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CASE REPORT

Endoscopic management of congenital meningo-encephalocele with nasal flaps

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Meningo-encephalocele;
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Summary The objective of this paper is to present a case of a 2-year-old girl diagnosed with a meningo-encephalocele after episodes of meningitis, and treated with a transnasal endoscopic approach using nasal septal flaps pediculated at the sphenopalatine artery. Endoscopic repair is a viable and minimally invasive alternative to traditional craniotomy, however technical difficulties encountered as well as questions that remain unanswered are discussed.

1. Introduction

Congenital encephaloceles, meningoceles or meningo-encephaloceles are protrusions of cranial contents beyond the normal limits of the skull [1,2]. The management of these congenital defects has changed dramatically with nasal endoscopic techniques. Historically, these lesions were approached with a bicoronal incision and frontal craniotomy. Often it was necessary a pericranial flap in order to reconstruct the skull base defects [3,4].

Unfortunately some disadvantages of these approaches were anosmia, intracranial hemorrhage or edema, epilepsy, and memory or concentration deficits [4]. Many of these complications were avoided after the use of nasal endoscopic techniques. The endoscopic repair of intranasal encephaloceles, meningoceles, meningo-encephaloceles, and cerebrospinal fluid (CSF) leaks in adults, mostly traumatic or iatrogenic, has become the standard of care [5–8]. However endoscopic management of congenital skull base defects present several challenges in pediatric patients [1].

Many of these children can present various signs and symptoms, including meningitis, nasal obstruction, craniofacial deformity or CSF leak, but, in some cases, the lesion may go undetected until it is noted incidentally on imaging studies [1]. In addition, the size of the nasal cavity can present a

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technical difficulty, especially with instruments and endoscopes handling [9].

The objective of this paper is to present a case of a 2-year-old girl diagnosed with an meningo-encephalocele after episodes of meningitis, and treated with an endoscopic approach using nasal septal flaps pediculated at the sphenopalatine artery. Technical difficulties encountered and questions that remain unanswered are discussed.

2. Case report

LP, 24 months old, was referred to our service in April of 2008 with a diagnosis of a meningo-encephalocele, after two consecutive episodes of bacterial meningitis. She was treated with intra-venous antibiotics and after the resolution of the second episode of meningitis, image studies were done. Computer tomographic (CT) exams showed a funnel-shaped bony skull base defect (Fig. 1) and the magnetic resonance imaging (MRI) showed a large meningo-encephalocele originated from the third ventricule, arising at the posterior part of the nasal cavity (Fig. 1). The girl also presented a cleft soft palate and hypertelorism, but did not present nasal obstruction or other symptoms. After a carefully preoperative evaluation, she was submitted to an endoscopic repair of the meningo-encephalocele and the skull base defect.

3. Surgical repair

The surgery was done under general anesthesia, with the dorsum slightly elevated in 30° angle. After a topical vasoconstriction of the nasal cavity with high concentrated adrenalin (1:1000) soaked cottonoids for 10 min, a nasal endoscopy was performed with a traditional 4-mm, 0° nasal endoscope.

The lesion was at the posterior part of the nasal septum. A navigation system was used, and an oral endoscopy was also performed using the cleft palate to have another way of accessing the nasal cavity (Fig. 2A).

After local infiltration of the anterior part of the nasal septum with a solution of xylocaine and adrenaline (1:100,000), an anterior incision was done at the right side of the nasal septum. Special pediatric instruments were used such as pediatric dissectors, aspirators, sickle knifes and small Kerrison punches. A mucoperichondral and periosteal dissection was done at the right side and at the left side. Two parallel incisions were done at the right side of the nasal septum in order to create a flap pediculated at the sphenopalatine artery. At the other side, the same incisions were done in order to create the contralateral pediculated flap.

The middle turbinate of the right side was removed in order to gain more space to work with instruments through both nostrils. The removed

