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TECHNICAL NOTE

Sublabial transsphenoidal microsurgical technique to treat congenital transsphenoidal encephalocele: a technical note

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Abstract

Encephalocele is a rare malformation consisting in herniation of cranial contents through a cranial defect. A transsphenoidal location is uncommon, representing 5% of all basal encephaloceles. The surgical treatment of transsphenoidal encephaloceles is challenging. An optimal approach has not yet been determined, and it varies according to the surgical experience. We report the surgical management of a transsphenoidal encephalocele. The encephalocele and the sellar defect were repaired through a sublabial transsphenoidal microsurgical approach (TSM). Preoperative magnetic resonance imaging (MRI) and computed tomography (CT) scans were crucial for surgical planning. The sublabial transsphenoidal microsurgical approach offered a good and complete exposure of both the sac and the bone defect. Therefore, the congenital defect was successfully repaired with complete resolution of the encephalocele without any surgical or medical complications. Postoperative CT scan and MRI showed the restoration of the bone defect and the recovery of a normal anatomy with herniated structures pushed back into the sella. The described sublabial transsphenoidal microsurgical approach represents a minimally invasive, safe, and effective treatment strategy for transsphenoidal encephalocele.

Keywords Transsphenoidal encephalocele · Basal meningoencephalocele · Transsphenoidal approach · Congenital · Microsurgery

Introduction

The herniation of brain tissue and cranial meninges through a defect in the skull base is called an encephalocele or meningoencephalocele. The herniations may be congenital, spontaneous, or secondary to trauma [5]. The classification system for encephaloceles is based on the location of the skull defect. The basal encephaloceles are rare and are further classified into transethmoidal, sphenoorbital, sphenomaxillary, and transsphenoidal [14]. A transsphenoidal location is quite uncommon, representing 5% of all basal meningoencephaloceles [3]. The size of the transsphenoidal defect is often large and these lesions are much harder to treat, since vital structures, such as the hypothalamus, the pituitary gland, anterior cerebral arteries, optic nerves, the optic chiasm, and the third ventricle might be inclined to herniate [6]. Transsphenoidal encephalocele treatment has been controversial for years with regard to indication and type of surgery. Most of transsphenoidal meningoencephaloceles are usually diagnosed during the first year of life after the appearance of clinical symptoms, such as visual disturbances, endocrine dysfunctions, water or electrolytic disturbances, dyspnea due to nasopharyngeal obstruction, CSF rhinorrhea, and recurrent meningitis [4, 5]. According to literature, surgical correction is generally recommended, in order to prevent the onset and/or resolve related symptoms [8, 10]. Surgical treatment of transsphenoidal encephaloceles is challenging and the optimal surgical approach has not yet been determined due to their deep location and the experience of the surgical team [1]. In our Institute, we recently operated a child affected by congenital transsphenoidal encephalocele who was successfully managed using a sublabial transsphenoidal microsurgical approach (TSM). We describe the TSM technique to treat transsphenoidal encephalocele and reconstruct the cranial base defect.

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Case presentation

History and examination

A 4-year-old child was admitted to our department because of delayed growth and several episodes of hypoglycemia. General examination showed short stature (< third percentile); weight 11.55 kg (− 3.85, SD), height 85.6 cm (− 4, SD). Endocrine workup showed low levels of IGF-1 (12 μg/L, range 50–190 μg/L) and a low GH peak (1.2 ng/mL, range 0–5 ng/mL) after stimulation test with arginine. The other hormone tests were normal. MRI scan revealed an enlargement of the craniopharingeal duct with a voluminous transsphenoidal encephalocele reaching up to the nasopharyngeal mucosa and choanae occlusion (Fig. 1). The high-resolution CT scans confirmed the large skull base defect (Fig. 2). Preoperative ophthalmological findings showed a defect of visual acuity (5/10 right eye; 3/10 left eye) and an amplitude drop of the left optic nerve at visual evoked potential (VEP). Once neuroradiology, laboratory, and clinical evaluations were completed, the indication was given for surgical treatment of transsphenoidal encephalocele. Standard informed consent relating to the procedure was obtained from child's parents.

