American Journal of Otolaryngology and Head and Neck Surgery

Research Article
Published: 10 May, 2021

The Endonasal Endoscopic Management of Pediatric CSF Leak and Meningoceles

Ali Al Momen, Ahmed Aldandan, Jamal Alhamam, Hussain Alsheef, Amirah Aldhurais, Balsamalawami, Salma Alhammad, Hussain Almulla, Zahra Almoumen and Fatima Alhassan*

Department of ENT, King Fahad Specialist Hospital, Saudi Arabia

Abstract

Introduction: The diagnosis and management of skull base defects in pediatrics are challenging. These defects can be con-genital or traumatic. Congenital lesion includes encephalocele which is a herniation of meninges, CSF and brain through a skull defect. The aim of this study is to present our experience with the endonasal endoscopic management of 9 pediatric patients presented with congenital encephalocele and traumatic CSF leakage.

Methods: A retrospective study of 9 pediatric patients (age ranging from 2-months to 15-years old) was conducted at our tertiary care center between the years 2010 to 2019. Three patients had congenital encephalocele while four children presented with traumatic CSF leakage. After detailed clinical evaluation and pre-operative imaging with CT and MRI scans, an endonasal endoscopic repair was carried out in all cases.

Results: All patients had a successful repair and recovery, without any post-operative complications. The follow-up period ranges from 1 to 9 years. All nine patients remain asymptomatic and recurrence-free.

Conclusion: The endonasal endoscopic approach is the preferred method for repairing skull base defects in pediatric patients. Compared to conventional operations, this endoscopic procedure is minimally invasive, efficient, and safe. A multidisciplinary team with experienced endoscopic nasal surgeon is required to perform this surgical procedure.

Abbreviations

CSF: Cerebrospinal Fluid; MEC: Meningoencephalocele; CT: Computed Tomography; MRI: Magnetic Resonance Imaging

Introduction

Pediatric skull base defects constitute a rare clinical entity [1]. The diagnosis and management of these defects in pediatrics pose a significant challenge [1,2]. Causes of skull base defects include congenital as encephalocele or traumatic skull base fracture [2].

Cephaloceles defined as extra-cranial herniation of any intracranial contents through a defect in the cranium or dura [3]. These lesions are classified based on their contents and location. Based on the herniated con-tents, cephaloceles are divided into two types: Meningocele which contain meninges and CSF only, and meningoencephalocele or encephalocele which contain meninges, CSF and brain tissue [3].

The epidemiological distribution of encephalocele has a worldwide variation. The incidence in the Southeast Asian countries is around 1 in 5,000 to 6,000 live births compared to 1 in 35,000 to 40,000 live births in the Western countries [4,5]. In Saudi Arabia, there are limited resources on the prevalence of encephalocele. However, several studies in different regions of Saudi Arabia had found a marked decline in the prevalence of neural tube defects including encephalocele [6,7].

A high index of clinical suspicion is required to diagnose pediatric encephalocele [1,2,8,9]. While encephalocele may be incidentally noted on imaging, clinical presentation can vary among patients [8]. The presenting symptoms include nasal mass, nasal obstruction, difficulty of breathing and feeding, CSF rhinorrhea and meningitis [8,9].

CSF rhinorrhea occurs due to a defect in the skull base bones, dura and arachnoid causing abnormal communication between subarachnoid cavity and nasal cavity [10]. There are two

OPEN ACCESS

Correspondence:
Ali Al Momen, Department of ENT, King Fahad Specialist Hospital, Dammam, MOH, Saudi Arabia, E-mail: alihalmomen@yahoo.com

Received Date: *20 Apr 2021*
Accepted Date: *06 May 2021*
Published Date: *10 May 2021*

Citation:
Al Momen A, Aldandan A, Alhamam J, Alsheef H, Aldhurais A, Balsamalawami, et al. The Endonasal Endoscopic Management of Pediatric CSF Leak and Meningoceles. Am J Otolaryngol Head Neck Surg. 2021; 4(4): 1132.

main causes of CSF leakage; traumatic and non-traumatic, with spontaneous leak being the most common cause [11]. Early closure of CSF leak is essential to avoid risk of meningitis [11].

The treatment of choice for skull base defects is surgical repair [4,8]. In the past, transcranial approach was used as the standard technique of surgical repair [4,8]. However, this approach was associated with a high risk complication [4,8,9]. In 1981, Wigand has reported the first successful endonasal endoscopic repair of CSF leak [4,8]. Thereafter, endoscopic approach has developed and becomes the preferred method of skull base defect repair [4,9].

Materials and Methods

A retrospective analysis involving 7 pediatric patients who underwent endonasal endoscopic repair of CSF leaks and encephaloceles between 2010 and 2019. This study was conducted at the department of ENT at King Fahad Specialist Hospital-Dammam, Saudi Arabia. Seven patients were included in the study, five male and two female, whose ages ranged from 2-months to 15-years old (mean age 10.8 years). The follow up period ranged from 6 months to 9 years.

Patient Evaluation

A detailed history and clinical evaluation with specific attention to the duration of symptoms, mode of onset, associated congenital malformations, features of raised intracranial pressure and meningitis. Nasal endoscopic examination was performed for all patients. Three children, 2 males and 1 female had congenital encephalocele. A 14 years old girls is a known case of Crouzon syndrome had previous history of encephalocele which was treated at other hospital. Three other male children and one female had previous trauma. The presenting symptoms and signs included CSF rhinorrhea (5 cases), recurrent meningitis (4 cases) and nasal obstruction (2 cases).

