

Sphenoidal Meningo-Encephalocele Revealed by Cerebrospinal Rhinorrhea: A Case Report and Literature's Review

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Abstract

Introduction: The meningo-encephalocele of the sphenoidal sinus is a rare lesion, it is a pathology that results from cranial trauma, iatrogenic causes or skull base'serosion. Spontaneous entities are rare and most often result from an increase in intracranial pressure. We report through this case the suggestive clinical symptoms, radiological features, as well as therapeutic approaches.

Case report: a 72-year-old male patient, without any pathological history in particular no traumatic one, was admitted to our ENT department for 4 years' history of a left unilateral clear rhinorrhea with ipsilateral hemicranial pain, complicated by two episodes of medically treated meningitis. The beta transferrin test of the fluid coming out from the nostril was positive, confirming the cerebrospinal rhinorrhea. Magnetic resonance imaging showed the presence of fluid formation in the left sphenoidal sinus communicating with the subarachnoid spaces through a bone defect of sphenoidal sinuslateral wall. Under general anesthesia, nasal endoscopy approach was performed. An ethmoidectomy followed by wide middle antrostomy was done on the side of the herniation. Then, the sphenotomy was carried out in order to visualize the defect present at the level of the lateral wall of the sphenoid sinus that was filled by abdominal fat and fascia latta muscle's fascia. The post operative's follow-up was simple without any abnormalities.

Conclusion: The meningo-encephalocele of the lateral wall of sphenoid sinus are uncommon but potentially life-threatening diseases. They can be an incidental finding or may be symptomatic, presenting multiple clinical manifestations. The ideal surgical approach for trans-sphenoidal encephalocele is still not clear due to lack of adequate experience in the literature.

Keywords: Meningo-Encephalocele; Sphenoidal Sinus; Rhinorrhea; MRI; Endoscopic Surgery

Introduction

Meningo-encephalocele is an entity of encephalocele, it's an abnormal sac filled with cerebrospinal fluid, brain tissue and meninges and extending through a bone defect in the skull base.

The meningo-encephalocele of the sphenoidal sinus is a rare lesion, involving different specialties including otorhinolaryngology surgery and neurosurgery; it is a pathology that results from cranial trauma, iatrogenic causes or skull base'serosion by an inflammatory or tumoral lesions. spontaneous entities are rare and most often result from an increase in intracranial pressure. We report through this case the suggestive clinical symptoms, radiological features, as well as the therapeutic approaches.

Clinical case

A 72-year-old male patient, without any pathological history in particular no traumatic one, was admitted to our ENT department for 4 years' history of a left unilateral clear rhinorrhea with ipsilateral hemicranial pain, complicated by two episodes of medically treated meningitis.

A complete neurological and ear nose throat examination; otoscopy as well as rhinoscopy; haven't found any abnormalities, notably no visible mass, nor swelling of cavum's roof.

The beta transferrin test of the fluid coming out from the nostril was positive, confirming the cerebrospinal rhinorrhea.

Magnetic resonance imaging (MRI) showed the presence of fluid formation in the left sphenoidal sinus in T1 hyposignal and T2 hypersignal measuring 18* 14mm communicating with the subarachnoid spaces through a bone defect measuring 5.2 mm of sphenoidal sinuslateral wall. We concluded to a meningo-encephalocele of sphenoidal sinus lateral wall.

Under general anesthesia after oral intubation, a nasal endoscopy approach was performed. An ethmoidectomy followed by wide middle antrostomy was done on the side of the herniation. Then, the sphenotomy was carried out in order to visualize the defect present at the level of the lateral wall of the sphenoid sinus, that was filled by abdominal fat and fascia latta muscle's fascia. Afterward, the patient was placed on intra venous antibiotics at meningeal doses. He was discharged after 5 days. The post operative's follow-up was simple without any abnormalities.