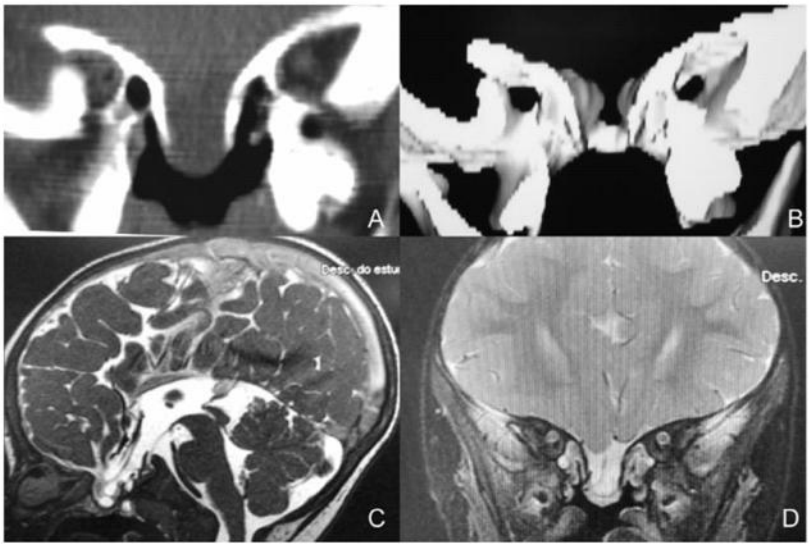

Fig. 1 (A) Coronal view of a computer tomography (CT) showing an extensive skull base defect. (B) 3D coronal reconstruction of a CT showing a "funnel-shape" skull base defect. (C) Saggital reconstruction of a T2-weighted magnetic resonance imaging (MRI) of a meningo-encephalocele arising from the third ventricule. (D) Coronal view of MRI showing a meningo-encephalocele.

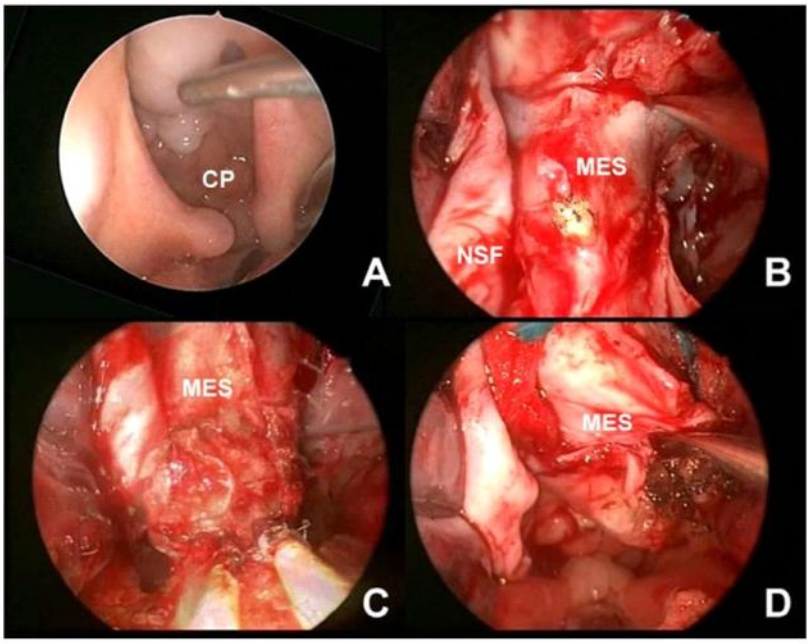

Fig. 2 (A) Oral endoscopy showing the cleft palate (CP) and the lesion at the posterior part of the nasal septum. (B) Meningo-encephalocele sac (MES) and nasal septum flap (NSF) of the right side pediculated at the sphenopalatine artery. (C) Resection and ablation of the meningo-encephalocele sac (MES). (D) Visualization of the neck of the meningo-encephalocele sac (MES).

turbinate mucosa was also used as a free-graft to help in the closure of the skull base defect.

Nasal septal cartilage was removed and saved for possible use in the reconstruction of the skull base defect. The meningo-encephalocele was identified and isolated (Fig. 2).

The bony skull base "V" or funnel-shaped defect could not support the nasal septum cartilage. Fascia lata and the nasal septum flaps were used for this purpose (Fig. 3).

Fibrin glue was positioned (Fig. 4) and a nasal packing with Gelfoam© was designed and inserted. A Folley© catheter was positioned, and inflated in order to keep the nasal packing in place. A lumbar drainage catheter was inserted.

4. Post-operative

After the procedure the child was maintained sedated for 2 days in order to keep the nasal packing in place. Antibiotics were given to the patient before the procedure, and for 10 days after the surgery. After 2 days, the Folley© catheter was removed. No CSF leak was noted. Five days after the procedure the child was discharged from the hospital with no signs of CSF leak, infection, respiratory distress or difficulty to eat.

Nasal exams and cleanings were done at the office during the visits. These nasal toilets were performed carefully, avoiding any stress to the patient. Imaging studies were done 30 days after the procedure. At this time no signs of complications such as infections or CSF leaks were observed.

5. Discussion

Endoscopic techniques can be performed in fairly young children for the treatment of many lesions such as choanal atresia, orbital cellulitis and meningo-encephaloceles [1,7–9]. However, the surgeon has to know that pediatric patients require meticulous preparation and special instruments. We agree with other authors that congenital intranasal encephaloceles rarely need to be treated immediately after birth, because they are nearly always covered with healthy tissue layers [1]. The surgeon can wait until the child is aged 2–3 years for significant facial growth that will facilitate an

