Endoscopic endonasal repair of a cerebrospinal fluid leak secondary to a meningoencephalocele using a posterior - based middle turbinate flap.

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ABSTRACT

Cerebrospinal fluid leaks are rare but remain an important differential diagnosis for patients presenting with persistent, unilateral rhinorrhoea. This case describes a middle-aged female with persistent left sided rhinorrhea. She was minimally responsive to treatment for chronic sinusitis. On re-evaluation, a cerebrospinal fluid leak secondary to a meningoencephalocele was identified. This was subsequently repaired with a pedicled, vascularized graft using an endoscopic endonasal approach. The discussion which follows reviews the management of CSF rhinorrhoea with an emphasis on the available surgical options as well as the materials used for repair.

INTRODUCTION

Unilateral rhinorrhoea infrequently presents to the Otolaryngologist. An important differential diagnosis to be considered despite its rarity is a cerebrospinal fluid (CSF) leak. Defects involving the dura mater, skull base and sinonasal mucosa can allow communication between the subarachnoid space and the nasal cavity[1]. Cerebrospinal fluid from the anterior cranial fossa can leak into the nose through defects in the cribriform plate, anterior ethmoid sinuses, frontal sinuses or posterior ethmoid sinuses.[2] Defects within the middle and posterior cranial fossae are less common and involve the sphenoid bones, temporal bones and inferior clivus.[2] Cerebrospinal fluid alone may escape through the defect but in some cases, additional tissue may also protrude.

This case describes a middle-aged female who developed spontaneous left-sided rhinorrhea for two years. She was treated for chronic sinusitis with minimal improvement in symptoms. On re-evaluation, nasal endoscopic examination and supporting radiological investigations identified a cerebrospinal fluid leak secondary to a meningoencephalocele. Almost all cerebrospinal fluid leaks in Trinidad and Tobago have been repaired via open or combined techniques. This case represents one that was successfully managed using a purely endoscopic endonasal approach using a pedicled, vascularized graft (middle turbinate) instead of non-vascularized or synthetic material. The following discussion will review the management of CSF rhinorrhea with an emphasis on the current surgical options available for repair.

CASE REPORT

A 59-year-old Indo-Trinidadian female, presented to the Otolaryngology clinic of the San Fernando General Hospital complaining of a left-sided colorless, nasal discharge for two years. This started after an episode of severe headache and fever. There were no reports of excessive sneezing, anosmia, postnasal drip, nasal pruritus or red and itchy eyes. She gave no history of prior head trauma, sinonasal or skull base surgery. The patient was treated for one episode of meningitis in the past. In addition, she had sought medical attention for a similar headache 18 years ago which had resolved with analgesia.

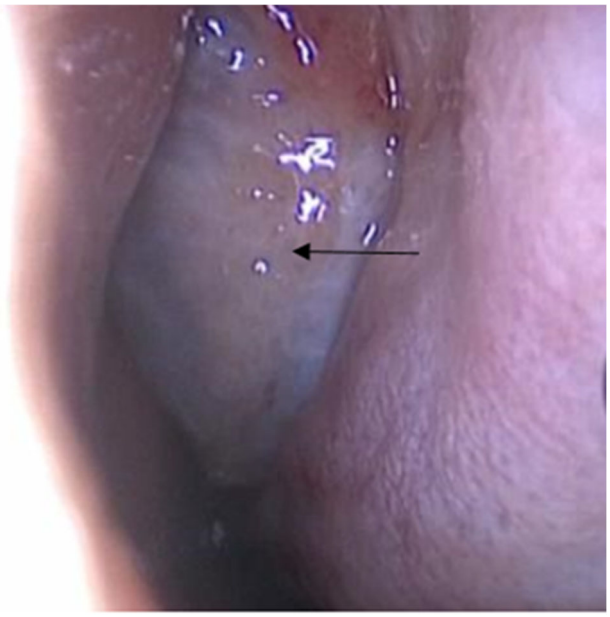


Figure 1: Endoscopic view of left superior meatus

The patient was initially treated for chronic sinusitis with nasal steroid sprays. However, there was little improvement. Repeat flexible nasal endoscopy identified a pale mass above the left superior turbinate. It was pulsatile with no visible surface ulcerations or dilated vessels (Figure 1, black arrow). Active leaking was not identified. A CT scan of the brain, paranasal sinuses and nose demonstrated a defect in the left cribriform plate (Figure 2, black arrow). A 2 cm x 1.6 cm low density, soft tissue mass was imaged in the left frontal sinus which extended into the left anterior nasal cavity. A meningocele was suspected and an MRI scan of the brain and paranasal sinuses was subsequently requested. Part of the brain and meninges were observed to protrude through a bony defect in the skull base into the upper part of the left nasal cavity (Figure 3a black arrow, Figure 3b white arrow). A meningoencephalocele was confirmed and the patient was listed for surgical repair.

