Successful endoscopic endonasal repair of nasal meningoencephalocele in a 21-day-old neonate

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ABSTRACT

Introduction: *Indications of surgical intervention in congenital nasal meningoencephaloceles includes presence of cerebro spinal fluid rhinorrhea having a risk of causing meningitis, episodes of prior meningitis and bilateral nasal obstruction causing respiratory difficulty in these obligate nasal breathers. Many authors would like to wait till the patient attains the age of 2 to 3 years for repair of the defect due to surgical feasibility. However, early intervention prevents further episodes of meningitis in the future. We present the youngest patient of nasal meningoencephalocele successfully repaired via endoscopic approach.*

Case Report: *A 21 days old neonate was referred to us with a nasal meningoencephalocele with active cerebrospinal fluid rhinorrhoea. Radiological investigation showed a cribriform plate defect on the right side. Repair was done by endoscopic route by multi-layered closure of the defect which was augmented with a mucoperichondrial flap from the septum. Patient was asymptomatic in the post-operative follow up period and did not have any episode of meningitis till date.*

Conclusion: *Early repair by transnasal endoscopic route is a feasible surgical option for congenital anterior skull base defects with meningoencephaloceles to prevent further episodes of meningitis. This is feasible even in the neonatal period due to improved technique and instrumentation now available for endoscopic nasal surgeries.*

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With the improvement in endoscopic techniques and high-resolution scans, more accurate localization of a defect and successful repair has become possible, thus avoiding the risks of craniotomy, along with a cosmetically acceptable transnasal route.[1–5] Dermoids, encephaloceles, and gliomas are included in the differential diagnoses for pediatric midline nasal mass, and, in the nasal cavity, nasal meningoencephalocele can often be easily mistaken for a benign nasal polyp. The difficulty in treating these lesions lies in the ability to adequately address the repair of the skull base defect and avoid the future risks of developing meningitis in cases of meningoceles, especially when it is associated with active cerebrospinal fluid (CSF) rhinorrhea. Endoscopic repair of these lesions has been reported in the literature of occurring earlier in infants and older children in recent years.[1,2] We report a case of congenital nasal meningoencephalocele in a 21-day-old neonate with active CSF leak. To our knowledge, this also represents the youngest patient (21 days old) reported in the literature to date, which emphasizes the important role of otorhinolaryngologists in the management of these patients at a young age, *i.e.*, even in their neonatal period.

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CASE REPORT

An 11-day-old infant was referred to us after attempted endonasal removal of a polyp at a different facility. When we examined her, the infant was having right-sided active watery nasal discharge, which was not foul smelling and which increased during crying. There was no history of fever, seizures, excessive cry, or feeding difficulties. Birth history was uneventful. There was an associated history of respiratory difficulty since birth, which was aggravated during sleep. Results of biochemical and cytologic analysis of the discharge were suggestive of CSF. Glucose and protein levels and total and differential leukocyte counts were within normal ranges for CSF. The fluid was positive for the presence of β transferrin. Magnetic resonance imaging revealed a meningoencephalocele protruding from a defect in the right cribriform plate (Fig. 1 *B, C*). Nasal endoscopy revealed a pale pinkish mass with an irregular surface that completely occupied the right nasal cavity and displaced the septum to the opposite side. (Fig. 1 *D*). By using zero degree 2.7-mm nasal endoscope, the meningoencephalocele sac was electrocauterized at the cribriform plate by using bipolar diathermy and was debrided at the site of the defect by using a pair of scissors. Mucosa at the margins of the defect was debrided by using a microdebrider to delineate the bony margin of the defect. At each attempt during the surgery the instrument was introduced first, followed by the endoscope to visualize the surgical field because the dimensions of the anterior nares were too small to accommodate both if introduced together (Fig. 1 *A*). A view could even be obtained by

