Giant Ethmoid Meningoencephalocele
F Aderito De Sousa, J Minaret Sandrea, V Aidmar Salas, G Fanny B. Medina

Citation

Abstract
The management of intranasal Meningoencephalocele has been historically a rare problem managed by the neurosurgeons, although their most outstanding clinical manifestations are rhinological. Nevertheless in the last years with advances in endoscopic sinus surgery (FESS) the treatment of these lesions has become easier to approach and it has diminished significantly the patient morbidity, despite its inherent morbidity. We report a case with an isolated transethmoidal meningoencephalocele, referred to us misdiagnosed as a left ethmoidal polyp with expansion to the ethmoidal roof which it was causing an important skull base defect. After multi-disciplinary evaluation (otolaryngologic, neurosurgical), a transnasal endoscopic resection of meningoencephalocele with endoscopic skull base defect reconstruction was successfully performed in this case, notwithstanding of the important size of the mass and the anterior skull base defect that this causes. They are easily misdiagnosed as nasal polyps and this can be potentially fatal after erroneous.

CASE REPORT
A 53-year-old female was referred a two years history of frequent and intense headaches with left nasal obstruction, recurrent post nasal drip and she experienced before consulting with us, a gradually anosmia and left intermittent watery rhinorrhea that worsens when she cough, sneezes and defecate. During the last 2 years, she consulted with several ENT which diagnosed rhinosinusitis that they prescribed antibiotics, decongestants, antialergics, topical and systemic steroids. One of them diagnosed left obstructive polyposis with concomitant middle skull base defect.

She has a history of obesity, AINES for chronic headaches, controlled hypertension with Captopril, and she denies antecedents of significant accidental cranial trauma, previous surgery and meningitis.

Left nasal endoscopy examination revealed an obstructive pulsatile giant mass with “polypoid” aspect that was occupying the superior, inferior and posterior portion of the left nasal cavity. The inferior portion of this mass was surrounded of scarce quantity of clear and non-viscous fluid. (Figures 1 and 2) Intraoral telescopic examination detected a large and roundish soft-tissue mass in the left choana. The left middle meatus was obstructed by the mass, and the left middle turbinate was medialized for expansible effect of the mass. The nasal septum was deviated to the right.

Figure 1
Figure 1: Left nasal endoscopy view (30Â°) revealed an obstructive pulsatile giant mass with appearance of a polypoid mass that was occupying the nasal cavity, and contact with de inferior turbinate. The clear and non-viscous fluid corresponds to cerebrospinal fluid (CSF)
Figure 2
Figure 2: Left nasal endoscopy view (30°) revealed upper level of meningoencephalocele which it’s descends among the attach of the middle turbinate and the lateral nasal wall.

Noncontrast 3-mm computed tomography (CT) of the sinuses showed a large left endonasal mass on coronal, sagital and axial projections, with an important osseous skull base defect that extends along the plate of left cribiform and the ethmoidal roof. All paranasal sinuses were clear except for some mild mucosal thickening of the floor of both maxillary antrum and left frontal sinus. (Figures 3 and 4)

Figure 3
Figure 3: Coronal thin-section CT scan shows defect in the left side of the cribiform plate and opacification of the left ethmoidal air cells and nasal cavity.

Figure 4
Figure 4: Sagital thin-section CT scan shows the meningoencephalocele dimensions and its projection toward the choana. In this projection the anterior skull base defect is shown.

T1-weighted magnetic resonance image (MRI) image showed downward left ethmoidal roof herniation High T2-weighted coronal, axial, sagital and spin-echo imaging identified a mass of liquid content (CSF-filled sac) in the left ethmoidal sinus and nasal cavity with intracranial connection. The sac is projected through the defect in the ethmoidal roof. T2 bright, nonenhancing soft tissue density in the left anterior ethmoid, which appears to be contiguous with brain parenchyma on some but not all the sequences, which are again limited by motion artifact. Thin-section high-resolution MRI was not obtained to delineate the location of the CSF fistula. There was no evidence of significant left frontal lobe herniation through the skull base defect. (Figures 5 and 6)

The patient was subsequently evaluated by a interdisciplinary team represented by neurosurgery and internal medicine. The patient was given a detailed explanation of her condition and different surgical alternatives (neurosurgical or rhinological or combined), including response, complications, and obtained written informed consents from for transnasal endoscopic approach whit endoscopic or combined repair of skull base defect reconstruction (transnasal endoscopic repair and transcranial repair of the defect). The patient no refuses to participate and fulfill its controls.
Giant Ethmoid Meningoencephalocele

Figure 5
Figure 5: Sagital T1-weighted MR image (90/10) shows downward a large meningoencephalocele sac with a CSF signal intensity inside the lesion (asterisk). The left frontal sinus showed a hyperintense signal that correspond to mucosal thickening.