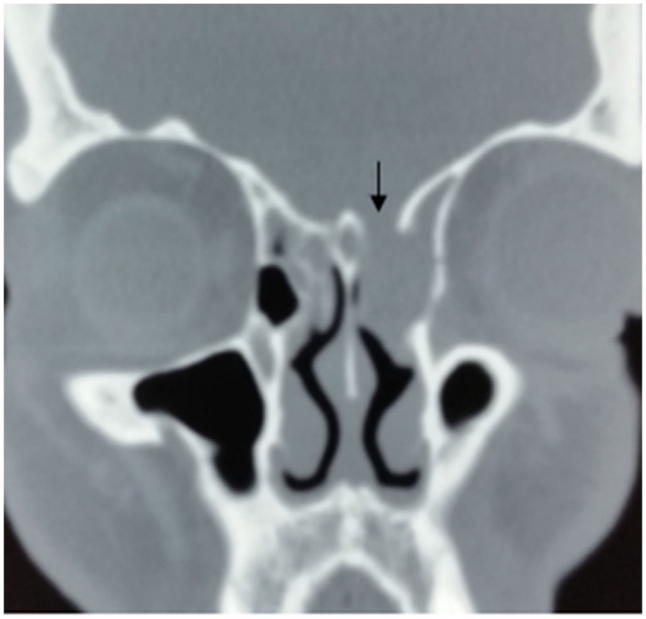


Figure 2: CT scan of the head (coronal view)

At surgery, a 30° rigid endoscope was inserted into the left nostril to visualize the meningoencephalocele. The nasal component was resected using a microdebrider.

