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Endoscopic repair of transsellar transsphenoidal meningoencephalocele; case report and review of approaches☆,☆☆

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ABSTRACT

We present an extremely rare case of transsellar transsphenoidal meningoencephalocele in a 36-year-old woman with pituitary dwarfism complaining of nasal obstruction. Imaging studies showed a bony defect in the sellar floor and sphenoid sinus with huge nasopharyngeal mass and 3rd ventricle involvement. Using endoscopic endonasal approach the sac was partially removed and the defect was reconstructed with fat and fascial graft, and buttressed with titanium mesh and septal flap. Visual field improvement was noticed post-operatively and no complication was encountered during follow-up. So, endoscopic endonasal approach with partial resection of the sac is a safe and effective treatment for this disease

Introduction

Meningoencephalocele is a herniated sac of brain structures and meninges into a defect of skull and dura. Its prevalence is estimated to be 7.5–30 cases in every 100,000 live births. Meningoencephaloceles are anatomically divided into frontal, occipital, parietal and basal types. Basal meningoencephaloceles can also be divided into transethmoidal, sphenoethmoidal, spheno-orbital and transsphenoidal types [1]. The transsphenoidal meningoencephalocele is the rarest one with the incidence of one in every 700,000 live births [2]. Subdividing this entity into midline (transplanum, transsellar), lateral (through Sternberg's canal), and trans-alar (through a defect of greater wing of sphenoid bone) meningoencephaloceles seems a practical classification.

A true transsphenoidal meningoencephalocele is the form in which sac contents pass the sphenoid sinus floor and get access to nasal cavity or nasopharynx while an intrasphenoidal meningoencephalocele is restricted to the sphenoid sinus cavity [1]. Transsphenoidal meningoencephalocele can be detected in early infancy due to their critical manifestations such as respiratory distress, feeding problems, endocrine abnormalities, and concomitant craniofacial defects [3]. But in the absence of those manifestations, the diagnosis can be delayed until the 7th decade of life [2]. Adult patients with transsphenoidal meningoencephalocele might present with spontaneous cerebrospinal fluid (CSF) leak, nasal obstruction, recurrent meningitis, and intranasal polyp. Visual disturbances are another common complaint presenting as visual field defects, decreased visual acuity, amaurosis, and amblyopia [1]. Endocrine abnormalities such as hypothyroidism, hypogonadism, hyperprolactinemia, and diabetes insipidus may also occur [1].

As far as we know, only 11 cases of true transsellar transsphenoidal meningoencephalocele (TTSME) in adults have been reported in the English literature so far [4]. In this study we describe the 12th case, a 36-year old woman with true TTSME who was treated by an endoscopic endonasal approach with partial resection of the sac. The surgical procedure was successful and the patient's recovery was excellent.

Case report

A 36-year old woman with pituitary dwarfism, diabetes insipidus and primary amenorrhea was referred to the ENT clinic due to her complaint of nasal obstruction and right eye's blurred vision that started and gradually deteriorated from two years before presentation and headache and nausea since six months ago. Nasal endoscopy revealed a large mass obstructing the posterior choana extending into the posterior part of the nasal septum. Paranasal sinus CT scan revealed a huge herniated sac with CSF isodensity, extending from sella to nasopharynx through a large osseous defect of sella turcica and the floor of the sphenoid sinus (Fig. 1). T1-weighted MRI showed a large CSF containing lesion extending from the interpeduncular space downward to extra cranial region and nasopharyngeal space with the 3rd ventricle inside. The sac was placed exactly behind the optic chiasm with a downward traction on the chiasma. There was no other skull base deformity or cerebral disorder (Fig. 2).

