Traumatic intranasal meningoencephalocoele with a cerebrospinal fistula

Zalilah MUSA [1], Irfan MOHAMAD [1], Nik Khairani NIK MOHAMAD [2]
[1] Department of Otorhinolaryngology-Head & Neck Surgery, School of Medical Sciences, Universiti Sains Malaysia, Kelantan, Malaysia and
[2] Department of Otorhinolaryngology- Head & Neck Surgery, Hospital Sultanah Nur Zahirah, Terengganu, Malaysia

ABSTRACT

Intranasal meningoencephalocoele is a rare clinical entity especially in the adult population. It is usually a congenital anomaly but can occur as a result of traumatic head injury or increased intracranial hypertension. We report the case of 25-year-old Malay man who presented with persistent headache of one-year duration that was associated with bilateral nasal blockage and intermittent right nasal discharge. He was also treated for meningitis and previously had two episodes of closed head injuries. Investigations showed that he had an intranasal transethmoidal meningoenchepalocoele. This was successfully treated with an endoscopic excision and fistula repaired using cartilage graft and tissue glue.

Keywords: Meningitis, cerebrospinal fistula, cerebrospinal leak

INTRODUCTION

An intranasal meningoencephalocoele is characterised by a protrusion of the meninges from the cranial cavity into the nasal spaces through a defect in the ethmoidal cribriform plate. [1, 2] It is usually associated with congenital anomalies but can also occur after traumatic fractures of the ethmoidal cribriform plate, post rhinological operations or through bone erosion from raised intracranial pressure, neoplasm, granulomatous or chronic infections. [2-4] Patients with intranasal meningoencephalocoele typically present with chronic sinus problems with or without with cerebrospinal fluid (CSF) leak and meningitis. We report the rare case of a 25-year-old Malay man with post traumatic meningoencephalocoele. The patient was symptom free after undergoing successful transnasal endoscopic excision of meningoencephalocoele and repair of the fistula.

Correspondence author: Irfan MOHAMAD
Department of Otorhinolaryngology-Head & Neck Surgery, School of Medical Sciences,
Universiti Sains Malaysia Health Campus,
16150 Kota Bharu, Kelantan, Malaysia.
Tel: 690-7676420
E mail: irfan@kb.usm.my

CASE REPORT

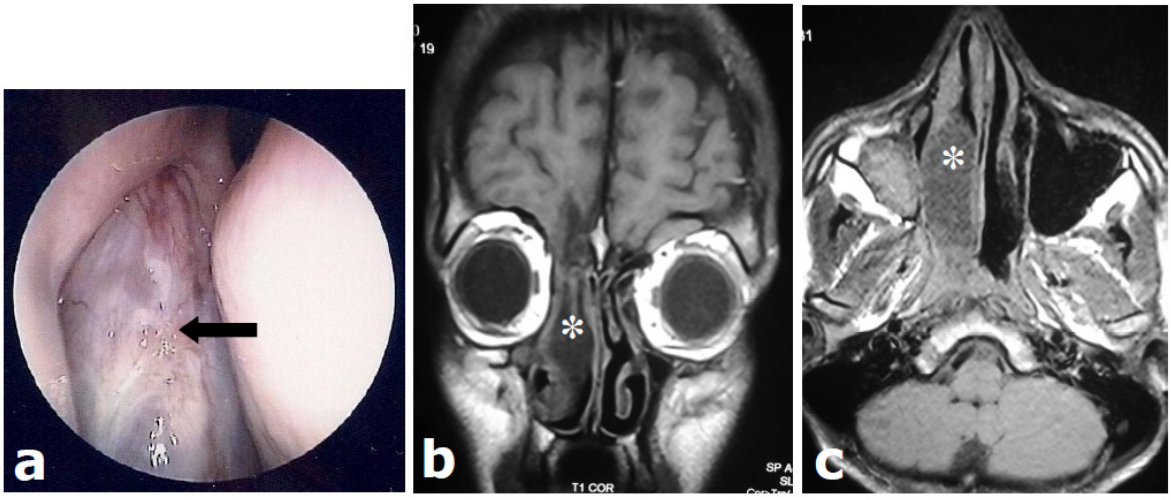
A 26-year-old Malay man was referred by the neuro-medical team with persistent headache.