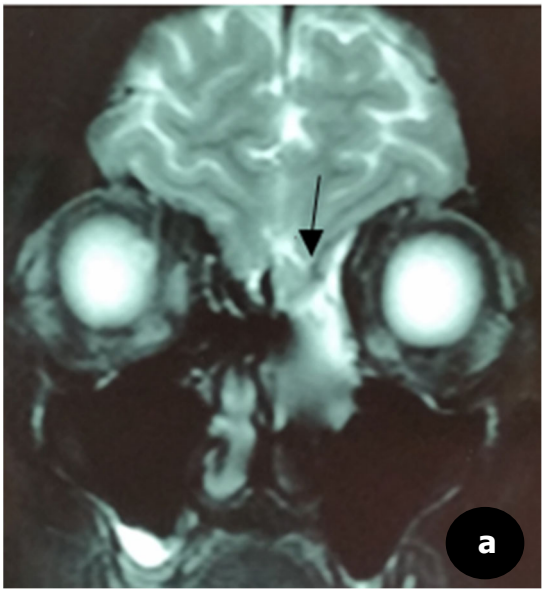


Figure 3a: MRI (Coronal T2) of the head

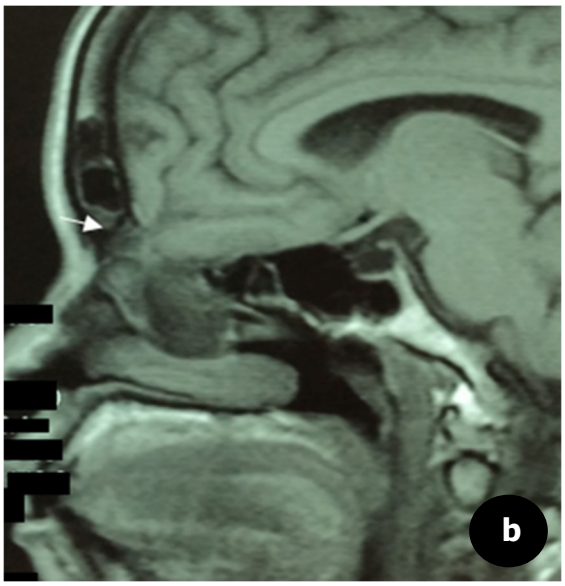


Figure 3b: MRI (**S**agittal T2) of the head

Other angled endoscopes were then advanced through the left frontal recess to enter the left frontal sinus. A bony defect was identified in the posterior wall of the sinus (Figure 4a). The stalk of the meningoencephalocele was cauterized and the mucosa surrounding the bony defect was debrided. A posteriorly-based middle turbinate flap consisting of mucosa and conchal bone was then used as a vascularized overlay graft (Figure 4b). Gelatin sponge was used to keep the flap in place and the repair was further bolstered with iodoform gauze.

After surgery, the patient was kept on intravenous ceftriaxone, stool softeners and bed rest with the head of the bed elevated at 30° for a total of ten days. At the end of this period, the iodoform gauze was removed and the patient was discharged. Nasal endoscopic examination performed at intervals of two weeks (Figure 5a) and then one month (Figure 5b, black arrow) in the clinic confirmed that the defect was completely sealed. The patient had an uneventful recovery with complete resolution of symptoms. She remains symptom free at one year post endoscopic endonasal repair.

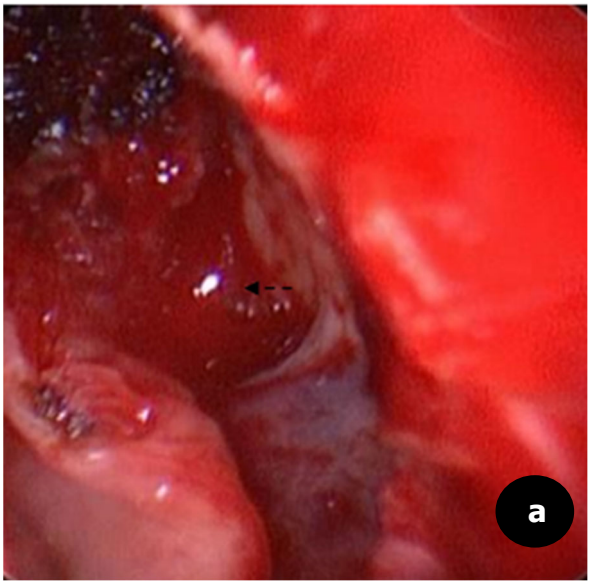


Figure 4a. Defect in posterior frontal sinus.