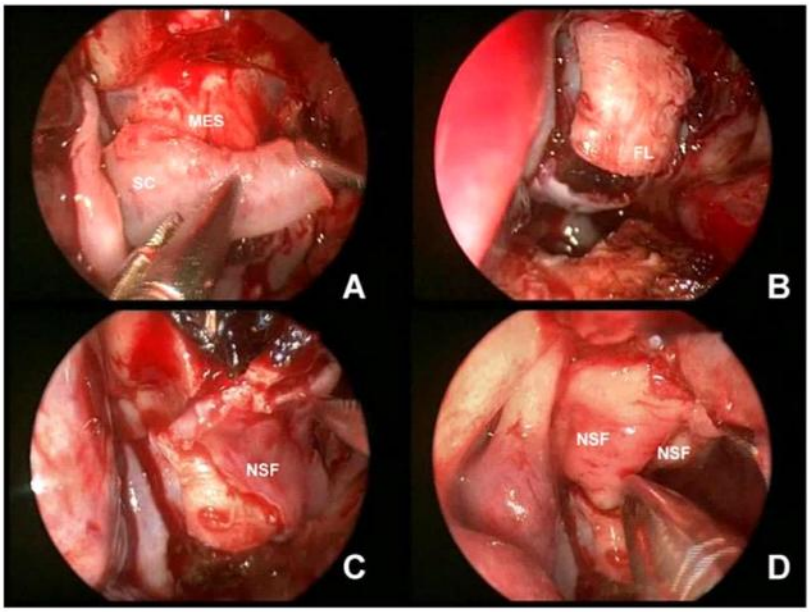

Fig. 3 (A) Meningo-encephalocele sac (MES) and nasal septal cartilage (SC). (B) Positioning of fascia lata (FL). (C and D) Positioning of the nasal septal pediculated flaps (NSF).

endoscopic management. However, the children's clinical signs and symptoms must be paramount for an early surgery intervention.

In this particularly case we used nasal septum flaps pediculated at the sphenopalatine artery to help in the closure of the skull base defect (Fig. 5). These nasal flaps, recently described, can be used in skull base surgery, when available, in the closure of CSF leaks in adults [10,11]. These pediculated flaps receive direct blood supply from the sphenopalatine artery, making them a much robust tissue to help in the closure of skull base defects. In adults its use is well described, however in pediatric patients it is still restrict and less described in the literature [10,11].

There are few studies about these flaps complications, such as mucoceles, synechia, necrosis and infection in adults. Few long-term follow-ups exist, but with short-term follow-ups, these flaps have become a safe and reliable treatment for skull base defects.

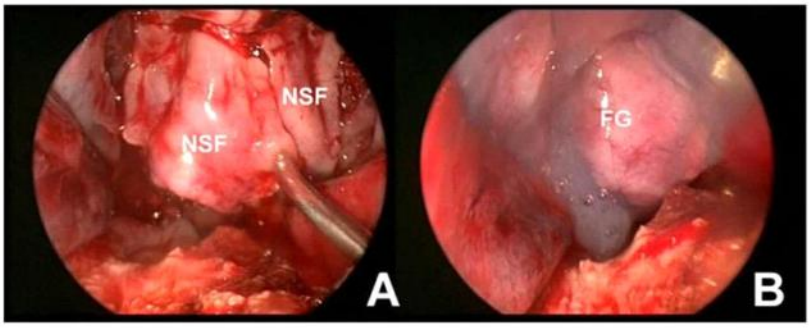

Fig. 4 (A) Final aspect of the nasal septal flaps (NSF). (B) Fibrin glue (FG).

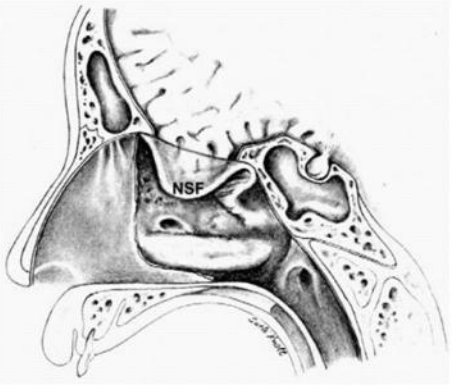

Fig. 5 Illustration of the right side nasal septal pediculated flap (NSF) positioned over the meningo-encephalocele sac.

A topic that deserves discussion is the nasal packing. There are few nasal packings specially designed for pediatric patients. We used Gelfoam© soaked in antibiotics in the nasal cavity. A Folley© catheter was also inserted to try to hold the flaps and the packing into the correct place.

The nasal packing time was another point in which we could not find support at the literature. We removed the Folley© balloon 2 days after the surgery, but absolvable material was kept in place for as long as it stayed.

Another important topic was the multidisciplinary team. A team with a pediatrician, pediatric anesthesiologist, otolaryngologist, neurosurgeon, intensive care pediatrician, and radiologist was paramount for the successful management of this case. This team agreed to maintain the child under general anesthesia for 2 days after the surgery. This decision was made because of several reasons: to keep the nasal packing into its right place and to remain the child in absolute rest.

6. Conclusion

We presented a successful reconstruction of a congenital mencephalocele with the use of bilateral nasal septum flaps pediculated at the sphenopalatine artery. No complications were observed with the nasal flaps, and the skull base defect was successful corrected.

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