This was associated with bilateral nasal blockage and intermittent watery right nasal discharge of more than one year duration. He was treated for meningitis with hydrocephalus a year prior to the current admission. He had two head injuries that were associated with two motor vehicle accidents approximately five and nine years ago.

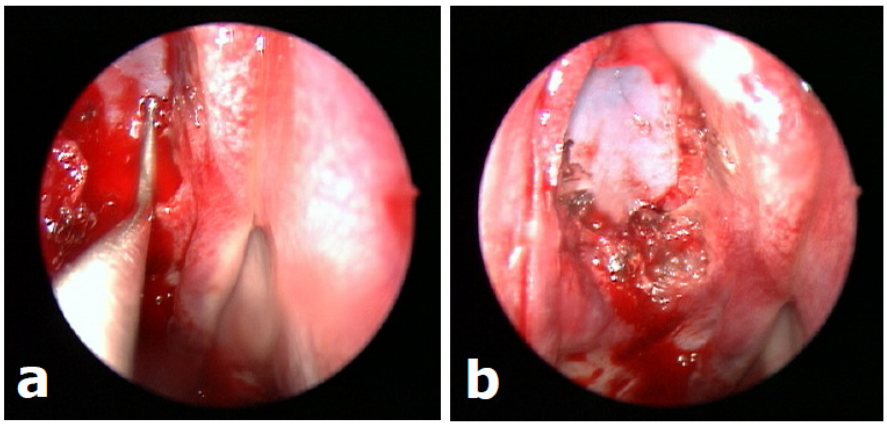
Clinical examination revealed no neurological deficits. An anterior rhinoscopic examination showed the presence of a right nasal mass. A zero degree nasal endoscopy which was used to visualise the whole intranasal anatomy confirmed the huge intranasal mass arising from the right middle meatus (Figure 1a). It was pulsatile in nature and increased in size with a valsalva manoeuvre. A computed tomography (CT) scan and a Magnetic resonance imaging (MRI) of the paranasal sinus and brain showed the presence of a right ethmoidal meningoencephalocoele extending to the right nasal cavity (Figures 1b and c). Blood investigations including white cell count and serum glucose were

The patient later proceeded for an endoscopic excision of meningoencephalocoele with fistula repair with prophylactic antibiotic (intravenous cefuroxime, 1.5 gm). A lumbar drain was inserted prior to operation to monitor the CSF pressure. Intraoperatively, there was a large pulsating intranasal meningoencephalocoele occupying the right nasal cavity. Trickling of CSF could be seen. The meningoencephalocoele was traced up to its pedicle and cauterisation was done at the pedicle prior to removal (Figure 2a). A septal cartilage was harvested through the right nasal cavity via a Killian incision. The graft was placed at the defect and sealed with fibrin glue (Figure 2b).

Postoperative nasal endoscopy revealed a healthy graft around the defect with no further CSF leak (Figure 3). Histopathology of the mass revealed only glial tissue. At follow-up, six months after surgery, the graft was intact and showed no further CSF leak. As the patient did not have any clinical symptoms and signs of recurrence, he was monitored clinically and endoscopically without any



Figs. 1: a) Endoscopic image showing an intranasal mass (black arrow) arising from the roof of nasal cavity at cribriform plate, b) A coronal MRI image and c) Axial MRI image showing the intranasal meningoencephalocoele (asterisks).



Figs. 2: a) Intraoperative image showing the defect (arrow) at the cribriform plate, and b) after closure with a nasal cartilage graft.

DISCUSSION

Encephalocoeles are herniation of cranial contents through the skull. They may contain meninges (meningocoele), meninges and brain (meningoencephalocoele) or meninges, brain and part of ventricle system (meningoencephalocytocoele). [1] Meningoencephalocoeles can be classified as occipital, sincipital, and basal. Basal meningoencephalocoele was first described by Richter in 1813. It was first classified by Gisselsson in 1947 into four types based on their anatomic locations: transethmoidal, sphenoethmoidal, transsphenoidal, and sphenomaxillary. [5] The transethmoidal type is the most common and the cranial defects can be found anterior to the sphenoid bone usually through the ethmoid or cribriform plates. The meningoencephalocoele may extend only as far as the ethmoidal sinus or may protrude through the superior meatus into the nasopharynx where it can be misdiagnosed as a nasal polyp or hypertrophied adenoidal tissue. [2]

