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Trans-septal endoscopic management of transalar transsphenoidal meningoencephalocele in a neglected adult patient with severe visual loss and pituitary dysfunction

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Congenital skull base meningoencephaloceles (MECs) are exceedingly rare lesions, occurring in approximately 1/35000 live births. Basal encephaloceles include spheno-orbital, sphenoethmoidal, sphenomaxillary, transethmoidal, and transsphenoidal lesions. Among them, the transsphenoid meningoencephalocele (TSME) are the rarest, occurring in 1/700000 live births.[1] Morning glory disk anomaly (MGDA) is described as a funnel-shaped excavation of the posterior fundus that incorporates the optic disc surrounded by an elevated annulus of chorioretinal pigment.[2] These congenital anomalies can be associated with moyamoya disease, Chiari malformation type I, agenesis of the corpus callosum, and basal encephaloceles.[2]

Transcranial versus endonasal approach, intradural or extradural handling of neurovascular content, and preservation or

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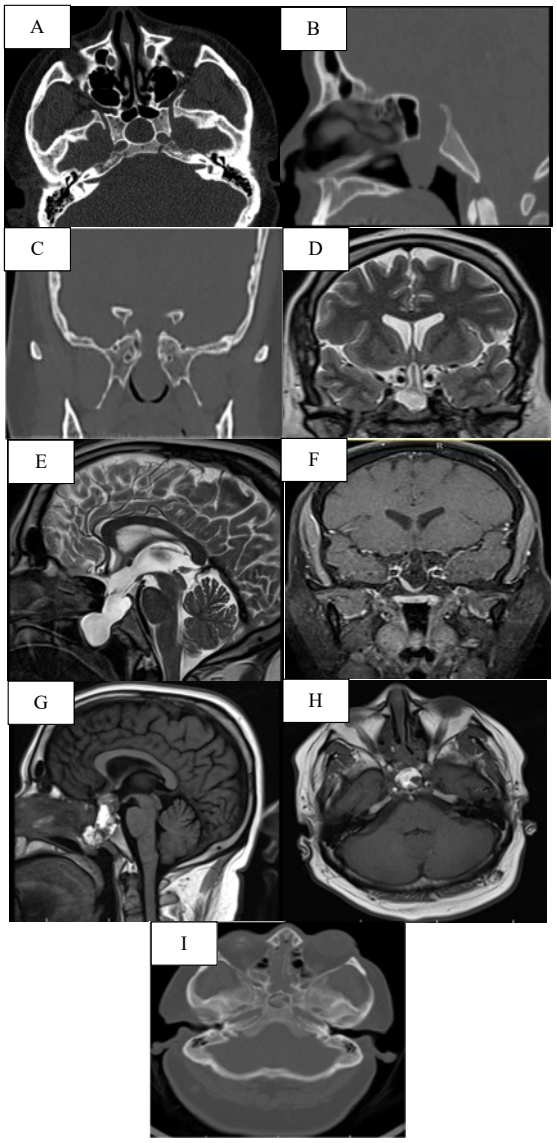
resection of the herniated material are different controversial subjects in surgical treatment of TSME. We intend to share our experience presenting this neglected case of TSME using the trans-septal endonasal endoscopic approach.

A 29-year-old man admitted with diabetes insipidus and bilateral severe visual loss. Ophthalmologic examination revealed right congenital cataract with microphthalmia and MGDA in the left eye. He was blind in the right eye since childhood and his vision dropped in the left eye during the previous one year from hand motion to light perception.

The computed tomography (CT-scan) of the paranasal sinuses showed a defect in the floor of the sella turcica (Figure 1).



