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Case Report

Endoscopic Endonasal Repair of Transsellar-Transsphenoidal Encephalocele in an Adult Patient

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Abstract

Background:

Meningoencephalocele is an abnormal sac containing brain structures and cerebrospinal fluid; herniating through a bony defect in the skull. Their origin could be related to previous head trauma or other congenital malformations. The transsellar transsphenoidal subgroup is rarely encountered in the adult population. We report a case of transsphenoidal encephalocele that occurred in an adult patient with specific circumstances of diagnosis and preoperative findings.

Clinical presentation:

An adult male complained of hypogonadism with intermittent rhinorrhea. The radiological investigation revealed a transsellar transsphenoidal encephalocele. it was managed using the endoscopic endonasal approach with an appropriate reconstruction of the bony defect.

Conclusion:

Endocrine disorders could be a revealing scenario of transsellar encephalocele. surgical management prevents repeated episodes of meningitis and can relieve the traction on the nervous structure and improve preoperative disorders.

***Keywords**: endoscopy, encephalocele, sellar.*

BACKGROUND AND IMPORTANCE:

An encephalocele is an abnormal sac filled with brain contents and cerebrospinal fluid protruding out through a bony defect in the calvaria or the skull base. Several subgroups were described in this entity considering anatomical parameters. they can be; frontal, parietal, or occipital, and when they affect the skull base, they are grouped within one of the following entities transethmoidal, transsphenoidal, sphenoethmoidal, and sphenoorbital encephalocele[1] . the transsphenoidal subgroup is very rare and

represents 5% of the basal encephalocele and its incidence is estimated to1 in 70000 births[2].

The transsphenoidal encephalocele is commonly revealed in the pediatric population, by the presence of endocrinological and visual disorders. this clinical scenario could be related to the distension of the pituitary stalk or the optic chiasm, routinely included within the herniating sac[3]. the origin of this malformation was also reported to be a persistent cricopharyngeal canal by an incomplete ossification of the sphenoid bone[4]. In the neonatal period, the patients present clinical features of some developmental craniofacial defects as; cleft lip and hypertelorism [5].

In the adult population; these malformations are discovered within a story of endocrinological disorders such as amenorrhea or delayed puberty. copious rhinorrhea was also encountered [6,7]. Headaches and meningitis were also reported in another work as some of the revealing symptoms[8]. the pituitary dysfunction is related probably to mechanical traction on the stalk and hypothalamus or even repeated meningitis[9]. At a later stage of evolution, serious complications, such as subdural empyema were associated with encephalocele [10]. we report a case of a transsellar transsphenoidal encephalocele in an adult patient with specific revelation mode and preoperative findings.

CLINICAL PRESENTATION:

A 43 years old male with a past medical history of cranial trauma at the age of 7 years, was admitted to the Department of Endocrinology for the investigation of secondary hypogonadotropic hypogonadism. He presented progressively during 2 years before gynecomastia with an ejaculation. At the clinical exam, we noticed that he presents a diabetes insipidus. He also reported several episodes of mild rhinorrhea. His ophthalmological evaluation didn’t reveal visual disorders.

Endocrinological evaluation revealed; Testosterone :0.42ng/ml, Luteinizing Hormones (LH): 2.56m IU/ml, Prolactin:8.43ng/ml(elevated), Corstisol:336.5nmol/l, FT4: 10.75pg/ml. those findings are concomitant with a profile of a hypogonadotropic hypogonadism.

The MRI showed the aspect of transsellar transsphenoidal encephalocele, that protrudes at the level of the floor of the sphenoid sinus. the pituitary stalk had an abnormally thickened aspect herniating through the same sac. A part of the optic chiasm was also included in the malformation. The CT sinus showed the location and the size of the bony defect in the sellar floor. (figure1)

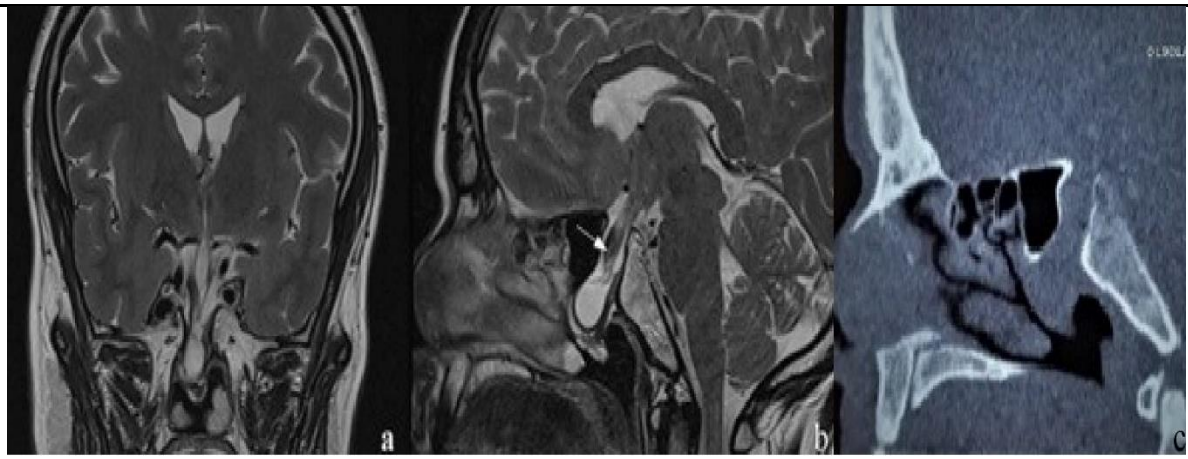

Figure 1: a- coronal section of MRI showing the importance of herniation if the sellar and part of the suprsellar contents .b- on the sagittal section , the abnormal and thickened pituitary stalk is showed by the white arrow .c- the sinus CT showed the bony limits of the defect from the clical recess to the anterior face of the sella.

