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CASE REPORT

Giant encephaloceles in adults: case report literature review

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Abstract

Encephalocele is herniation of cranial contents through a skull bony defect. Its contents may include the meninges (meningocele), meninges and brain (meningoencephalocele). It is usually seen in pediatric age group. The type of encephalocele may be classified as occipital, parietal, basal, and sincipital or frontoethmoidal. Encephalocele is mainly congenital problem with the overall incidence about 0.8–4/10,000 live births. Although, many cases of encephalocele were reported in adults' population either due to traumatic, iatrogenic or spontaneous causes. Cerebrospinal fluid (CSF) rhinorrhea, nasal obstruction, smell loss or recurrent meningitis is the most common symptoms. Here we present two cases of trans ethmoidal meningoencephaloceles managed with endoscopic approach.

Key words: Encephalocele, cranial, meningocele, frontoethmoidal, Cerebrospinal fluid

1 | CASE 1

A 25-year-old male patient presented with a history of rhinorrhea, right-sided nasal obstruction and an episode of meningitis two months prior to presentation, which began after a diving activity. Examination revealed a large reddish-bluish mass filling the right nasal cavity. Computed tomography (CT) scan showed a homogeneous opacification of 43 x 31.5 x 41mm with a right cribriform defect. T2 Magnetic resonance imaging (MRI) reported a hyperintense mass surrounded by cerebrospinal fluid (Figure 1). Based on nasal and imaging findings, an endoscopic endonasal approach to skull base reconstruction was planned. The endonasal mass was cauterized and removed until the skull base defect was identified. Reconstruction was performed with a multilayer technique with two layers of fascia lata inlay and a pedicled middle turbinate flap overlay. Histological examination revealed sinonasal mucosa with an underlying component of well-differentiated cerebral meningocortical tissue. Long-term follow-up showed no evidence of recurrence.

2 | CASE 2

A 23-year-old male patient presented with a history of rhinorrhea, left nasal obstruction, headache and an episode of meningitis one month prior to presentation. Nasal endoscopy revealed a reddish pedunculated mass located anterior to the left middle turbinate. CT scan showed a 45 x 23mm mass occupying the left nasal fossa and ethmoidal cells with a left cribriform plate defect and displacement of the nasal septum to the contralateral fossa. MRI revealed a T1 hypointense and T2 hyperintense mass occupy-

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ing the left nasal fossa with a small component in apparent continuity with the left fronto-basal brain parenchyma with heterogeneous signal, and another peripheral component surrounded by a hypointense wall on T2 containing fluid with a resonance signal identical to that of CSF. After administration of intravenous contrast, peripheral enhancement of the lesion was observed (Figure 2). An endoscopic endonasal approach was performed, involving bipolar cauterization and resection of herniated tissue and skull base reconstruction with two layers of fascia lata inlay with a vascularized nasoseptal flap. Postoperative course was uneventful with no complications. Histological examination confirmed the diagnosis of meningoencephalocele. One-year follow-up revealed no evidence of recurrence.

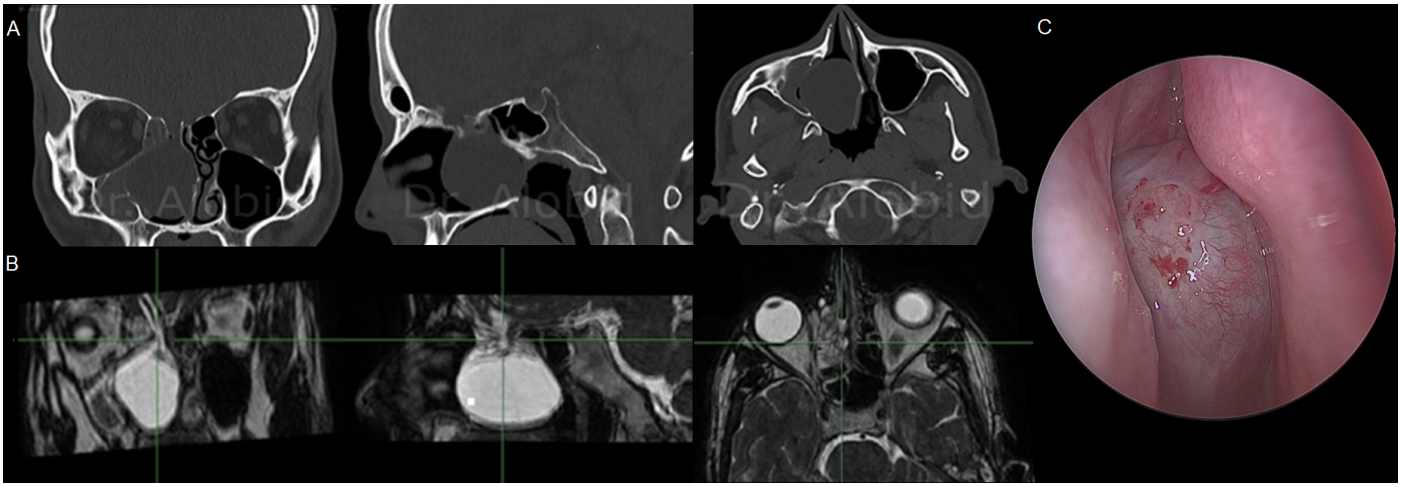

Fig. 1: Clinicalcase 1. A) CT scan coronal, sagittal and axial cuts showing a homogeneous massopacification from right cribriform defect occupying the middle and posteriorthird of the right nasal fossa. B) MRI T2-weighted coronal, sagittal and axial imagesshowing a hyperintense mass surrounded by CSF. C) Endoscopic intraoperativeview.