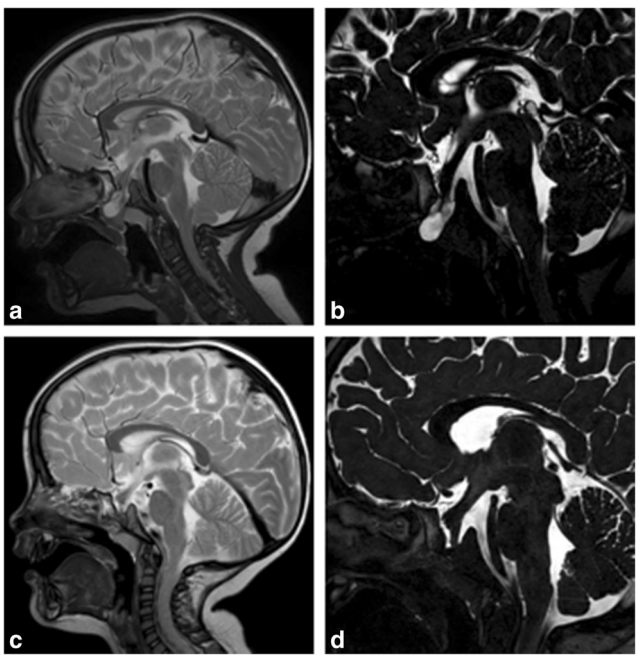

Fig. 1 **a, b** Preoperative sagittal high-resolution T2-weighted and drive T2-weighted MR images without contrast, respectively, showing the transsphenoidal encephalocele and its contact with the nasopharyngeal mucosa. **c, d** Follow-up postoperative sagittal high-resolution T2-weighted and drive T2-weighted MR images without contrast, respectively, obtained 1 month from surgery demonstrating the resolution of the encephalocele and the repair of the sphenoid defect

Positioning of the patient

The patient was placed in a supine position on the operative table (Maquet, Rastatt, Germany) with a 30-degree angulation at the trunk-thigh hinge, a 20-degree head extension and a 25-degree left head rotation. The surgeon was located on the right side of the patient.

Preoperative care

The skin of the face, the nostrils, and the buccogingival junction were disinfected with a clorexidine solution. A 10-ml solution of mepivacaine and adrenaline 1/100,000 (mepiforan; Baxter, Glendale, CA) was injected bilaterally at the caudal end of the nasal septum and at the buccogingival junction to allow analgesia and to facilitate dissection of the nasal mucosa.

Operation

A microscope (OPMI Pentero 900, Carl Zeiss Meditec, Jena, Germany) was used at the beginning of surgery. The upper lip was gently retracted superiorly. A vertical 2-cm incision was made at the buccogingival junction (Fig. 3). Care was given to incise the mucosa quite far from the gingival border to avoid postoperative retraction. The mucosa was elevated from the maxilla subperiosteally to ensure total bone exposure of the nasal spina. The nasal spina was left in place. The septal cartilage was detached from the nasal spina and a septal submucosal unilateral tunnel was created via a blunt subperichondrial dissection. The dissection was continued using a Killian nasal speculum and a blunt suction tube. The entire left side of the nasal septum is pushed back to the perpendicular plate of the ethmoid (the bony part of the septum). The cartilaginous portion of the septum was subsequently dislocated and tilted to the right. A longer nasal speculum was introduced, and blunt dissection was continued. The bony nasal septum was then isolated from the mucosa bilaterally and the anteroinferior wall of the sphenoid was exposed (the absence of the sphenoidal rostrum was noted). A piece of the bony septum was removed and stored in antibiotic solution for the subsequent reconstruction. The barrel of a 5-ml syringe (Emerald syringe, Becton Dickinson, Mequinenza, Spain) without the distal end was used as a self-retractor. It was inserted gently allowing an angular view in horizontal and vertical direction. The anterior and inferior border of the sphenoid was widely thinned using a Kerrison punch. The mucosa of the nasopharynx was cut down and dissected off the encephalocele sac in all direction. The sac was gradually separated from the surrounding mucosal layer. The herniated encephalocele was progressively dissected with bipolar coagulation for wall shrinkage and gradually pushed back into the sella (Fig. 3). The reconstruction of the sellar floor was thus