Figure 6
Figure 6: Sagital T2-weighted fast spin-echo MR image (4,000/90) shows the large hyperintense meningoencephalocele sac.

Figure 7
Figure 7: Pre-operative lumbar puncture. The patient is in right lateral decubitus position Image in which we observe the intermittent left rhinorrhea of CSF.

MANAGEMENT AND SURGICAL RESOLUTION
The patient was managed closely with an interdisciplinary neurosurgical support. The surgical approach scheduled was a transnasal endoscopic resection of meningoencephalocele and a endoscopic skull base defect reconstruction.

Prior to the procedure, a lumbar drain (Codman external lumbar drain Johnson&Johnson Co.) was placed by lumbar puncture. It was performed to enter the intrathecal space and withdraw 10 mL of cerebrospinal fluid CSF. The CSF was mixed with 25 mg of fluorescein (0.25 mL of injectable 10% solution, Akorn Inc. Buffalo Grove, IL) and is slowly reinjected into the intrathecal space through the lumbar drain. (Figure 7)
**Giant Ethmoid Meningoencephalocele**

**Figure 8**
Figure 8: Slowly reinjection into the intrathecal space through the lumbar drain of 10 mL of CSF, mixed with 25 mg of fluorescein (0.25 mL of injectable 10% solution, Akorn Inc. Buffalo Grove, IL)

The meningoencephalocele was resected successfully by endoscopic endonasal partial middle left turbinectomy approach with a microdebrider and fulgurated using bipolar cautery to the level of the skull base. (Figures 9, 10, 11)

**Figure 9**
Figure 9: Endoscopic view shows microdebrider resection of left meningoencephalocele. The opening lesion shows a fluorescein-stained CSF (green color) inside the meningoencephalocele 

**Figure 10**
Figure 10: Endoscopic view of bipolar coagulation of ethmoidal anterior artery in the ethmoidal roof. This artery was displaced and elongated by the mass effect of the lesion.

**Figure 11**
Figure 11: Endoscopic view (30°) of ethmoidal roof with skull base defect (2.3 x 1.8 cms) and the dural herniation with Fluoroscein staining. The frontal recess is observed superiorly (asterisk)
Figure 12

Figure 12: Endoscopic view (30°) The mucosa surrounding the skull base defect was removed to prepare the recipient bed for the graft and the temporalis fascia graft was placed custom-shaped to fit the defect in the epidural space under the bony defect (underlay technique).

DISCUSSION

Intranasal meningoencephaloceles are uncommon lesion with a herniation of the adjacent meningeal and brain substances through of a skull base defect, mainly of congenital, traumatic or spontaneous origin(1). Some meningoencephaloceles concern anterior cranial fossa and then are observed as hernias in nasal cavity or in paranasal sinus (2). The lesion's appearance may suggest the diagnosis of a nasal polyp. They are easily misdiagnosed as nasal polyps and this can be potentially fatal after erroneous (3). This may lead to an inappropriate surgical approach and serious neurological complications. For such reason it is important to have an index of suspicion of intranasal meningoencephalocele when examining adult patients with nasal polyps.

The presence of a skull base defect can lead to major complications such as cerebrospinal fluid leak, meningocele, encephalocele and meningitis (4).

Transethmoidal meningoencephaloceles usually present with CSF rhinorrhea(5). The clinical history may also include headache, nasal obstruction, clear rhinorrhea, or rarely seizures, meningitis and brain abscess.

Several imaging studies features aid in the preoperative characterization of a transethmoidal meningoencephaloceles and it subsequent management. Fine-cut CT of the anterior and middle fossa may reveal the location of the bony defect. Although CT can clearly identify bony defects with the aid of a bone algorithm, it cannot as easily distinguish herniating brain tissue from mucosal thickening, ethmoidal mucocele and ethmoidal polyp (6).

MRI provides valuable information on detecting brain tissue and dural herniation that were useful for the proper diagnosis and surgical planning. The most important feature is its ability to identify a stalk of soft tissue traversing the bone defect from the temporal lobe to the sac of the meningoencephalocele. Multiplanar imaging allows for the detection of soft-tissue continuity and provides the ability to view the gyral pattern; the characteristic signal intensity of the herniated brain makes MRI the ideal imaging method for diagnosis and surgical planning (7).