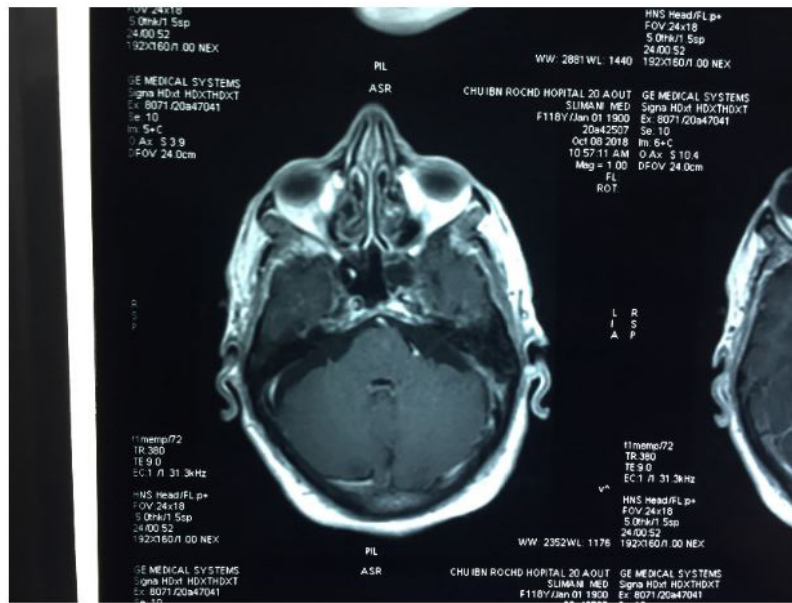


Figure 1: MRI image showing meningo-encephalocele of sphenoidal sinus lateral wall

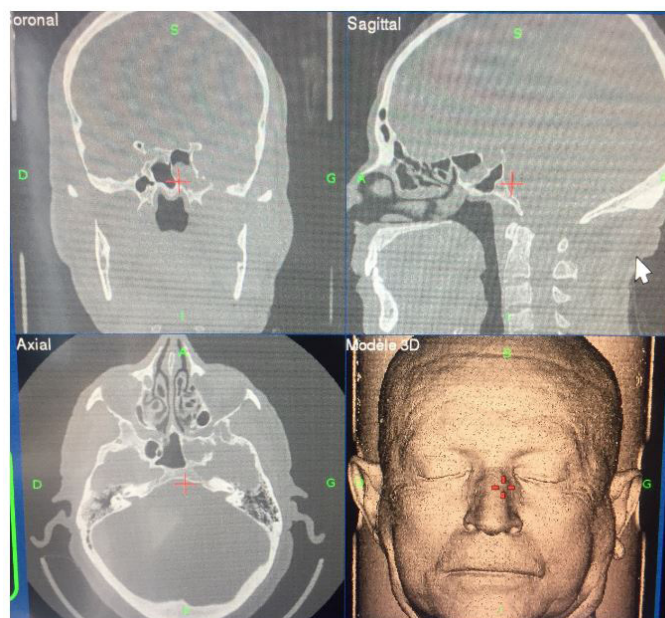


Figure 2: Intra-operative image guided navigation showing the meningo-encephalocele

Discussion

SME is a rare entity of encephalomeningocele. Spontaneous forms are usually attributed to a congenital anomaly at the lateral craniopharyngeal canal, also known as Sternberg canal [1], caused by the incomplete fusion of deferent sphenoid bone parts; At birth the embryonic portions of sphenoid bones fuse, leaving some cartilaginous portion corresponding to the future lateral wall of sphenoid sinus normally, The ossification of this cartilage occurs in the neonatal time. If fusion fails, a bony gap, located in the posterior part of sphenoid sinus and lateral to V2, called Sternberg canal or lateral craniopharyngeal canal [2-4].

The location of the defect is specific in the parasellar area, in the lateral recess of the sphenoidal sinus, laterally to V2 course [5]. From here the herniated tissue protrudes downward into the lateral recess of the sphenoid sinus and can involve also the ethmoidal sinuses or the pterygopalatine or infratemporal fossae [6,7]. Another alternative suggestion is that the defect is secondary to the cumulative effect of CSF pressure on naturally occurring defects in the floor of middle cranial fossa leading in a fistulous tract [8,9]. Considering the presence of empty sella and recurrence of rhinorrhea after proper package of the sella, the last theory can explain development of the anomaly in our second case.