The repair of the encephalocele was performed using an endoscopic endonasal approach. the sac was resected at the most distal part after being coagulated. We have performed a usual an endoscopic enodonasal binostril approach. After the sphenoidotomy , we can appreciate the bony borders extensions of the encephalocele the long of the sellar floor (image a - figure 2). The sac of was dissected 360 degrees from the bony defect that was individualized clearly. after clipping the lower extremity of the malformation we have tried first to puncture it sing a syringe to reduce tension inside the sac without getting CSF. we have coagulated and sectioned it just superior to the vascular clip. the reconstruction of the bony defect was secured with double layers of fascialata and a nasoseptal flap that was harvested in the right nostril.

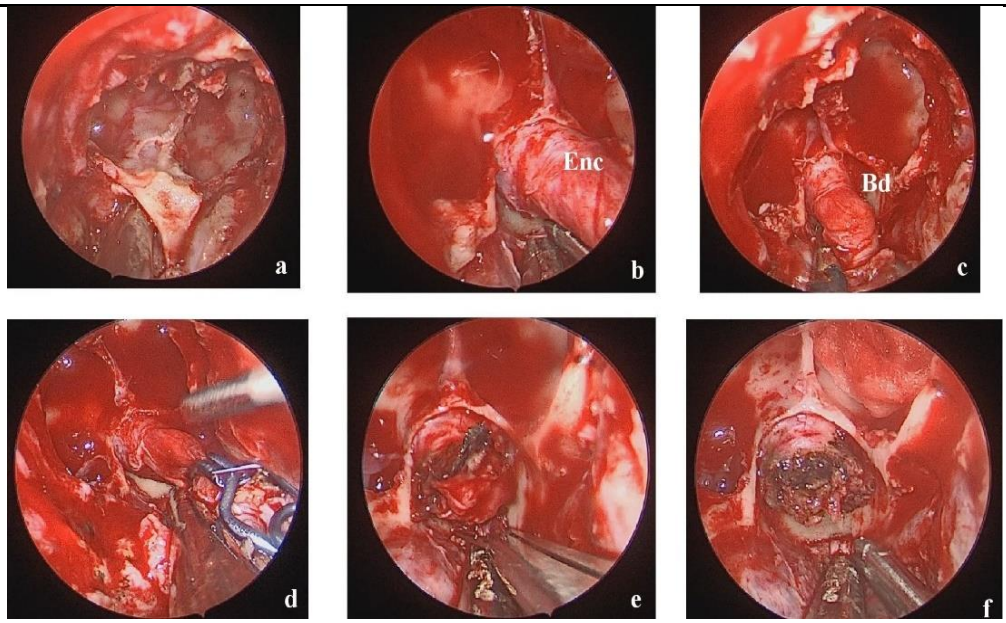

Figure 2: different important surgical steps .a- the endoscopic view of the encephalocele after the sphenoidotomy .b+c – the circumferential dissection of the sac and the definition of the bony borders of the defect .d- the most lower extremity of the sac was clipped using aneurysm clip to prevent eventual important CSF leaking after section during surgery .e+f – section just distal to the clip and the final view of the treated malformation .

The postoperative course was uneventful. we have noticed a transitory DI that was managed using desmopressin. the patient was discharged 4 days after surgery. He didn't present CSF leak.

DISCUSSION:

Encephaloceles are secondary to a bony defect through which a part of the brain contents and CSF protrudes. The transsellar form of basal encephalocele is rare however essentially encountered in the pediatric population. They are associated with some craniofacial abnormalities such as cleft lip and hypertelorism [11]. Such forms of encephalocele, remain challenging due to their contents as optic chiasm, pituitary stalk, and the anterior communicating complex [11] . Polyuria and nasal obstruction were also some of the revealing symptoms in other candidates[12]. our patient was an adult candidate and the revelation was made during the investigation of a hypogonadism. He presented also a polydipsia that was a part of a diabetes insipidus. these symptoms are probably related to the mechanical tension on the stalk and pituitary gland. The same

circumstances of diagnosis were reported by other authors[7], however, additionally, their patients had rhinorrhea which was also reported by our patient. In another work, ADH deficiency was also noticed in a patient with transsellar encephalocele [13]. Several approaches were described to manage these malformations, including transcranial and transsphenoidal routes[12,14]. We have performed a binostril endoscopic endonasal approach. the nasoseptal flap was harvested at the beginning of surgery and packed in the nasopharynx. after the sphenoidotomy, the bony defect was individualized as a bony tunnel that extends the sella to the floor of the sphenoid sinus. the bony structure was drilled and the encephalocele was circumferentially delimited than excluded distally with a clip. after coagulating; we have resected distally to avoid eloquent structures. We have used two layers of fascialata with a vascularized septal flap. after surgery, we didn't notice visual disorders. A transitory worsening of diabetes insipidus was noticed that was managed using desmopressin. some authors described a modification of the endoscopic technic, where they aspirated partially the cystic part of the encephalocele to reduce tension and the bony defect was firstly closed using titanium mesh plate [12].

The transcranial route for this form of basal encephalocele remains challenging because of the retrochiasmatic position of the structures and the sac[12], whereas the endoscopic approach is a straightforward trajectory and has better options for reconstruction.

CONCLUSION:

The transsellar encephalocele remains a challenging entity because of the highly eloquent structure that usually is present in the sac. The endoscopic route is a targeted approach with an efficient reconstruction technic that offers good healing of the dural and bony defects.

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