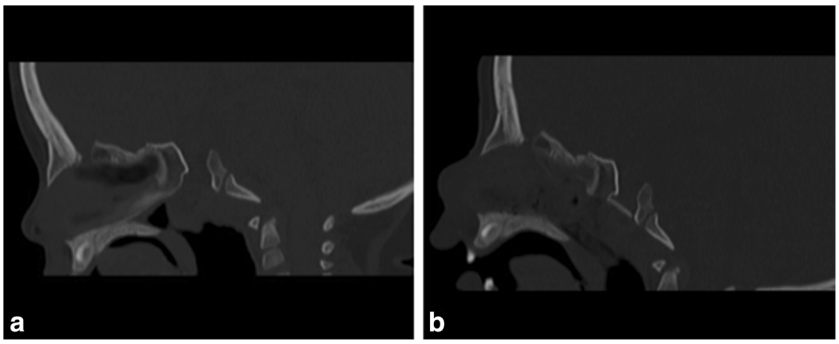

Fig. 2 **a** Preoperative sagittal CT scan without contrast (bone view) showing the congenital defect of the sphenoid bone causing the transsphenoidal encephalocele. **b** Early postoperative sagittal CT scan without contrast (bone view) showing the bone fragment from the nasal septum used to close the sellar defect

obtained using the bone fragment of the nasal septum previously removed. The fragment was tailored and placed overlying to the dura. It was kept in place by inserting it under the borders of the sellar defect (Fig. 3.). Fibrin glue (Tissucol; Baxter Corp.) was then applied as a sealing system.

An antibiotic solution of rifamycin (250 mg; rifocin; Lepetit S.p.A, Lainate, Milan, Italy) was left in the sphenoid sinus after washing it with peroxide and saline solution. The septal mucosa was pushed medially to cover the residual bony and cartilaginous septum, which was replaced in its original location. Bilateral anterior nasal packing was then performed under microscopic view using two soft standard nasal dressings (merocel; Medtronic Xomed Surgical Products, Jacksonville, FL). The sublabial wound was sutured with absorbable thread (Safil Quick; Braun, Tuttlingen, Germany). The patient was woken up immediately after surgery.

Postoperative management and follow-up

The postoperative course was uneventful. Daily fluid balance, serum electrolytes, serum, and 24-h urinary osmolality were monitored daily until discharge. No hormonal or electrolytic disturbances were recorded during hospitalization. No surgical postoperative complications occurred; in particular, there was no cerebrospinal fluid (CSF) leak. The patient was discharged on the fifth postoperative day.

Postoperative and follow-up data were collected both by patient's visit and by report from the patient's pediatrician.

Postoperative visual acuity, 1 week, 6 months, and 12 months after surgery remained unchanged in comparison to preoperative evaluation (5/10 right eye; 3/10 left eye). Moreover, postoperative VEP, performed at 6 months after surgery, showed and confirmed a low amplitude of signal on the left optic nerve compatible with a damage of the nerve.

Postoperative computed tomography (CT) scan performed immediately after surgery (Fig. 2), and magnetic resonance imaging (MRI) studies performed 1 month, 6 months, and 1 year later demonstrated complete resolution of the encephalocele (Fig. 1). The growth hormone deficiency was treated with synthetic GH replacement, while the other hormone functions were stable (last follow-up 14 months after surgery).

