in the midline of the skull base, causing glial tissue, meninges and CSF to herniate into the nasal cavity.⁷ They are rare with an incidence of about 1 in 40 000 live births.¹⁰ Traditionally, they are classified depending on the site of herniation as ‘frontoethmoidal’, ‘sphenoorbital’, ‘sphenomaxillary’ and ‘nasopharyngeal’, further being divided into transtethmoidal, sphenothmoidal and sphenopharyngeal meningoencephalocoeles.⁷ Lately, there has been increasing evidence of a separate category from those, the herniation occurring in the parasellar area, adjacent to the lateral recess of the sphenoid sinus.^{7 8} In the present patient, the meningoencephalocoele was localised to the pterygoid fossa, sparing the orbit, both the orbital fissures and the nasal cavity. The main theory of this location of herniation concerns failure of fusion of ossification centres of the sphenoid during the early embryologic stages of sphenoid bone formation.⁸

A review of the literature revealed very few studies relating to patients presenting with temporal lobe meningoencephalocoeles; most of these were limited to case reports.^{4 8} They typically presented with seizures and patients treated with a temporal lobectomy via a transcranial or infratemporal approach. The cause of the seizures is presumed to be either related to irritation or a traction effect of the glial tissue herniating through the skull base defect into upper nasal cavity, or by coexisting malformations; the most important being diffuse cortical dysplasia.^{7 8 11} Historically, these patients were treated with large lobe resections through a transcranial approach. However, there has been an increase in the recent reported prevalence of meningoencephalocoeles in patients with medical refractory epilepsy, with Campbell *et al* going as far as reporting a 12.5% among these patients.¹² These new findings stress the need for a less invasive treatment for these patients suffering from epilepsy as a consequence of temporal meningoencephalocoeles. Although there are some reports of tonic-clonic seizures from temporal lobe encephalocoeles, more typically, one expects to see a partial seizure followed by secondary generalisation.¹³

The transpterygoid approach was first described in 2005 and provides excellent visualisation and direct access to the lesion extracranially, thereby limiting manipulation of the brain.⁶ It potentially affords less morbidity and is continuing to gain popularity.⁵ Faulkner *et al*¹¹ reported a similar case which was also treated successfully endoscopically, although the patient remained on anti-epileptic medical treatment despite being seizure-free. It should, however, be noted that this case could potentially have a good seizure outcome because preoperative seizures started 6 months before surgery. In epilepsy refractory to drug treatment over several years, more complex epileptogenic network may be present.

Where the present paper differs from the aforementioned studies, our patient was not only seizure-free with normal encephalography, but deemed to also no longer require anticonvulsant medications; to the authors’ knowledge, this has never

Learning points

- ▶ Meningoencephalocoeles may present with a plethora of symptoms; not only headaches and cerebrospinal fluid rhinorrhoea, but also epileptic seizures.
- ▶ The endoscopic transnasal transpterygoid approach affords a safe approach to such lesions at the pterygoid base, providing excellent visualisation, and minimal morbidity through reduced manipulation of brain tissue.
- ▶ In carefully selected cases, the endoscopic transnasal transpterygoid approach has the potential to render the patient seizure-free and to no longer require anticonvulsant medication.

previously been reported in the literature. The patient remains on regular follow-up by both neurology and otolaryngology. While the results in the present case have been striking, further more substantial studies are warranted in the future.

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