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Original Article

The Endonasal Endoscopic Management of Pediatric Anterior Meningoencephaloceles, a Tertiary Care Hospital Experience

Shatha AlGhamdi,[1] Fatimah Al Hassan,[2] Abdulrahman Al-khatib,[3] Ali Almomen[3]
[1]College of Medicine, King Faisal University, AlAhsa, SAUDI ARABIA
[2]Department of Surgery, King Fahad Specialist Hospital, Dammam, SAUDI ARABIA
[3]ENT Department, King Fahad Specialist Hospital, Dammam, SAUDI ARABIA

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BACKGROUND: Skull base defects are considered one of the rarest conditions in children. The diagnosis and management of skull base defects in pediatrics are challenging. These defects can be congenital or acquired in etiology.

OBJECTIVE: To assess our experience with the endoscopic endonasal management of pediatric sinonasal meningoencephaloceles and anterior skull base defects in a tertiary referral hospital.

PATIENTS AND METHODS: This is a retrospective study involving 6 pediatric patients, with age ranging from 2 months to 15 years, who underwent endoscopic endonasal sinonasal meningoencephaloceles excision and skull base reconstruction between the period of 2010 and 2020. Three patients were repaired with multilayer technique using septal bone as an underlay, mucosal graft as an overly and supported with gelfoam. The other 3 patients were managed by single layer repair with mucosal graft that was supported with tissue glue and gelfoam.

RESULTS: The study included a total of 6 pediatric patients with main presenting symptoms of nasal obstruction, cerebrospinal fluid (CSF) leak, and meningitis. All patients had a successful excision of their sinonasal meningoencephaloceles and skull base repair using the image guided endoscopic endonasal approach with no postoperative complications. The follow-up period ranged from 3 to 9 years, with a mean follow up period of 5.5 years. All of our patients were asymptomatic and recurrence-free during the follow-up period, with 100% success rate.

CONCLUSION: Careful history taking, endoscopic examination and radiological investigations are important for accurate diagnosis and management of pediatric sinonasal meningoencephaloceles. The endoscopic endonasal approach is the method of choice in repairing anterior skull base defects in pediatric patients. The advantages of this approach are direct visualization, minimal invasiveness, and safety, with low morbidities and short hospital stay.

KEYWORDS: Defect, Encephalocele, Endonasal endoscopic, Meningoceles, Skull base.

INTRODUCTION

Skull base defects are considered one of the rarest conditions in children. It can be caused by acquired traumatic event whether iatrogenic or accidental, or can be congenital in nature.[1] Cephalocele is defined as extracranial protrusion of intracranial matter through a defect in the cranium and dura. It can be further subdivided according to its content into meningoceles, in which the protruding material consist of meninges and CSF, and meningoencephaloceles, in which the protruding structures consist of meninges, CSF and brain.[1,2]

Meningoencephaloceles slightly predominate in females, and its incidence is approximately 1 in 6000 live births in Asian population.[3] Sinonasal meningoencephaloceles mostly present as clear CSF leak, while other symptoms including progressive nasal obstruction, snoring or nasal discharge have been reported.[4] Due to the possibility of fatal complications such as meningitis, pneumocephalus, and brain abscess, surgical intervention is almost always required for treatment of sinonasal meningoencephaloceles.[5]

The aim of this study was to assess our experience with the endoscopic endonasal management of pediatric sinonasal meningoencephaloceles and anterior skull base defects in a tertiary referral hospital.

PATIENTS AND METHODS

We conducted a retrospective consecutive case series analysis involving 6 pediatric patients who were referred to our hospital with sinonasal meningoencephaloceles, and who underwent endoscopic endonasal excision of meningoencephaloceles and skull base reconstruction in the period between 2010 and 2020. Patients' demographics, clinical presentation, etiology, location and side of defect, and reconstruction methods were all recorded. Postoperative complications, recurrences, and follow up records were evaluated as well.