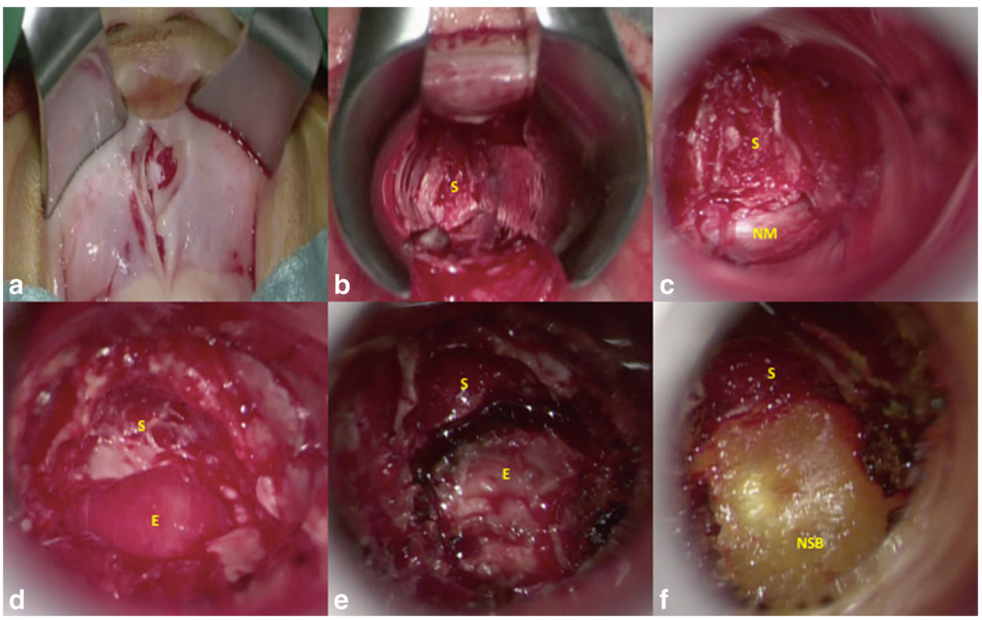

Fig. 3 **a** Microscopic view of the incision at the buccogingival junction. **b** View through the nasal speculum used during the dissection of the nasal septum from the mucosa to allow identification of the anteroinferior border of the sphenoid (S). **c** Anteroinferior wall of the sphenoid partially thinned to see the nasopharyngeal mucosa (NM) adherent to encephalocele sac (E). **d** The nasopharyngeal mucosa is cut down to expose the sac (E). **e** Encephalocele reduced by the use of bipolar coagulation and then pushed back between the anteroinferior and posterior border of the sphenoidal defect. **f** Fragment of bone taken from the nasal septum (NSB) and used to close the sphenoid defect

Discussion

The ideal surgical approach to treat transsphenoidal encephalocele has not yet been established and the treatment method varies according to the experience of the surgical team involved [1]. Preoperative MRI and CT scans remain crucial for surgical planning [13].

Currently, the approaches reported in the literature are transcranial, endoscopic endonasal, and transpalatal [15]. In previous series of patients treated by the transcranial approach, the mortality and morbidity rate approached 50 and 70%, respectively [4, 16]. This procedure can be performed with retraction of the frontal lobes, keeping the dura intact. The floor of the anterior cranial fossa is accessed to visualize the encephalocele. The position of the sac in a median perisellar transsphenoidal encephalocele is quite low and requires for a significant traction force to be exerted on the frontal lobe for the encephalocele to be reached intracranially. Furthermore, the exposure of the bone defect with an intracranial approach is limited and the traction could cause brain edema, ischemia, and sometimes damage to the encephalocele sac and its structures, producing new visual and/or hormonal problems. In certain cases, it might be necessary to resect the encephalocele in order to elevate the sac above the epipharynx with possible subsequent hypothalamic damage.

In cases of transsphenoidal encephaloceles with large cleft palate defects, some authors used an extracranial transpalatal approach. In these cases, the soft palate was split along the midline and when necessary the hard palate was opened risking palatal wound dehiscence, infections, meningitis, and prolonged enteral tube [11].

Zeinalizadeh et al. reported an endoscopic endonasal approach to treat transsphenoidal encephalocele with no excision of encephalocele sac and good cranial base reconstruction. They performed CSF aspiration from the anterior inferior portion of the sac to decompress the encephalocele sac before pushing it upwards. They did not report major surgical or medical complications showing the safety of technique [17].

The sublabial transsphenoidal microsurgical technique to treat transsphenoidal encephalocele has never been described in the literature. The sublabial exposure is historically preferred for transsphenoidal lesions in the pediatric population due to the small diameter of the nares and lack of aerated sphenoid sinuses. We think that TSM, like the endoscopic approach, offers a good and complete exposure of both the sac and the bone defect with no need to resect the herniated sac. It allows the dissection of the sac from nasopharyngeal structures under continuous visual control with no need to exert any traction. This decreases the risks of damaging the sac's contents or dural tearing.

The major potential risks related to transsphenoidal surgery for the treatment of transsphenoidal encephalocele are CSF leak, meningitis, vascular complications, visual complications, hypopituitarism, diabetes insipidus, and sinonasal alterations. Although to date, there is no scientific evidence finding the endoscopic approach for pituitary illnesses better than microscopic one, the former is known to be associated with significantly higher rates of endonasal and bleeding complications [2, 12]; in fact, the lateral displacement or resection of lateral turbinates with the posterior nasal septum during the endoscopy cause an alteration of the normal nasal cavity [7, 9].

Like the endoscopic approach, TSM is minimally invasive compared to transcranial and transpalatal approaches with good resolution of the congenital defect and low risks of complications.

Conclusion

Surgical treatment of transsphenoidal encephaloceles is challenging. The optimal surgical approach has not yet been established and varies according to the experience of the surgical team involved and confidence with the technique. In experienced hands, TSM is a safe technique to treat a transsphenoidal encephalocele and has low risks of surgical complications, recurrence and mortality.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval The study has been performed in accordance with the ethical standards as laid down in the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.

Informed consent Informed consent was obtained from parents of the child included in the study.

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