A SINGULAR CASE OF FRONTAL MENINGOENCEPHALOCELE ASSOCIATED A WITH LHERMITTE-DUCLOS SYNDROME

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ABSTRACT

Meningoencephalocele (MEC) refers to a herniation of intracranial contents, through a defect of the skull base into the nasal or paranasal cavities. It is a rare entity, typically congenital, originating from a defect during or immediately after the neurulation process. Less frequently it can be secondary, especially in adults as in the present case, resulting from traumatic injuries, surgery, hydrocephalus, and chronic sinusitis. Diagnosis is based on clinical evaluation, CT (computed tomography) scans and MRI (magnetic resonance imaging). B2-transferrin dosage may be useful in unclear cases. The clinical case herein presented refers to an unusual presentation of a MEC in the frontal sinus associated with Lhermitte-Duclos Syndrome, a condition which led over time to hydrocephalus and intracranial hypertension, well-known predisposing factors for MEC genesis. A combined transnasal endoscopic and external approach was performed to completely dominate the herniation and adequately reconstruct the bony defect with a gasket seal technique. To date, the patient has no recurrence of MEC.

INTRODUCTION

Meningencephalocele is a herniation of brain tissue into the nasal or paranasal cavities. [1] It can be congenital or acquired after certain insults to the brain such as traumas, surgery, hydrocephalus, and chronic sinusitis. [1-2] Diagnosis is achieved through clinical assessment, CT scans and MRI. Beta-2-transferrin can guide the diagnostic process, since it is a form of the transferrin protein normally present in cerebrospinal fluid (CSF), but not usually found in blood, nasal secretions, or other body fluids. [3] Surgical removal of non-vital herniated brain and bony defect reconstruction are mandatory to allow resolution of symptoms and to avoid brain superinfection. [2] The most frequent locations of meningoencephalocele and CSF-leak include cribriform plate and sphenoid sinus. Frontal sinus is a rare

localization of MEC and requires adequate and specific surgical management.

CLINICAL CASE

A 43-year-old man was admitted to our Department for an unilateral left limpid rhinorrhea onset about two months before.

He was also affected by a particular syndrome, the Lhermitte Duclos' disease, a condition characterized by a rare genetic disorder, often associated with Cowden's syndrome. It consists of a slowly growing benign cerebellar tumor, a dysplastic gangliocytoma. Symptoms are nonspecific and slightly progressive, due to an increasing intracranial pressure and a progressive alteration in cerebellar functions; those include headache, ataxia, tremors, visual impairment, and electroencephalogram abnormalities. For this condition he had undergone twenty years earlier a surgical resection of the neoplasm through an occipital craniotomy approach. To note, literature reports the frequent association of this syndrome with obstructive hydrocephalus, megalencephaly, syringomyelia, and skeletal anomalies. [4]

Therefore, the history of unilateral rhinorrhea and central nervous system abnormalities led to suspicion of a CSF-leak. Transnasal endoscopy showed no anatomical changes or clear signs of rhino liquorrhea. However, confirmation of the suspicion came with the dosage of B2-transferrin on the liquid collected from nasal secretions and with a CT scan which clearly demonstrated the interruption of the posterior wall of the left frontal sinus. A clear definition of the pathology was finally obtained through MRI with T2-weighted and FLAIR sequences which documented the presence and extension of the frontal sinus MEC.

In particular, FLAIR sequences, by suppressing the liquor signal, distinguished CSF from any other inflammatory signals which conversely would appear hyperintense (Figure 1).

In the present case, MEC was probably secondary to the condition of obstructive hydrocephalus, which generated a high pressure in the liquor circulation and therefore the onset of a fistula. Only one case of frontal meningoencephalocele in Lhermitte Duclos disease had been previously reported in literature, likewise, related to the presence of obstructive hydrocephalus. [4]

Ascertained that surgery is mandatory, the question is which surgical approaches should be performed. Even if the transnasal endoscopic technique allows to dominate the meningoencephalocele from the bottom, the superior-lateral control of the pathology as well as the bony defect reconstruction are not possible exclusively with that approach. At the same time, the exclusively external technique through a frontal osteoplastic flap could not manage the meningoencephalocele in its lower portion. For that reason, a combined approach was revealed to be the most appropriate strategy. After the removal of the MEC in its no longer vital portion up to his peduncle, the reconstruction took place through a first layer of pericranium, a second layer composed of modeled quadrangular cartilage introduced with a gasket seal technique, and a third layer of pericranium. The layers were stabilized with hemostatic material and fibrin glue. [5]

Post-operative control with CT scans demonstrated good surgical outcomes with no complications or evidence of leakage (Figure 2).