This study was conducted at the department of ear, nose and throat (ENT) at King Fahad Specialist Hospital-

Correspondence:
Shatha AlGhamdi
College of Medicine, King Faisal University, AlAhsa, SAUDI ARABIA
Email: ishathaalghamdi@gmail.com

Dammam, Saudi Arabia, which is a tertiary referral hospital covering 4 million population of the eastern province of Saudi Arabia. Prior to commencing the study, ethical approval was obtained from the institutional review board of King Fahad Specialist Hospital-Dammam, and written informed consents were obtained from patients' guardians.

A careful history taking and clinical examination in addition to in-office endoscopic examination was performed for all patients. All patients underwent preoperative imaging using contrast enhanced computed topography (CT) and magnetic resonance imaging (MRI). All of our patients were managed with image guided endoscopic endonasal excision of their sinonasal meningoencephaloceles and skull base repair by a senior rhinologist and a skull base surgeon. Intraoperatively, topical fluorescein dye was used in 2 out of 6 patients; the advantages of this dye are the intraoperative confirmation of the CSF leak, the delineation of the defect and tight seal of the reconstructed skull base. No intrathecal fluorescein dye was used in any of our pediatric patients. Three of our patients were repaired with multilayer technique using septal bone as an underlay, mucosal graft as an overly and supported with gelfoam. The other 3 patients were managed by single layer repair with mucosal graft that was supported with tissue glue and gelfoam.

All patients were kept in the hospital for 3-5 days postoperatively for observation. Postoperative antibiotics were given prophylactically for 7 days. No lumbar drains were used in any of our patients. Patients were followed up by serial outpatient department (OPD) visits and endoscopic examinations, one week postoperatively, one month postoperatively, every 4 months for the first year, then annually. The plan was to perform imaging as needed but it was never required.

RESULTS

Six patients were included in this study, 5 males and one female, and age ranging from 2 months to 15 years (Mean age: 10.8 years). Interestingly, our patients had various presentations; 2 patients had nasal watery clear discharge, nasal obstruction and upon in-office endoscopic examination a nasal mass was noted, 3 patients had recurrent meningitis and CSF rhinorrhea, and one patient presented with single episode of meningitis and CSF rhinorrhea. Table 1 summarized the clinical characteristics of our patients including age, gender, clinical presentation, etiology, site and side of the skull base defect, material and method of reconstruction, and follow-up period. There were no postoperative complications nor recurrences appreciated in any of our patients, during a mean follow up period of 5.5 years following surgical repair.

Illustrative cases:

Case 1

A 2 months old baby boy referred to our hospital with unilateral nasal mass obstructing the left nasal cavity. On examination the left nasal cavity was filled with a soft tissue mass obstructing the nasal passage. CT scan of the sinuses showed a large soft tissue mass filling the left nasal cavity protruding through a large skull base defect **(Fig. 1a).** Confirmation of congenital left ethmoidal meningoencephalocele was done by MRI **(Fig. 1b).** The child underwent an endoscopic endonasal excision of the left large ethmoidal meningoencephalocele with multilayer skull base reconstructions with free grafts, with the aid of image-guided surgery **(Figs. 1c,1d,1e).** There were no intraoperative or postoperative complications. The patient remained symptoms free with no recurrence after 6 years follow-ups.

Case 2

A 15 years old boy presented to our clinic complaining of right-side nasal obstruction and headache. Clinic endoscopic examination showed a large, pulsating, pale mass medial to the middle turbinate in the right nasal cavity **(Fig. 2a).** CT scan of the sinuses showed a large soft tissue mass occupying the right nasal cavity with right cribriform plate of ethmoid skull base defect **(Fig. 2b).** MRI confirmed a skull base defect with meningocele sac **(Fig. 2c).** An endonasal endoscopic approach was performed to excise the meningocele and reconstruct the skull base defect by using a single layer free mucosal septal graft as onlay technique **(Fig. 2d).** There were no intraoperative or postoperative complications. After 5 years follow-up, the patient is symptoms free with no recurrence **(Fig. 2e).**