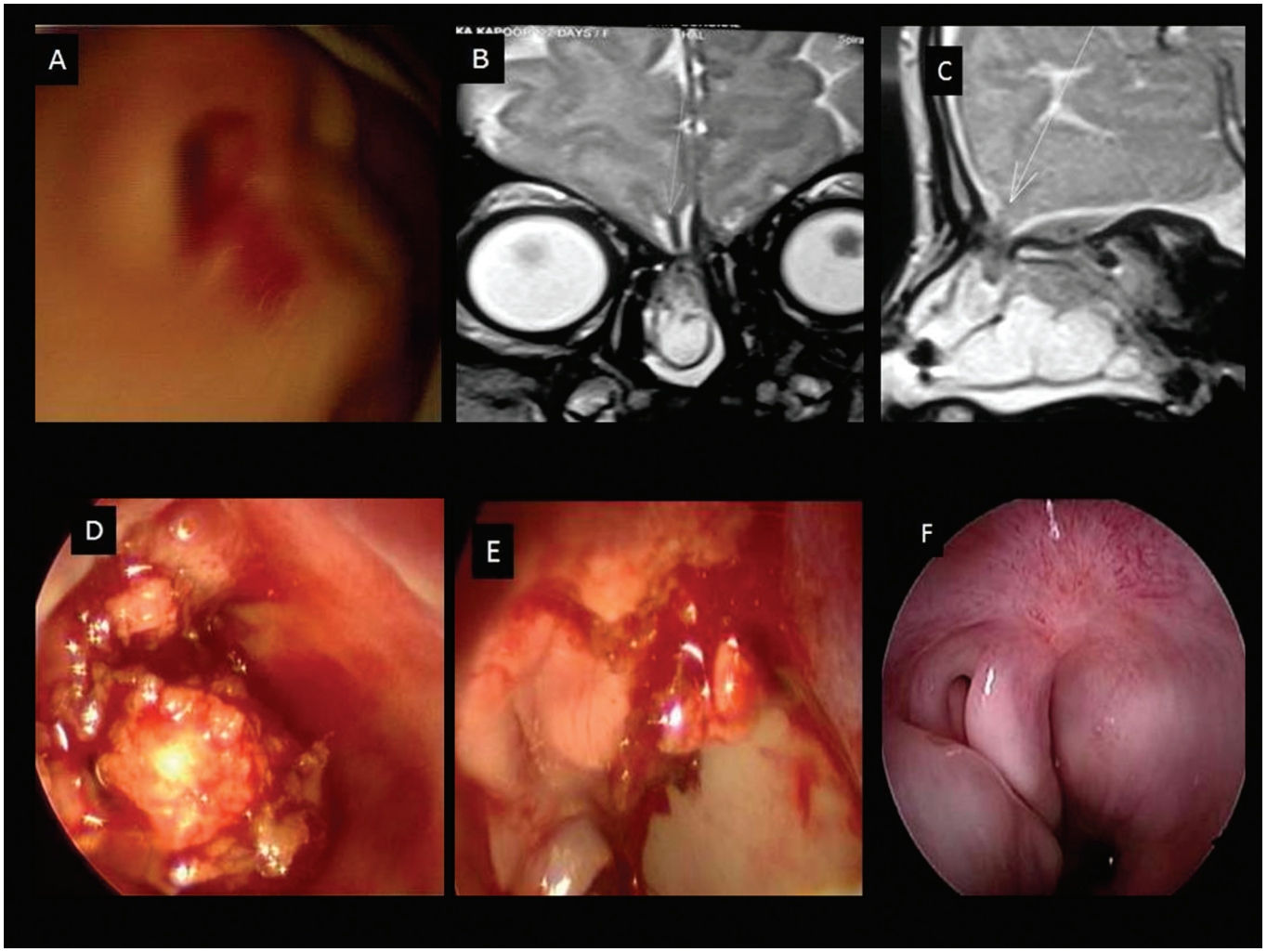

Figure 1. *(A) Clinical photograph immediately after diagnostic endoscopy; note the stretching of the nostril due to the endoscope. (B) T2-weighted MRI, coronal section, showing an intense soft tissue density in the right nasal cavity, which is communicating with intracranial contents; the white arrow points to a possible flow void. (C) T2-weighted MRI, sagittal section, showing an intense soft tissue density occupying the whole right nasal cavity; the white arrow points to a defect in the cribriform plate. (D) Endoscopic view of the meningoencephalocele sac; note the irregular surface due to an earlier attempted removal. (E) Endoscopic view of the septomucosal flap that was used to plug the defect after excision of the meningoencephalocele. (F) Repeat endoscopy at 5 months postoperative period, showing a completely healed right nasal cavity. MRI, magnetic resonance imaging.*

keeping the endoscope at the nares. The defect was sealed by a rotated posterior-based nasal septomucosal flap (single-layered repair) (Fig. 1 *E*). The repair of the defect was further augmented with the application of fibrin glue, and hemostasis was achieved to end the procedure. Lumbar drainage was not done during the postoperative period because of the age of the neonate to avoid any procedure-related complication and infections. The patient was given antibiotic prophylaxis with antimeningitic doses of ceftriaxone during the postoperative period for 7 days. She was discharged on postoperative day 7. The patient is under our continuous follow-up to date. An nasal endoscopy was performed at the 5-month follow-up, which showed a well-healed skull base without any CSF leak or skull base defect or encephalocele (Fig. I *F*). A magnetic resonance cisternography picture taken during the follow-up showed the absence of any encephalocele or active CSF leak. (Fig. 2).