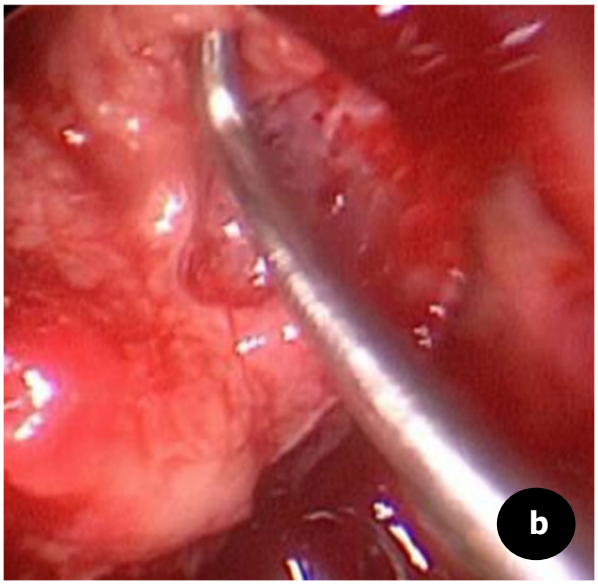


Figure 4b: Middle turbinate flap rotated over defect

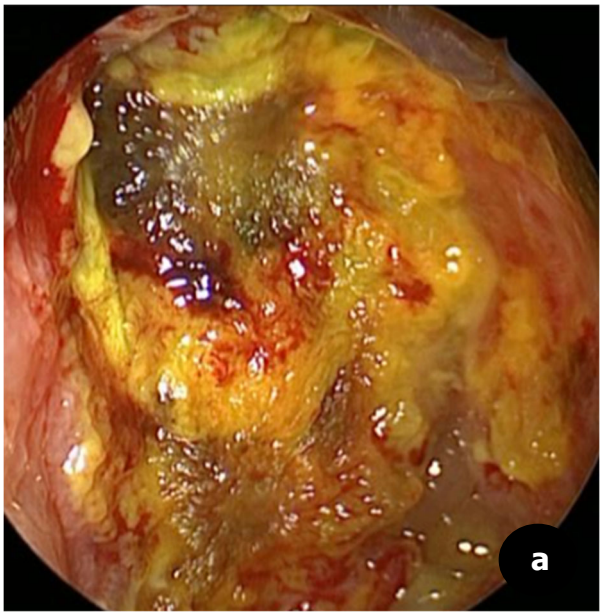
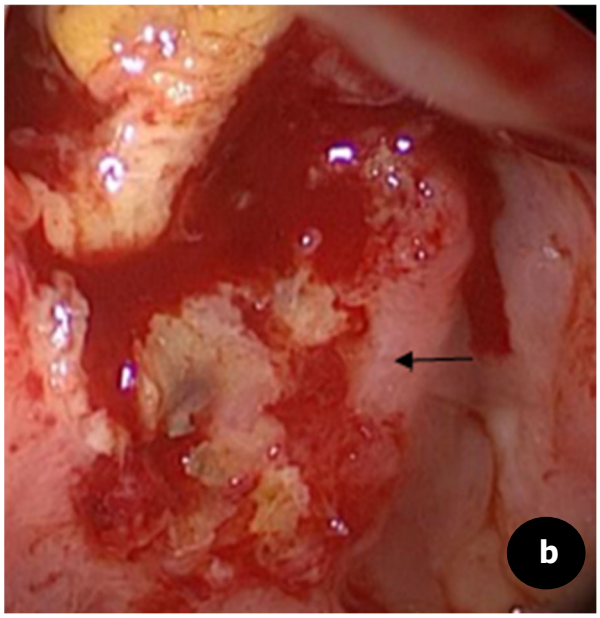


Figure 5a: Surgical site after 2 weeks



5b: Surgical site after 1 month

DISCUSSION

A meningoencephalocele occurs when CSF, meninges and brain tissue herniate through a defect in the skull base.[1,3] It is a rare cause of CSF rhinorrhea and has an incidence that ranges from 0.008% to 0.03%.[4] Meningoencephaloceles are usually congenital but a few acquired cases have been reported.[5] The patient in this case report had a history of meningitis and was also suspected of having a second meningitic episode 18 years before presentation. The absence of antecedent head trauma or skull base surgery suggests a congenital lesion. Radiological imaging confirmed the diagnosis of a left meningoencephalocele. A meningocele is present when only a small collection of CSF surrounded by meninges protrudes through a skull base defect. An encephalocele occurs when brain matter herniates through the bony defect. Even rarer than these are the encephalocystoceles (brain and fluid filled parts of the ventricles) and encephalocystomeningoceles (large parts of brain, ventricles, large volume of CSF and meninges).[1,3] If the defect is not repaired, serious sequelae may occur such as recurrent meningitis, subdural abscesses, brain abscesses and even death.[6,7]

The treatment of CSF rhinorrhea in general is influenced by the type (traumatic or non-traumatic), volume (high or low flow), size, location and number of sites involved.[8] In cases of CSF leaks without cranial tissue protrusion, most can be managed conservatively.[9] The use of antibiotics in this group is controversial but there seems to be no overall benefit with its use.[10] Acetazolamide is used to reduce intracranial pressure in leaks caused by benign intracranial hypertension.[11] In addition, a lumbar drain can be placed for one week in moderate flow CSF leaks in order to decrease the intracranial pressure and facilitate healing.[8,9] Within two weeks, approximately 90% of CSF leaks resolve. High flow leaks and defects that are >1.5 cm however are unlikely to respond to conservative management and surgical intervention is usually required.[12,13]