Case 3

A 14 years old girl, a known case of craniosynostosis and dilated ventricles, presented to the clinic with active right nasal CSF leak and past history of recurrent meningitis. The patient was found to have a large right ethmoidal meningoencephalocele through fovea ethmodalis skull base defect on CT cisternography **(Fig. 3a)**. The patient underwent image-guided endonasal endoscopic excision of the meningoencephalocele **(Fig. 3b)** with the help of topical fluorescein dye to confirm the CSF leak **(Fig. 3c),** and repair by three layers skull base reconstruction. The first layer was septal bone underlay free graft, second layer was free mucosal graft and the third layer was middle turbinate pedicled rotation flap, as demonstrated in **(Figs. 3d,3e,3f,3g).** The patient is symptoms free after 5 years of clinical and endoscopic follow up.

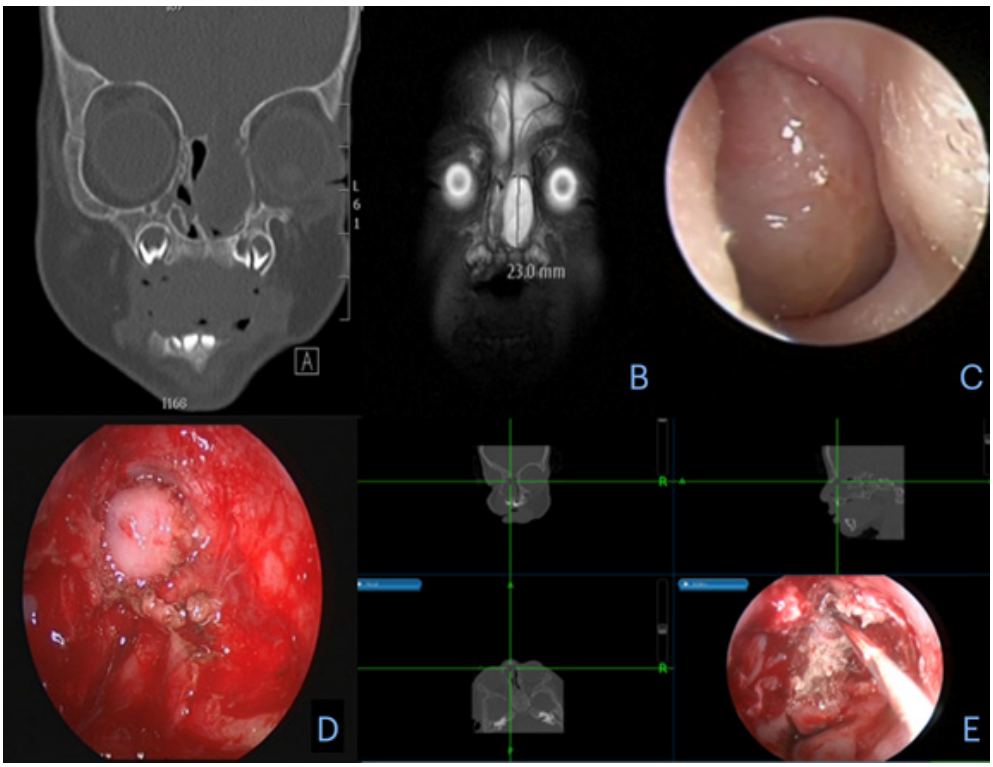

Fig 1: (A, B): Coronal CT and MRI scans of a left ethmoid encephalocele, (C): Endoscopic view of meningocele sac filing the left nasal cavity. (D): Ntra-op view of the reconstructed skull base defect with free graft, (E): Image guided surgery for accurate placement of graft and skull base reconstruction.