The cribriform plate is a fragile and vulnerable structure. Trauma from closed head injury is actually the most common cause of CSF leak occurring in one to three percent of all closed head injuries. [6] Persistence of these defects can lead to the formation of meningoencephalocoeles. It is considered rare in the adult population. Immediately after head trauma, patients may present with CSF leak (most within 48 hours and 95% manifest within three months). [6] This act a as route for ascending infection leading to bacterial meningitis. [2-4] Intranasal meningoencephalocoele usually develops months later manifesting with symptoms of chronic sinusitis, nasal obstruction, headache and CSF rhinorrhoea. However, CSF leakage may stop or

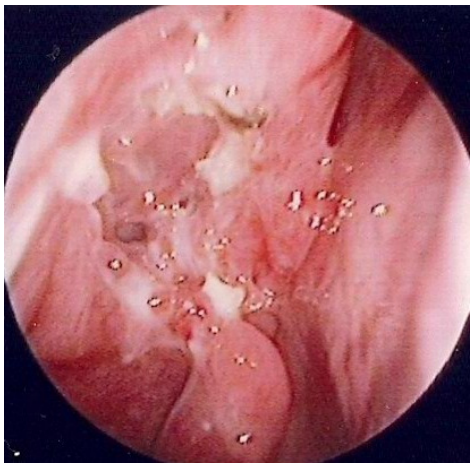


Fig. 3: Endoscopic image showing a healthy cartilage graft without evidence of further CSF leak.

become intermittent as results of closure of the fistula from adhesions or herniation of brain. However, this is usually insufficient. [4] Bacterial meningitis usually develops within the first three months of head injury and this can be recurrent. [1, 4] Gienta *at el.* reported a atypical patient who presented with recurrent meningitis over a five-year period after a closed head injury. [4] In our case, our patient presented with a single episode of meningitis five years after the second head injury. Generally, such a presentation as in our patient is rare and unusual.

A patient with intranasal meningoencephalocoele may have hypertelorism with a widened nasal root. [1] This is more pronounced in the congenital type, as the growing frontonasal bone (which determines the interpupillary distance) may expand according to the space occupied by the herniated mass. Both the congenital and post-traumatic types may be visible on inspection of the nasal fossa. It typically appears as a soft whitish, non-pedunculated, pulsatile mass which must be differentiated from a polyp and other congenital intranasal mass such as haemangioma, dermoid cyst, nasal glioma or ectopic adenoid tissue. [1-4] The encephalocoele is compressible and may display a positive Furstenberg sign (expansion with valsalva manoeuvre or with compression of internal jugular vein). [1] Failure to differentiate between a meningoencephalocoele from other pathologies can lead to life threatening consequences after surgical interventions.

A skull radiograph may show a defect in skull base on a submento-vertex projection but is generally not helpful. Small defects in the cribriform plate are usually not obvious on plain radiography. [4] Lateral skull views may show a nasal or nasopharyngeal mass. [1] CT scan and MRI are very useful and they can demonstrate the defect in cribriform plate with extension of soft tissue defect into ethmoidal region. [7] MRI provides better soft tissue resolution and can distinguish between other intranasal masses like inflammation, ectopic brain tissue or neoplasm without intracranial extension. [2, 8] However, it does not demonstrate the bony defect as well as a CT scan.

Treatment of any meningoencephalocoele is mainly with surgery. Khan and Salehuddin reported an interesting case of a large intranasal meningoencephalocele which was successfully treated with excision and closure of the cribriform plate defect with tissue and fibrin glue. [9] Endoscopic closures of anterior skull base CSF leaks are now a recognised treatment of choice. [10-12] Wormald and McDonogh reported an endoscopic 'bath plug' repair using fat to close the cranial defects and reinforced with free mucosal graft and fibrin glue. [12] In our case, we performed an endoscopic excision and closure of the CSF fistula using a septal cartilage graft and tissue glue.

In conclusion, we report an interesting case of post-traumatic intranasal meningoencephalocoele that was successfully managed with endoscopic excision and closure of the fistula with cartilage graft and tissue glue.

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