A multi-disciplinary management is recommended in the diagnosis and treatment, the surgical procedure includes a transnasal endoscopic resection with skull base repair, anterior fossa craniotomy in combination with a transnasal approach (8).
With the advent of transnasal endoscopic procedures of the paranasal sinuses and advanced skull-base procedures, the endoscopic approach of meningoencephaloceles techniques have been increased popularity. Thus the meningoencephalocele is carefully resecting with a microdebrider to the level of the skull base and it stalk is ablated slowly using bipolar cautery.

The mucosa surrounding the skull base defect is removed to prepare the recipient bed for the graft.

The choice of graft material is dictated by defect size and configuration, underlying pathophysiology, and surgeon preference (13). A multi-layer repair is preferred not only to stop the CSF leak, but also to “shore-up” the thin skull base and thereby prevent meningoencephalocele recurrence (16,17).

Autologous free or pedicled mucoperichondrial grafts, fascia, composite turbinate grafts, cartilage grafts, pericranial-galeal and bone grafts, have all been used successfully. Heterologous grafts have also been described used as subdural graft of collagen matrix (Duragen, Integra Life Sciences) and acellular dermis (AlloDerm, Lifecell Corp.) (15).

The ability to repair large skull base defects may be limited by pressure on the graft by the brain. If such opening to the subarachnoid space exceeds 2.5 cm, intranasal repair becomes more difficult (12).

We consider that the skull base defect size and herniation of brain in this case the best option to skull base repair it was to use a multi-layer grafts of thin, pliable, and well vascularized graft, as temporalis fascia and septal mucoperichondrial , to will prevent a recurrent brain herniation and a CSF leak.

A temporalis fascia graft, custom-shaped to fit the defect is placed in the epidural space under the bony defect (underlay). A second layer of septal mucoperichondrial was placed over the bony defect in the ethmoidal roof, previous removal of surrounding mucosa (s001011212617).

Historically, lumbar drainage has been used after surgery as an adjunct to surgical repair. Lumbar drains can be used intraoperatively to assist with encephalocele localization (after intrathecal administration of fluorescein) as well as to shunt CSF away from the defect, allowing easier placement of graft material. In the early post-operative period their utility lies in preventing graft-threatening spikes in intracranial pressure due to inadvertent coughing, sneezing, or emesis (s01920c).

Nevertheless, the indications for a postoperative lumbar drain have been debated and it is very controversial in the literature revision (1,2). The lumbar drain is used depending on the surgeon’s preference. In this patient’s surgical management the skull base defect size justifies the lumbar drain use. Nevertheless it has been demonstrated that the lumbar drain is not routinely necessary for successful skull base repair in smaller skull base defects (13,24).

Surgical follow-up and post operative care are always an important concern and they includes bedrest with the head of the bed elevated 30° for 1 to 2 weeks and the use of laxatives or stool softeners. In the absence of obvious infection the prevention with antibiotic is fully justified but the efficacy of prophylactic antibiotics continues to be unproven (5).

Patients are advised against coughing, sneezing, nose blowing or straining (6).

The nasal packing is commonly removed after 48 hours (mean, 64.5 hours; median, 48 hours) and the average hospital stay was 4.3 days for all patients the patient.

**CONCLUSIONS**

Otolaryngologists are increasingly being challenged to care for patients with increasingly complex problems through minimally invasive endoscopic methods. The transnasal endoscopic approach makes possible a direct approach to the meningoencephalocele and it stalk, allowing the repair of the defect skull base defect. That was very difficult with the previous approaches.

In selected cases, this approach enables the otolaryngologist to meet modern demands to treat neurosurgical conditions in otolaringological areas with the use of endoscopic techniques. The surgical approach, and the initial experience described here in, is presented in the hopes that it can aid fellow neurosurgeons in helping patients with challenging conditions as a meningoencephalocele management.

**References**

Author Information

F. Aderito De Sousa, MD
Director and Career Counselor, Unit in Endoscopic Sinus & Skull Base Surgery Endoscopic Otorhinolaryngology Unit, Instituto Médico La Floresta

J. Minaret Sandrea, MD
Senior Fellow, Otorhinolaryngology Unit, Instituto Médico La Floresta

V. Aidmar Salas, MD
Senior Fellow, Otorhinolaryngology Unit, Instituto Médico La Floresta

G. Fanny B. Medina, MD
Junior Fellow, Otorhinolaryngology Unit, Instituto Médico La Floresta