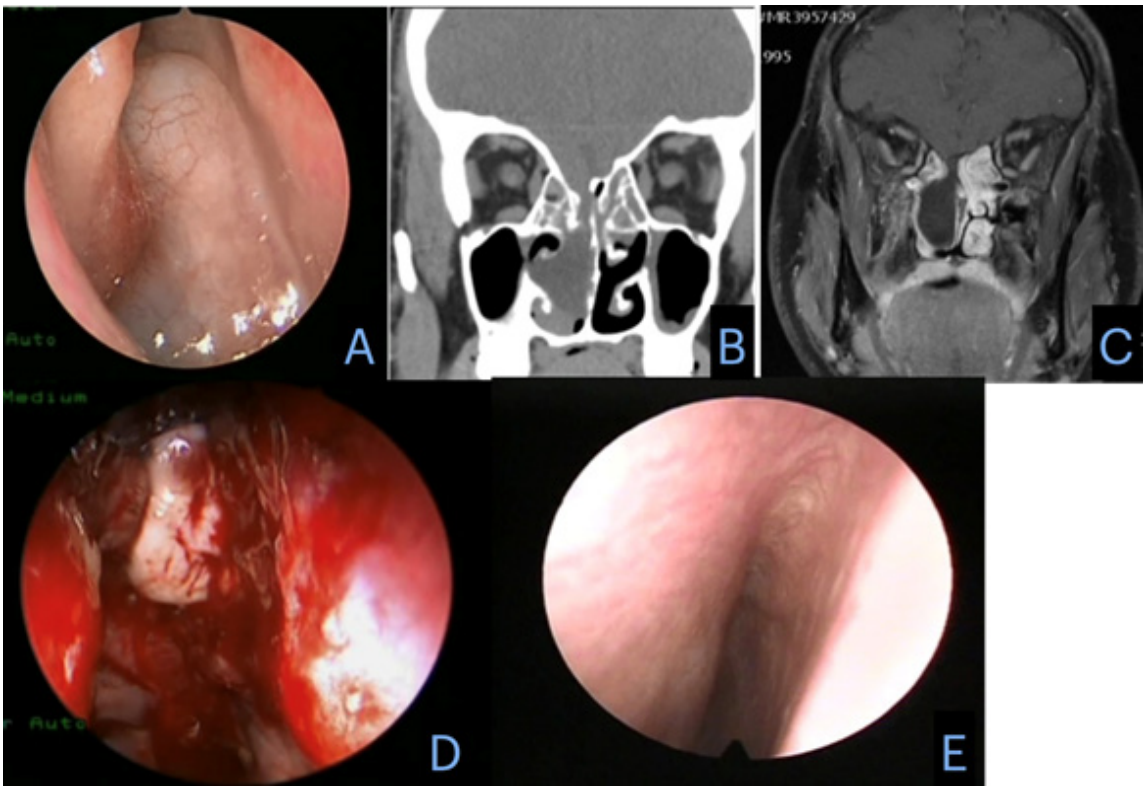

Fig 2: (A): Nasal endoscope shows meningocele mass medial to middle turbinate in the right nasal cavity, (B): A CT sinuses show a large meningocele filling nasal cavity with cribriform plate of ethmoid defect, (C): MRI sinuses meningocele filling nasal cavity with cribriform plate of ethmoid origin, (D) Intraoperative view skull base defect closure with septal mucosa onlay free graft, (E): Postoperative endoscopic view showing the healed skull base defect.