The aim of surgery is to repair the defect by creating a watertight closure. In cases where cranial tissue prolapses through the defect as was the case with our patient, surgical repair must be performed. Single or multiple layer repair can be done using various non-vascularized materials such as fat, fascia, mucoperichondrium, cartilage, bone or acellular dermis.[14,15] Synthetic materials inclusive of gelfoam, fibrin glue and cellulose can also be utilized. Intranasal and extranasal vascularized grafts/flaps can be used to reinforce the repair. Intranasal rotational mucosal flaps include the Hadad-Bassagasteguy vascularized pedicled nasoseptal flap, inferior turbinate flap, middle turbinate flap and lateral nasal wall flap. In our patient, a posteriorly-based middle turbinate flap was used to repair the defect and gelfoam was also utilized to keep the flap in place. The repair was further bolstered with iodoform gauze for a period of ten days. Extranasal pedicled flaps such as the transpterygoid temporoparietal fascial flap, buccinator flap, palatal flap, occipital flap and the endoscopic-assisted pericranial flap can be used but these require osteotomies or tunneling for correct placement.[12,16] Free flaps can also be used for repair. Tissue sealant or other synthetic materials can be placed at the tissue edges to further stabilize the repair. There is no evidence that specific grafts or flaps are better than others once the leak has been successfully repaired. Larger defects and more complex repairs however may combine several layers in an underlay, inlay or overlay pattern.

The introduction of endoscopic sinus surgery in the 1980s revolutionized the way CSF leaks were approached. Endoscopic endonasal surgery has become the preferred method for repair. It is indicated for defects that are less than 1.5 cm in size as well as those that involve the cribriform plate, ethmoid, sphenoid and frontal sinuses. Endoscopic techniques can access ethmoid and sphenoid sinus leaks and the posterior wall of the frontal sinus via an osteoplastic frontal sinusotomy. Newer techniques performed under endoscopic guidance include the direct tension free suturing of the dura mater and dural laser welding (duraplasty).[17,18] Endoscopic CSF repairs have rates of resolution as high as 90 % on first attempt with minimal complications.[9] Defects >1.5 cm, leaks in the superolateral aspect of the frontal sinus and in the lateral sphenoid sinus may present a challenge to repair endoscopically and an open approach is preferred in these instances.[12,13]

CSF leak repair can also be performed extracranially (combined endoscopic and open) or transcranially (open alone). Extracranial approaches such as external ethmoidectomy and frontal sinusotomy attempt to balance the morbidity from the technique of skull base exposure with successful repair. Limited external access is needed for entry into the sinuses and subsequent endoscopic skull

base repair. Reported success ranges from 86 – 97 %.[19,20] Transcranial and extended open cranial repairs on the other hand require a bifrontal craniotomy to access the skull base. This method provides the best exposure of the skull base but has the highest complication rates and the poorest cosmetic outcomes. Additionally, it is successful in only 73 % of cases.[21] Thus, it is reserved for cases with co -existing intracranial tumours, recurrent CSF leaks, multiple leaks and failure of other surgical methods.[11]

CONCLUSION

A high degree of suspicion for a cerebrospinal fluid leak should be maintained in the presence of unilateral rhinorrhoea. Most leaks involving defects <1.5cm with low flow rates and/or lack protrusion of cranial tissues are managed conservatively. The use of antibiotics is controversial and has not been proven to be beneficial to overall outcome. Additionally, acetazolamide is utilized in the treatment of leaks due to benign intracranial hypertension. Lumbar drains can be temporarily placed for moderate flow CSF leaks. The patients who fail conservative management as well as those with protrusion of cranial tissue through the defect require surgical correction.

Three types of surgical approaches – endoscopic, extracranial (combined endoscopic and open) and transcranial (open) are described for CSF leak repair. The materials used include synthetic materials, non-vascularized biologic tissues, pedicled flaps as well as free flaps. The transnasal endoscopic approach is best suited for defects < 1.5 cm or those involving the cribriform plate, ethmoid, sphenoid and frontal sinuses. It has the highest success rates, lowest morbidity and best cosmetic outcomes for CSF leak repair. The extracranial or combined approach attempts to balance the morbidity from the technique of skull base exposure with successful repair. Transcranial and extended open cranial repairs provide the best exposure of the skull base but has the highest complication rates and the poorest cosmetic outcomes. Therefore, it is reserved for cases with co-existing intracranial tumours, recurrent CSF leaks and multiple leaks. Extracranial and transcranial approaches can also be offered to patients who fail initial endoscopic treatment.

Ethical approval: not applicable

Funding: none

Patient consent: obtained

Conflicting interests: none to declare

Author contributions: Dr. Jugmohansingh and Dr. Peng wrote the manuscript. Dr. Clarke provided the pictures and imaging reports for the case.

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