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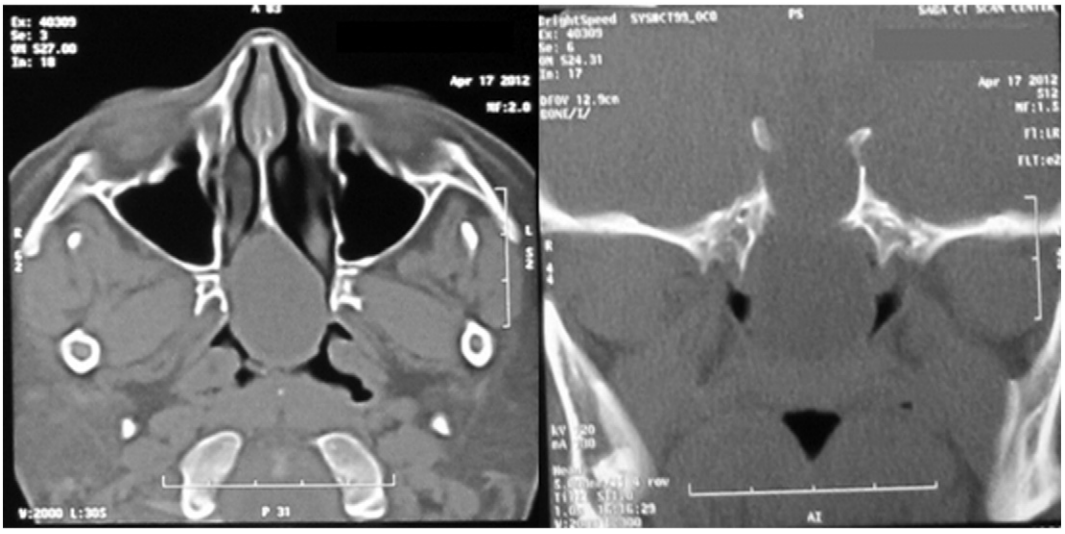

Fig. 1. Axial and coronal CTscan of paranasal sinuses showing the extension of the mass and bony defects of sellar floor and sphenoid sinus.

Ophthalmologic evaluation revealed normal pupils with normal pupillary light reaction bilaterally. The visual acuity was 10/10 in the left and 3/10 in the right eye. Fundoscopy showed a right large optic disc with total pallor (morning glory syndrome). The left eye fundoscopy only showed a pallor disc. Perimetry of the right eye showed severe generalized depression and significant localized scotoma with blind spot enlargement (Mean Defect = −14.7, Pattern Standard Deviation = 9.7). The left eye perimetry showed only mid peripheral scotoma (Mean Defect = −1.1, Pattern Standard Deviation = 2.7). The perimetry results were in accordance with right temporal hemianopia and intact central vision. Endocrine evaluation was indicative of pan-hypopituitarism and total GH deficiency. No history of CSF leak was present.

Binostril endoscopic endonasal approach was adopted as previously described [5,6]. After general anesthesia, the right middle turbine was partially removed. A right-sided septal mucosal flap was developed based on the posterior septal artery and meticulously dissected from the lesion. The septal mucosa of the left side was tightly adhered to the sac postero-inferiorly which was removed along with mucosa of the left small sphenoid sinus. The remnant of sphenoidal rostrum was observed. A transsphenoidal sac protruded from the floor of the sphenoid sinus. To reduce the size of the sac, electrocautery was used, but because of the large size of the sac and its thick wall, it was minimally effective.

Considering the huge size of the sac, any attempt to push it up would probably result in optic nerve or chiasma compression, therefore partial removal of the sac was intended. Additionally this strategy might probably minimize the risk of relapse. So, in the next step, an incision was made in the anterior wall of the sac using a sickle knife which led to CSF flow and after enlargement of the whole with a Blakesley forceps, the endoscope was introduced into the sac for close inspection of its contents. The web of the floor of the third ventricle