The last follow-up, 3 years after treatment, shows a stable surgical outcome, with no recurrence of MEC. The hyperintense signal in both MRI T2-weighted and FLAIR sequences into the frontal sinus (Figure 3) is actually compatible with inflammatory tissue. The patient does not complain any symptoms for the latter finding.

CONCLUSIONS

Even with a low frequency, frontal sinus CSF-leak may occur, especially when associated with particular conditions, such as central nervous system disorders. The best management of a bony defect in this specific district may be achieved endoscopically with less morbidity. However, in far lateral lesions or larger defects requiring a complete dominance of the frontal sinus, a combined endoscopic and external approach is required.

References

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FIGURES CAPTIONS

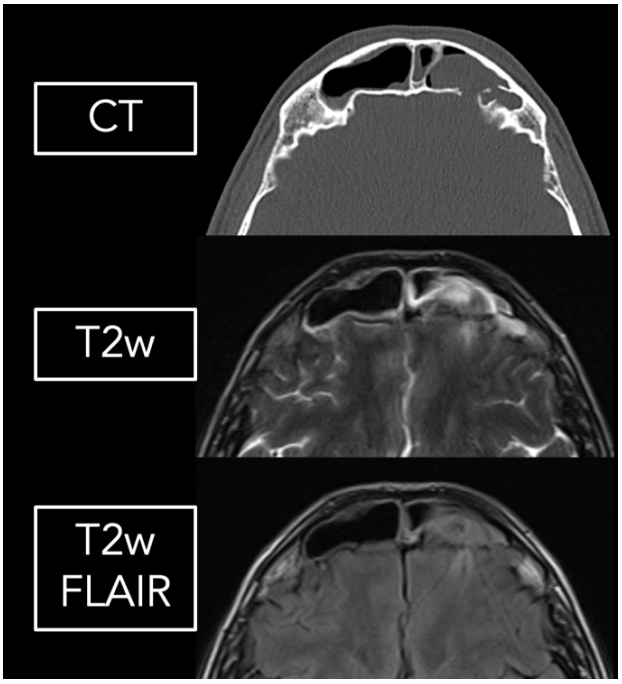

Figure 1

Preoperative CT scan showing the posterior frontal wall interruption; MRI showimg the extent and the signal characteristics of the MEC, hyperintense in T2-weighted sequences and hypointense in FLAIR sequences

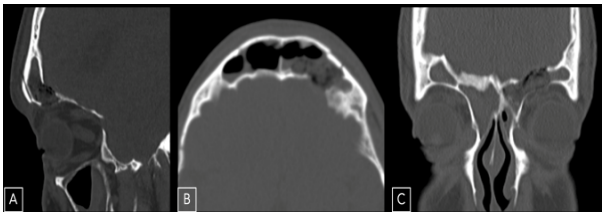

Figure 2
Postoperative CT scan, in sagittal (A) axial (B) and coronal (C) planes, performed 48 hours after surgery displaying good surgical outcomes free from complications.

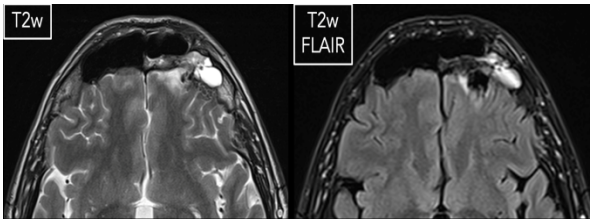

Figure 3
MRI, performed 3 years after treatment, showing no evidence of MEC relapse and a hyperintense signal in both T2-weighted and FLAIR scans, consistent with inflammatory tissue