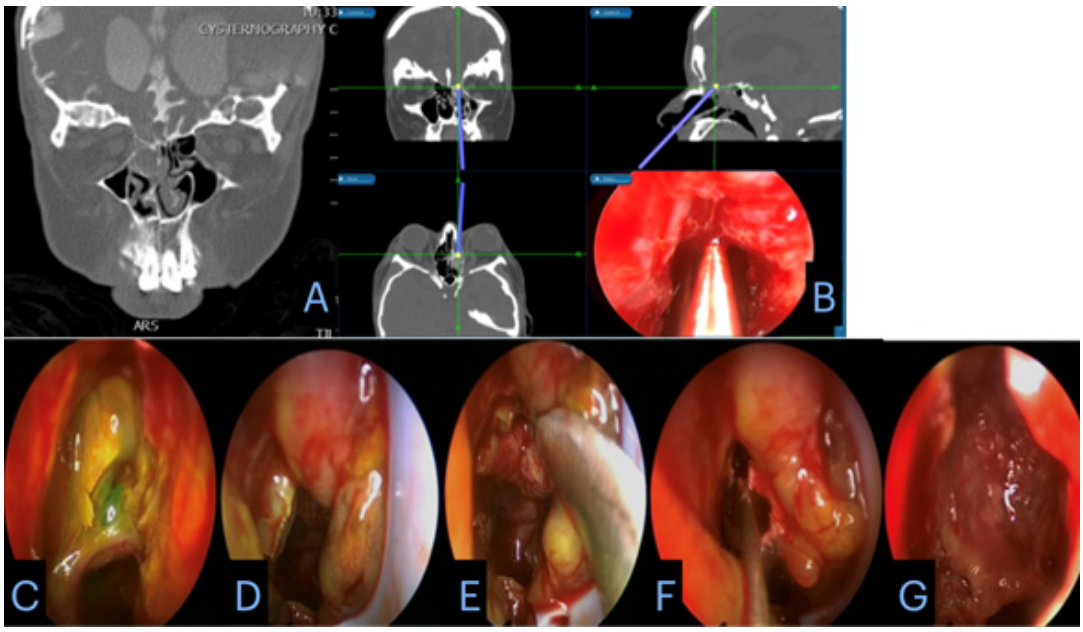

Fig 3: (A): Large right ethmoid meningoencephalocele on CT cisternography, (B): Image guided endoscopic endonasal excision and repair of skull base defect, (C): Intraoperative view of the meningocele with positive fluorescein dye of CSF leak, (D): First layer of septal bone underlay graft reconstruction, (E): Second layer of free mucosal graft underlay reconstruction, (F): Third layer middle turbinate pedicle rotation flap reconstruction, (G): Final cavity post excision MEC and three layers skull base reconstruction.

Table 1: Results and summary of 6 pediatric patients with skull base defect

Case	Age	Gender	Presentation	Etiology	Skull defect location	Side	Diagnosis	Graft	Follow up
1	2 months	Male	Nasal obstruction, nasal discharge and nasal mass	Congenital	Fovea ethmoid	Left	Nasoethmoidal meningoencephalocele	Multiple layers underlay & overlay	6 years
2	15 years	Male	Nasal obstruction, nasal discharge and nasal mass	Spontaneous	Cribriform plate	Right	Nasoethmoidal meningoencephalocele	Single layer mucosal free graft	5 years
3	14 years	Female	Recurrent meningitis & CSF rhinorrhea	Spontaneous	Fovea ethmoid	Right	Ethmoidal meningoencephalocele	Multiple layers underlay & overlay	5 years
4	12 years	Male	Recurrent meningitis & CSF rhinorrhea	Traumatic	Posterior wall frontal sinus	Right	Nasofrontal meningocele	Single overlay layer	9 years
5	14 years	Male	Recurrent meningitis & CSF rhinorrhea	Traumatic	Posterior wall frontal sinus	Right	Nasofrontal meningocele	Single overlay layer	5 years
6	15 years	Male	CSF rhinorrhea & meningitis	Traumatic	Fovea ethmoid	Right	Ethmoid meningocele	Multiple layers underlay & overlay	3 years

DISCUSSION

Cephalocele is defined as an extracranial protrusion of any intracranial structures through a defect in the dura or cranium. It can be subdivided into meningocele, which is the protrusion of CSF and leptomeninges, and meningoencephaloceles, which comprise of CSF, leptomeninges, and brain tissue. It can also be classified according to the etiology of skull base defect into congenital or acquired, the latter can be further divided into spontaneous and traumatic.[1,2] Another way to classify anterior skull base encephalocele is based on the location of the defect. There are sincipital encephaloceles which has three types; nasofrontal, nasoethmoidal, and nasoorbital, and basal encephaloceles which include four types; sphenoethmoidal, sphenoorbital, transsphenoidal and transethmoidal which is the most common type. Both can cause hypertelorism and external deformity of the nose.[3]

Clinical presentation of anterior skull base defects in pediatric age group varies and can manifest itself as nasal obstruction, CSF rhinorrhea, breathing disorder, headache, or in some instances it may remain asymptomatic.[3] Others may present with more critical situations as meningitis, as we have witnessed in four out of our six patients.