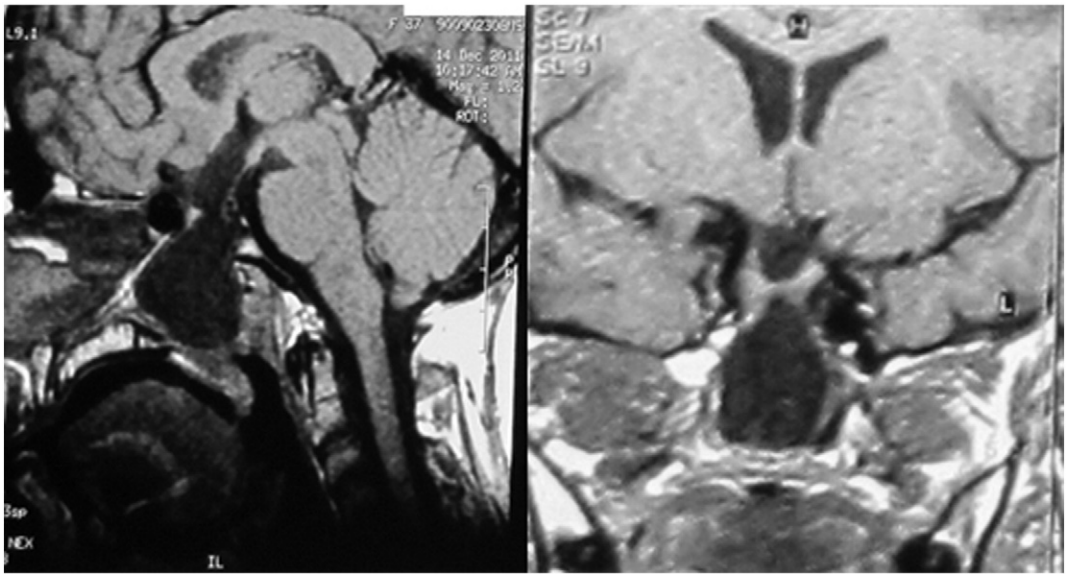

Fig. 2. Sagittal and Coronal T 1-weighted MRI showing the meningoencephalocele positioned posterior to the chiasma and the 3rd ventricle involvement and extension of the lesion to the nasopharynx.

was visualized. Partial resection of the sac at the level of the floor of the sphenoid sinus was made by a turbinate scissors afterwards. Electrocautery of the remnants was carefully performed to weld the dura. The defect was reconstructed by fat and fascial graft. As there was no bony ledge, a titanium mesh was screwed to the remnant of the rostrum and covered with the pedicled nasoseptal flap (*Please watch the attached* video). The patient had nasal packing for the next 48 h. No CSF diversion method was used.

Post-operative period was uneventful and no CSF leak or newly developed anterior or posterior pituitary dysfunction was encountered. Visual field testing on three-month follow-up showed marked improvement. Visual acuities in both sides were 10/10. Perimetry showed marked reduction of the blind fields in the right eye (on the right side: Mean Defect = −9.6, Pattern Standard Deviation = 7.4 and on the left side: Mean Defect = −1.1, Pattern Standard Deviation = 1.7). Post-operative CT scan one year later and nasal endoscopy 18 months after surgery revealed no relapse of the lesion (Fig. 3).

Discussion

A true transsphenoidal meningoencephalocele could not be managed easily because of its proximity to vital structures and the importance of sac contents, such as pituitary gland, cerebral arteries, and optic system [4]. In those cases who are candidates for surgical management, the surgical approach should be chosen based on sac characteristics, its boundaries, proximity to vital structures, the size of the sphenoid bone defect, and concomitant anomalies. Although new surgical methods have been introduced, their usefulness remains controversial mostly due to the very low prevalence of the true TTSME.

Only eight cases of true TTSME were introduced in the adults until the last century. Although most cases were managed conservatively, the transcranial approach was used in the others [1]. Due to higher morbidity, mortality and more post-operative pituitary dysfunction, the transcranial approach is not well accepted in transsphenoidal meningoencephalocele [4,7].

Transsphenoidal route was chosen by Abe et al. [7] for a 26-year-old woman presenting with secondary amenorrhea, diabetes insipidus, bitemporal hemianopia, nystagmus, and morning glory syndrome. The sac was reduced and the defect was repaired by a bony graft, muscle, and fibrin glue. Because of the continuation of the symptoms, the revision surgery performed via the sublabial transmaxillary transsphenoidal approach to push the sac into the sellar cavity. Although the reduction of sac was achieved, the neurological and endocrine abnormalities remained thereafter.

The trans-palatal approach is another alternative which is usually preferred in children due to simultaneous craniofacial malformations [3]. However, this approach has been associated with high rate of post-operative complications and incomplete restoration of the sac. The only successful adult case was an 18-year-old boy with panhypopituitarism and visual loss who underwent microscopic transpalatal approach for correction of TTSME. Eight-year follow up revealed improvement in visual and hormonal functions [8].

In transsphenoidal meningoencephalocele, the endoscope provides a high quality close-up view of herniated sac while its wide working view angle makes the panoramic vision of anatomic landmarks possible. The instrument is utilized with minimum tissue dissection, causing less discomfort and cosmetic problems. Smaller and ergonomically adjusted equipment provide bimanual working field. The best results are achieved by a team of neurosurgeons and otolaryngologists.

Franco et al. [9] reported the first use of endoscopic endonasal approach for the repair of a true TTSME in a child with satisfactory results. Chen et al. [10] described another successful use of endoscopic endonasal approach in a two-year-old boy with apneic–cyanotic spells. The patient became symptom-free and follow-up imaging showed no sign of relapse although mild diabetes insipidus developed post-operatively. The only adult case was described by Saito et al. [4] who presented a 36-year-old man with TTSME managed successfully with endoscopic transsphenoidal approach. They used a septal cartilage and a pedicled vascularized nasoseptal flap for reconstruction. Oxidized cellulose with fibrin glue was used for the sac fixation. Upper airway obstruction was relieved but the visual field defect improved slightly.