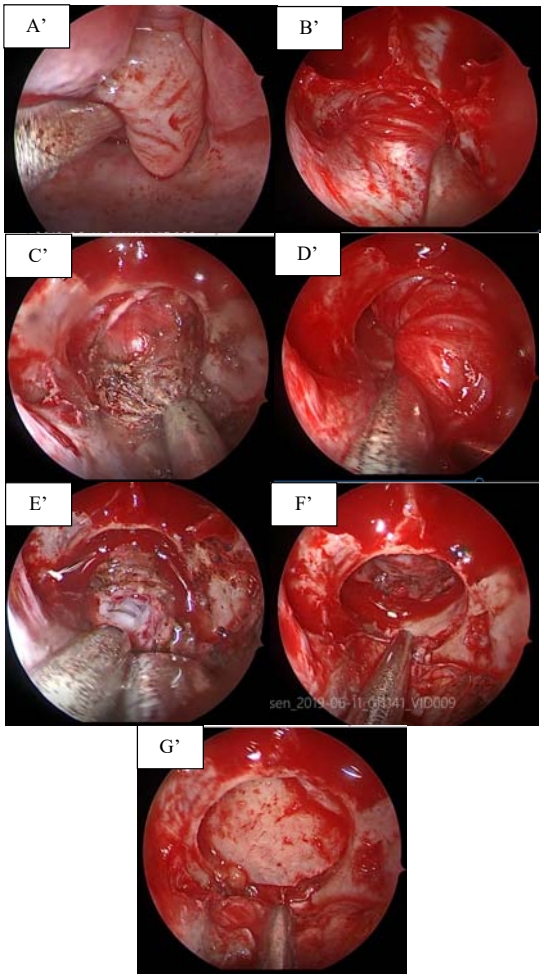


Figure 1. A, B, C: Preoperative axial, sagittal, and coronal computed tomography (CT) scan revealed a skull base defect with cortical bone formation around the hole, D and E: preoperative coronal and sagittal T2-weighted magnetic resonance imaging (MRI) presented the herniated meningoencephalocele (MEC) into the sphenoid sinus and nasopharynx. F: preoperative dynamic coronal view of pituitary gland displaced in the right side into the sphenoid sinus. G, H: postoperative sagittal and axial T1 MRI presenting the basal defect reconstruction. I: postoperative axial bone window view showing the reconstructed basal defect with nasal septum bone. A': endoscopic view of nasopharyngeal part of MEC, B' and C': central corridor exposure with subperiosteal dissection in the left and right sides. D': bipolar coagulation of MEC to shrink the cyst. E': MEC opening to drain the CSF and continuing dissection all around the cyst wall. F': pushed up MEC. G': reconstructed basal defect using fat and septal bone

Brain magnetic resonance imaging (MRI) revealed downward displacement of optic chiasm, pituitary stalk, and pituitary gland into

the sphenoid sinus resembling herniation of a MEC into the nasopharynx (Figure 1). The result of the pituitary assay was T3 = 52 ng/dl (70-204 ng/dl), T4 = 4.12 mug/dl (4.5-12.5), thyroid stimulating hormone (TSH) = 0.734 muIU/ml (0.4-4.2), follicle-stimulating hormone (FSH) = 0.3 mIU/ml (1.5-12.4), luteinizing hormone (LH) ≤ 0.1 mIU/ml (1.7-8.6), Prolactin = 9 ng/ml, Cortisol = 0.56 mug/dl (4.8-19.5), growth hormone (GH) ≤ 0.030 ng/ml, and Insulin-like growth factor 1 (IGF1) = 32.7 ng/ml (103-271).

Under general anesthesia, the central trans-septal corridor approach was used for submucosal dissection around the nasopharyngeal mass to mobilize the herniated MEC. The sac was incised to drain cerebrospinal fluid (CSF) and shrink its volume. After which, the sac could be dissected completely from the surrounding elements. These structures were pushed back into the dural defect and the whole shrunken cyst was pushed up. Abdominal fat was used to fill up the defect. A piece of the septal bone was used to reconstruct the bony defect (Figure 1).

The post-operative CSF leakage was controlled with Acetazolamide and lumbar puncture. The vision worsened in the left side but improved after two weeks back to the preoperative status. The control MRI showed the reconstruction of the basal defect (Figure 1).

Skull base MEC is a rare congenital lesion. Among these anomalies, the trans-sellar type is the rarest one. TSME can herniate through midline defect (transsellar or transplanum), via the lateral wall of the sphenoid sinus (sternberg's canal), or transalar (greater sphenoid wing).[1] Association between TSME and some eye abnormality like MGDA has been reported previously.[2] MGDA was diagnosed for our patient in another hospital six years ago without any work up for associated conditions. Skull base MEC may enlarge and cause respiratory distress, hormonal insufficiency, visual disturbances, and recurrent meningitis. The technique of surgical repair of these lesions should be tailored according to the size and location of the bone defect and the herniated sac, functional or non-functional content of MEC, and concomitant anomalies. The endoscopic endonasal approach is the safe and effective approach to repair these lesions.

Gaab recommend that conventional microsurgery is more difficult than endoscopic technique in these skull base lesions.[3] London et al. reported endoscopic repair of a basal encephalocele in a 4-year-old child using porous polyethylene plate, but the MEC recurred due to the displacement of the plate. They successfully revised the case using calvarial bone graft.[4] Zeinalizade et al. used titanium mesh to rigidly reconstruct large sphenoethmoidal defect and restore nasopharyngeal airway in cases of basal MEC.[1] Repair of MEC needs delicate technical considerations because of the close relationship between the neurovascular structures (optic pathway, circle of Willis and hypothalamo-pituitary complex) and the herniated content of TSME.

We would like to highlight two main points to be considered in the management of these lesions: 1) repair of the herniated sac and bone defect is possible, both intra or/ and extra-durally and should be performed in MEC cases in the skull base region and, 2) preservation of the function of neurovascular component after releasing and replacing them in their correct anatomical locations is necessary. Intradural release of the neurovascular components usually prevents tractional ischemic damage.

In our case, we used central trans-septal access to get to the rostrum and base of the bone defect. With this approach we were able to see around the lesion well and releasing the MEC was feasible. We decided not to open the meningeal sac and release the intradural anatomic structures because of the severe and long-term hypopituitarism and visual loss. The anatomic structures located within the sac were not manipulated to prevent further damages. Furthermore, the low flow CSF leak could be controlled more easily. Visual apparatus and pituitary gland were placed into the normal position and MEC repaired by autologous nasal septum bone without significant complication. However, visual impairment and pituitary hormone dysfunction did not improve.

Skull base MECs are exceedingly rare lesions with a challenging management. A high rate of caution to evaluate these lesions is recommended in cases with eye abnormality like MGDA. Early detection of this congenital lesion is highly important to prevent further neurological damages. Endonasal trans-septal with panoramic endoscopic view is a proper technique for the management of these midline lesions.

Conflict of Interests

The authors declare no conflict of interest in this study.

Acknowledgments

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