Diagnosis can be very challenging especially in pediatric patients. A high level of suspicion combined with endoscopic evaluation and neuroimaging is essential. MRI permits a detailed exploration and gives clear view of the vascularity of the herniated material. It also can help in differentiating meningoencephaloceles from other benign nasal masses such as nasal gliomas and nasal dermoids. CT allows best visualization for the bony anatomy of the skull base and can confirm the possibility of coexisting multiple skull base defects. Both MRI and CT should be obtained before endonasal endoscopic surgery is performed. When exposure to radiation is a serious concern in the pediatric age group, CT scan can be excluded but MRI is essential to be done before the operation.[4-6] Laboratory investigations are of paramount importance as well to confirm the diagnosis. Beta-2 transferrin has a high sensitivity rate of 98% in detecting CSF leak, however it mandates active rhinorrhea at the time of evaluation, which can be hard to obtain or collect in the pediatric age group.

Intraoperatively, intrathecal fluorescein can aid in identification and reconstruction of multiple skull base defect sites as highlighted by Castelnuovo et al. study.[4] There were no complications related to the injection of fluorescein when the dosage was precisely constituted and the intrathecal lumbar route was adopted. The technique should be considered safe.

From a surgical point of view, many approaches have been proposed. Conventional anterior skull base surgery in children can potentially cause disruption of the growth centers in the craniofacial skeleton that may result in facial asymmetry.[7] On the other hand, endoscopic endonasal approach achieves definitive repair of most anterior and middle skull base CSF leaks.[8] Furthermore, it minimizes the surgical scars on the brain tissue, helps to preserve the nasal and sinuses physiology and spares the growth and development centers of the facial skeleton.[9,10] In addition, it permits a short inpatient time. The safety of endoscopic endonasal surgeries are well established in the literature.[6] Although endoscopic endonasal surgery (EES) is historically less preferred in pediatric age group, several studies exist that show these restrictions are minimal and that most pediatric patients' nasal size is adequate for the use of standard endoscopic equipment.[11] However, a well skilled surgeon is essential for better outcome.[12]

A systematic review compared EES to open transcranial and transfacial approaches for managing encephaloceles, meningoceles and CSF leakages. The review found that there was no significant difference between repair success rates, but that complications and perioperative mortality were significantly lower in the EES cohort.[13] Few publications exist highlighting cases of purely endoscopic repair of sinonasal meningoencephalocele.[4,14-19] Of these, Tabaee et al. and Castelnuovo et al. report the largest case series, with 92% and 100% success rates on 13 patients and 11 patients, respectively.[4,14]

As noted in the literature, congenital or spontaneous defects are usually much larger than the traumatic defects hence the use of multilayer technique are usually needed.[11] This comes in agreement with our experience, where skull base reconstruction of small traumatic defects are ideally managed by a single layer overlay graft harvested from the nasal septum or the inferior turbinates. On the other hand, larger congenital or spontaneous skull base defects are managed by multilayers skull base reconstruction by using the septal bone as underlay followed by free mucosal overlay graft, and finally a pedicled middle turbinate rotational flap as the final layer of reconstruction.

CONCLUSION

In our hands, endoscopic endonasal skull base repair is the favorable approach in management of pediatric anterior meningoencephaloceles. It has the advantages of being a direct minimally invasive technique, with a short hospital stay, minimal morbidities, and 100% success rate.

List of abbreviations

CSF: Cerebrospinal fluid.
CT: Computed tomography.
EES: Endoscopic endonasal surgery.
ENT: Ear, nose and throat.
MRI: Magnetic resonance imaging.
OPD: Outpatient department.

Disclosure

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