3 | DISCUSSION

This paper provides an educational step-by-step diagnosis and management of giant meningoencephaloceles alongside a current literature review.

Encephalocele is a condition characterized by the herniation of cranial contents through a skull base defect, which may involve the meninges (meningocele) or both the meninges and brain tissue (meningoencephalocele). The condition is primarily congenital (1) with some reports in adults, often resulting from traumatic, iatrogenic or spontaneous causes. It can be classified into several types based on their occipital, parietal, basal or frontoethmoidal location. (2)

The main symptom at presentation includes CSF leak. It has also been reported seizures, headaches, vision disturbances, respiratory and endocrinal abnormalities. (3, 4) A detailed medical history, background and the progression of symptoms are of vital importance, considering the chronicity of the clinical presentation and previous treatments.

Physical examination may reveal an intranasal soft-tissue reddish or bluish mass that can be pedunculated or sessile, sometimes pulsatile, with water-clear fluid. (2, 4, 5) It will often be located near the cribriform plate or other areas of the skull base. Craniofacial defects may be observed depending on location and extent of the lesion. (3)

Imaging studies are important for accurate diagnosis and surgical planning. CT scan often reveals a bony defect in the skull base or bone thinning. MRI provides excellent delineation of the protruded

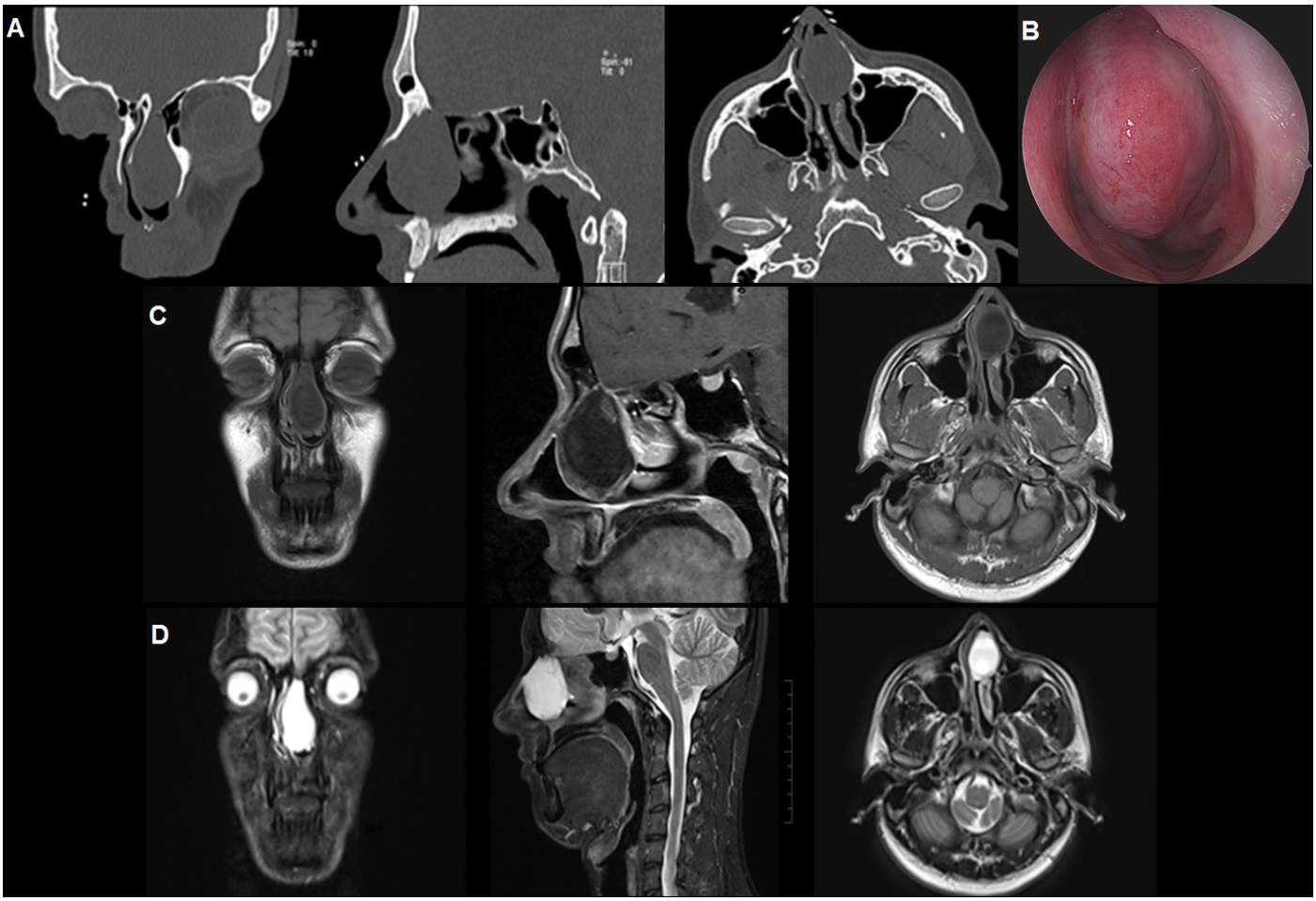

Fig. 2: Clinicalcase 2. A) CT scan coronal, sagittal and axial cuts showing a homogeneous massopacification from left cribriform defect. B) Endoscopic intraoperative view.C) MRI T1-weighted coronal, sagittal and axial images showing a hypointensemass occupying the left nasal fossa. d) MRI T2-weighted coronal, sagittal andaxial images showing hyperintense mass with peripheral enhancement aftercontrast administration.

meninges through the skull base defect and shows the continuity of the mass with intracranial structures in cases of meningoencephaloceles. Typically, a signal intensity similar to CSF will be shown; hypointense on T1 and hyperintense on T2-weighted images, with peripheral enhancement after contrast administration. (5, 6)

The treatment is solely surgical for the prevention of complications such as meningitis, intracranial abscess, and pneumocephalus as well as preservation of functional neural tissue. (1) Different approaches can be performed, depending on the location, size and the surgeon´s experience. A meta-analysis by Hegazy et al. (7) found a success rate of 90% using endoscopic approach, similar to Komotar et al. (8) results, but with lower incidence of postoperative morbidity and rate of recurrence. Open approaches, such as transcranial o transfacial approaches, are preferred for large cranial base defects with intracranial complications. (7)