As the brain parenchyma inside the sac is presumed to be non-functional, the use of electrocautery to reduce the size of the sac to the level of skull base defect is an acceptable and commonly used strategy. It also has the benefit of removing the mucosa that adhered to the wall of the sac, preventing mucocele formation cephalad to the site of reconstruction.

It is neither our routine nor our suggestion to use titanium mesh cage for reconstruction of sellar and skull base defects with any size and characteristic. We usually deal with the dural defect using either of the two following techniques: harvesting fascia lata plus its overlying fat or using abdominal wall fat plus a septonasal flap mobilization. The first author has performed hard reconstruction with titanium mesh in only two cases out of more than eight hundreds patients operated in the past ten years; In a case of Rathke's cleft cyst [5], a severe herniation of optic system and third ventricle into the sphenoid sinus persuaded us to use a mesh to minimize the risk of recurrence. In the current case a severe anatomic distortion was present. After opening the sac, diaphragma sella was not present and CSF was freely flowing. An abnormal bony tunnel from diencephalon to the sphenoid sinus was present and there was no bony edge for settlement of the flap. There is no study to compare different

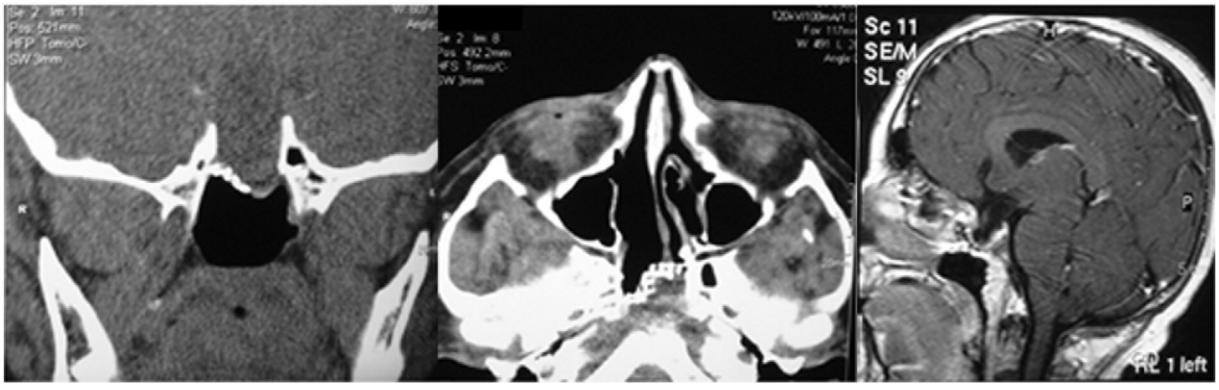

Fig. 3. Left: Post-operative coronal CT scan showing the repaired site. Middle: Post-operative axial CT scan showing the reduction of the size of the sac. The titanium mesh is also seen. Right: Sagittal view MRI showing the reconstructed sella.

reconstruction methods in such an extreme anomalous situation. So we decided to use the maximal reconstruction technique with fat, fascia lata graft, nasoseptal flap mobilization and a titanium mesh screwing for buttressing them.

Although the remaining scar would be annoying, fascia lata grafts are safe, available and have had excellent outcomes. Artificial dural grafts, also, have been used widely with comparable results; but we do not use them commonly because most patients would not afford the expenses. Little high-quality evidence exists to compare autologous versus artificial grafts. So, the choice largely depends on the patient preference and the surgeon's experience [11].

The use of lumbar drain is another uncertainty. While intraoperative CSF drainage through a lumbar drain could reduce the size of the sac and minimize the risk of damage to vital structures [4], chance of relapse remains in addition to other complications [8].

Conclusion

As there are limited reported uses of endoscopic endonasal approach for the repair of true TTSME, this case is a good example of the ability of this minimally invasive approach to be used in repair of huge meningoencephalocele in this region. Reduction of a bulky sac might induce some pressure on the optic chiasma; however careful resection of inferior part of the herniated sac with concomitant repair of the CSF leakage can be successfully performed with improvement of the visual field defect and no further complication.

Selection between different reconstruction techniques is really challenging and should be individualized for each patient depending on the surgeon's experiences, presence and intensity of CSF leak, the size and bony anatomy of the defect and patient preferences, until high-quality evidence becomes available to help decision making.

Supplementary data to this article can be found online at http://dx.doi.org/10.1016/j.inat.2015.02.002.

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