DISCUSSION

The development of congenital nasal encephaloceles, gliomas, and dermoids begins during the early stages of gestation.[6,7] These lesions, depending on their location, are classified as occipital (75%, most common), frontonasal (sincipital), or basal.[6,8] Frontonasal lesions can project anteriorly onto the nasal dorsum or laterally into the orbit, whereas basal lesions present intranasally and can be subdivided into transethmoid (intranasal), sphenoeth-

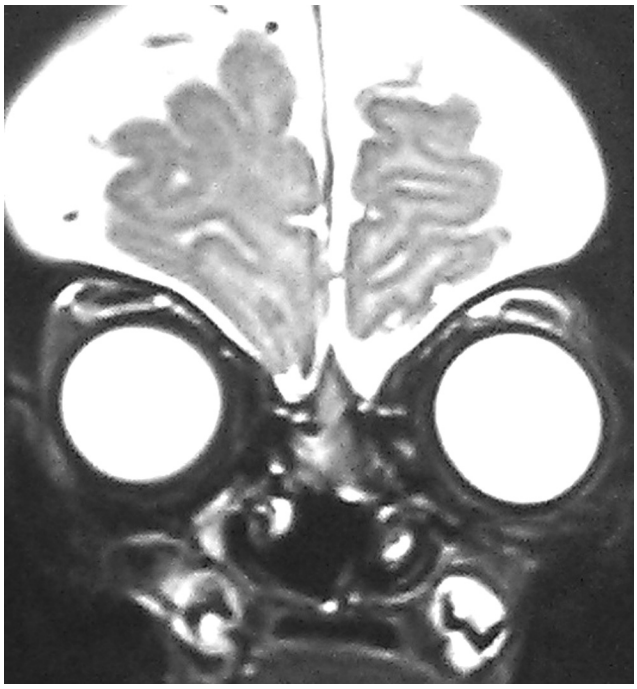

Figure 2. Magnetic resonance cisternography picture during follow-up, showing the absence of encephalocele or active CSF leak.

moidal (nasopharynx), transsphenoidal (nasopharynx), and sphenomaxillary (pterygopalatine fossa).[5] Encephaloceles that contain only meninges are called meningoceles, and those that have brain and meninges are called meningoencephaloceles.

Woodworth and Schlosser[2] showed that there were three episodes of meningitis encountered in patients with active CSF leaks. The actual risk of ascending meningitis in a congenital skull base defect without an active CSF leak is likely low. However, intact mucosa and dura may not provide an impermeable barrier against the spread of an ascending infection. Hence, the concept of early intervention in cases of congenital anterior skull base defect has emerged, especially when it is associated with active CSF rhinorrhea.

Identification and delineation of the bony defect at the skull base, bipolar diathermy of the encephalocele sac, resection of the neck of the meningoencephalocele sac and multilayer closure of the resultant defect are the key steps in the surgical management of congenital nasal meningoencephaloceles.[4–6] We suspected a flow void in the intracranial communication of meningoencephalocele in our patient. Bipolar cautery provides safe excision in such cases. Multiple techniques have been reported in the literature that advocate underlay, overlay, cartilage or bone, fat plug, and fascia for the closure of these defects.[9–12] We used a single-layer closure to plug the defect in the skull base after resection of the meningoencephalocele sac. To reduce the donor site morbidity in a very small child, no fat, muscle, or bone was used. Fibrin glue was used to seal the intranasal mucosal graft in place and has been successfully used in the past.[9–13]

Preoperative imaging should be obtained to look for intracranial connections and the presence of herniated brain parenchyma or cerebral vasculature. High resolution computerized tomography gives excellent bony detail of a defect in the skull base. However magnetic resonance imaging allows for better evaluation of the soft tissue, CSF, and flow voids associated with vasculatures,[14] as in our case (Fig. 1 *B*). Because our patient was a neonate of barely 21 days old, to limit the radiation exposure, we used magnetic resonance imaging as the only radiologic investigation.

The endoscopic endonasal approach also avoids facial incisions or a craniotomy with frontal lobe retraction. This helps to facilitate management of these lesions at an earlier age given the potential for reduced morbidity compared with the open approaches. The endonasal approach is common these days for removal of anterior skull base lesions and repair of anterior skull base defects. Pediatric sinus surgery also lacks any effect on long-term facial growth compared with the open approaches.[15,16] The availability of good pediatric nasal endoscopes and improved visualization has immensely improved the use of endoscopic endonasal procedures for the anterior skull base. Our case supports the belief that endoscopic endonasal repair can be accomplished with good success in a neonate. To date, the youngest a successful repair of nasal meningoencephalocele has been done was the age of 2 months.[1]

CONCLUSION

An endoscopic approach to the repair of anterior skull base defects in cases of congenital meningoencephaloceles is less morbid and offers a shorter hospital stay to the patient and avoids the incisions and facial growth abnormalities. This approach can be successfully used, even in infants, due to the availability of good endoscopes and visualization instruments, *e.g.*, cameras. Hence, the age of intervention is also decreasing. Those patients with symptoms of nasal obstruction and CSF leak and, hence, an inherent risk of developing meningitis should be intervened early in the life. To our knowledge, we report the youngest patient reported in the literature to date, 21 days in age, operated successfully by using a single-layered repair. Hence, age does not seem to be a contraindication any more with better instrumentation available for obligate nasal breathers such as neonates, who also have associated breathing difficulty for large congenital nasal masses along with the inherent risk of developing meningitis in the

presence of active CSF rhinorrhea, when associated with meningoencephaloceles.

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