Clinical findings and management of giant encephaloceles in adults described so far in the literature are summarized inTable 1 .

The choice of graft and technique is highly variable. Multilayer reconstruction is preferred in cases of defects over 1 cm to ensure CSF leak cessation and prevent recurrences. Different materials have been used, such as fascia lata, fat plug, nasoseptal flap and middle turbinate flap by the majority of authors. (5, 7, 9, 10)

Surgical follow-up and postoperative care are crucial. Recommendations include bed rest with head elevated 30 degree for 2 weeks and the use of antitussives or stool softeners.

Table 1. Literaturereview of giant encephaloceles in adults.

Author & year	Location	Size	Cause	Clinical findings	Treatment
Pollock et al. (1968)	Sphenoid	6 cm	Spontaneous	Vision problems	Not reported
Thijssen et al. (1976)	Cribriform plate	3 cm	Spontaneous	Rhinorrhea, anosmia, headache, facial hypoesthesia, meningitis	Open approach
Essene et al. (1981)	Cribriform plate	1.5 x 1.5 x 0.5cm	Iatrogenic (previous surgery)	Headache, vision problems, rhinorrhea, meningitis	Open approach
Choudhury et al. (1982)	Cribriform plate	2.5cm	Spontaneous	Recurrent meningitis, rhinorrhea	Open approach
Dempsey et al. (1988)	Posterior ethmoid	5 × 3 × 0.9cm	Spontaneous	Rhinorrhea, seizure	Open approach
Jabre et al. (2000)	Sphenoid	4 cm	Spontaneous	Bitemporal field deficit	Observation
Mukherjee et al. (2010)	Sphenoid	5.5 × 4.1 × 3.8 cm	Spontaneous	Difficulty breathing, nasal obstruction	Open approach
Albert et al. (2011)	Anterior midline skull base	3 cm	Cocaine-Induced	Altered mental status	Open approach
Sharifi et al. (2014)	Cribriform plate	1.5 cm	Iatrogenic (previous surgery)	Intermittent rhinorrhea	Endoscopic approach
Kosmidou et al. (2020)	Fronto-ethmoidal	1.6 cm	Iatrogenic (previous surgery)	Rhinorrhea, meningitis, coma	Endoscopic approach
Lam et al. (2021)	Cribriform plate	3.6 × 3.5 × 3.3 cm	Spontaneous	Headache, blurry vision, anosmia, rhinorrhea	Open approach
Chauhan et al. (2023)	Sphenoid	2.4 × 1.6 cm	Congenital	Difficulty breathing	Open approach

4 | CONCLUSION

Although meningoencephalocele is rare in adults, careful attention is required for patients presenting with longstanding symptoms such as rhinorrhea, nasal obstruction, headache or recurrent meningitis. Endoscopic endonasal approach for mass resection along with effective multilayer repair is essential for symptom improvement and to prevent complications and recurrence.

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