The incidence of congenital encephalocele is approximately 1 in 3000-5000 live births [10]. Trans-sphenoidal encephaloceles comprise less than 5% of all basal meningo-encephaloceles and has an estimated incidence of 1 in 700000 live births [11-13].

Cerebrospinal fluid rhinorrhea is the most common presentation of SME of the sphenoid wing with lateral sphenoid sinus extension. Other presentations include headache, recurrent meningitis, seizures, and cranial nerve impairment. It can also be diagnosed as an incidental finding.

MRI is essential in diagnosis and evaluation of trans-sphenoidal encephalocele to confirm the extent of lesion, the possible associated abnormalities and to plan for the safest approach. MR angiography is helpful to evaluate intracranial vasculature. [14] MRI with contrast material injection can evaluate the content of cyst. CT scan can visualize the bone defect in the skull base.

Classification of sphenoid wing encephaloceles is based on the location of the bony defect connecting the subarachnoid space to the extracranial compartment and the extension of the meningoencephalocele itself [15,16]. These anatomical classification schemes can potentially guide the surgical approach. Traditionally, an open transcranial, an infratemporal, or a transfacial approach has been required to treat these lesions successfully. These approaches are associated with significant morbidity mainly caused by brain retraction and the extensive osteotomies needed to directly visualize the defect. The endoscopic approach is a less invasive technique with the advantages of avoiding brain retraction and large skin incisions. Initial reports of treating these lesions transnasally suggested a higher failure rate [17,18]. The poor outcomes were due to the inadequate visualization of defects lateral in the lateral recess of the sphenoid sinus. However, more precise appreciation of the surgical anatomy of the nasal sinuses [19,20] and improved endoscopic illumination and instrumentation, have led to greater use of endoscopic transnasal approaches for SME of the sphenoid wing. Recent reports recommend the transpterygoid route to treat these lateral lesions [21].

The indications for treatment and choice of surgical approach for transsphenoidal encephaloceles remain controversial. The main indications for intervention are: Obstruction of respiratory pathway, repeated meningitis, rhinorrhea, and progressive visual defect attributable to the lesion [22]. Treatment methods for meningoencephaloceles consist of a conservative approach and surgery. The objective of surgery for a sphenoid sinus meningoencephalocele is complete extracranial–intracranial blockade. The surgery can be performed by a transcranial, lateral rhinotomy, sublabial transseptal (Hardy's method) or endonasal approach [23,24].

Faulkner *et al.* in 2010 had reported the use of endoscopic endonasal treatment in a meningo-encephalocele causing refractory epilepsy [25]. These Authors demonstrated that a limited resection of the herniated brain through a ventral route could be enough to manage the associated epileptic seizure. Thus, they conclude that even in presence of epilepsy the endoscopic endonasal approach can be a valid and minimally invasive alternative to conventional transcranial surgery. Beyond its good tolerability, low complications rate and fast recovery time, it avoids the infrequent but serious complications related with the resective surgery on the temporal lobe [25-28].

Otherwise to Repair the persistent CSF leakage is the major indication for surgery in intrasphenoidal encephalocele. Transsphenoidal or transethmoidal approaches must be decided individually depending upon the exact location of the defect in skull base diagnosed by the precise preoperative imaging and the experience of the surgeon.

Conclusion

The meningocele and meningo-encephalocele of the lateral wall of sphenoid sinus are uncommon but potentially life-threatening diseases. They can be an incidental finding or may be symptomatic, presenting multiple clinical manifestations: mainly CSF leak and meningitis, more rarely seizures. The surgical approach must be tailored according to the individual patient's age, anatomical characteristics and associated anomalies [29]. The ideal surgical approach for trans-sphenoidal encephalocele is still not clear due to lack of adequate experience in the literature.

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