For the radiological evaluation, a Computed Tomography (CT) scan and a Magnetic Resonance Imaging (MRI) of the brain & paranasal sinuses were done to localize the defect in the skull base, to decide the surgical exposure necessary, and to rule out any associated pathologies. CT cisternogram was performed in one patient with rhinorrhea to localize the leakage site.

Following a full clearance for general anesthesia and consent from the child's guardian, an endonasal endoscopic repair of skull base defects was performed in all patients. Systemic antibiotic was administered to all children for 7 days starting from the time of surgery.

Operative Procedure

All skull defects were repaired using rigid endoscopes and micro-instruments. Surgical procedure was performed under general anesthesia with patient on supine position and slight head extension. Adrenaline soaked tampon were applied intranasally to minimize bleeding. After removal of tampon, local anesthesia injected in the submucosa for easier dissection of the mucosa from cartilage or bone.

Adequate exposure of the entire skull defect is crucial. Following release of mucosal adhesion, encephalocele was resected meticulously using bipolar cautery. Then, the repair was designed based on the defect and intra-operative circumstances. Several materials were used for grafting including mucosal free graft from inferior turbinates, nasal septum and middle turbinate bone. Single layer free graft was used for skull defect reconstruction in 5 patients, while multilayered reconstruction was used in the remaining 2 patients. Finally, the graft fixed into position with fibrin glue supported by absorbable hemostatic sponge and nasal pack.

Postoperative Care

All patients received intravenous antibiotic postoperatively for 5 to 7 days. The nasal pack was removed 48 h to 72 h after the surgery. One of our patients required a post-op lumbar drain. This patient had multiple defects, high pressure leaks and multiple encephaloceles. The drain was removed 7 days after the surgery.

All measures were taken to prevent elevation of intracranial pressure. After discharge, patients underwent endoscopic evaluation and followed up at a range from 6 months to 9 years. Imaging was reserved for patients with history of rhinorrhea, meningitis, or seizures in the postoperative period.

Results

Among the 9 pediatric patients, 5 had CSF rhinorrhea, 4 had a history of meningitis, and 2 had symptoms of nasal obstruction. One infant with nasal obstruction was identified by symptoms of nasal obstruction, oral breathing, feeding difficulties, and nasal discharge. The clinical characteristics, including age, symptoms & signs, etiology, and location of the skull defects are shown in Table 1.

Five patients had skull base defects in the form of meningoceles and meningoencephaloceles (2 meningoceles and 3 meningoencephaloceles). On endoscopic evaluation, the two patients with unilateral nasal obstruction had a unilateral polypoidal mass protruding into the nasal cavity. One patient with Crouzon syndrome who presented with rhinorrhea and recurrent meningitis had a cystic, grayish mass filling the nasal cavity.

Four children with traumatic skull base lesions presented with unilateral rhinorrhea, and 3 of them had re-current episodes of meningitis. On endoscopic examination, all patients were found to have a normal anatomy. All patients with active rhinorrhea were tested for beta-2 transferrin and had positive results.

All patients underwent pre-operative CT scan and MRI, which revealed skull base defects in all cases. The skull base defect were successfully located in (the cribriform plate of ethmoid 4 cases, roof of anterior ethmoid 2 cases ,posterior frontal sinus wall 2 cases, cribriform plate and ethmoid bone 1 case). All patients had a successful repair with no post-operative complications or recurrence.

Discussion

Skull base lesions are rare and infrequently identified in children [4,8,9]. There are several theories describing the pathogenesis of encephalocele. The most acceptable thesis is the non-separation theory which is related to the abnormal development of embryological layers [12]. Any defect or disturbance in the separation process at the site of final closure of the rostral neuropore can lead to a persistent communication between neuroectoderm (brain) and surface ectoderm (epidermis of the skin) [12]. This connection would prevent mesodermal formation at this site causing midline skull defect, which eventually leading to formation of encephalocele [12].

Encephaloceles are classified according to their location as occipital, cranial vault, frontoethmoidal (sincipital), and basal encephaloceles; which are subdivided into transethmoidal, sphenoethmoidal, transsphenoidal, and frontosphenoidal, Table 2

Table 1: Results summary of 9 pediatric patients with skull base defects.

Case	Age	Sex	Presentation	Etiology	Location of skull defect	Side	MEC	Diagnosis	Graft	FU
1	2 M	Male	Nasal obstruction, nasal discharge & nasal mass	Congenital	Cribriform plate	Left	Yes	Nasoethmoidal MEC	Multiple layers mucoperichondrium free graft	5
2	15 Y	Male	Nasal obstruction, nasal discharge & nasal mass	Congenital	Cribriform plate	Right	Yes	Frontoethmoidal MEC	Single layer mucosal free graft	3
3	14 Y	Female	Recurrent meningitis & CSF rhinorrhea	Congenital	Cribriform plate & Ethmoid	Right	Yes	Anterior & posterior MEC	Multiple layers Underlay & overlay	4
4	12 Y	Male	Recurrent meningitis & CSF rhinorrhea	Traumatic	Posterior wall of the frontal sinus	Right	No	-	Single overlay layer	9
5	13 Y	Male	Recurrent meningitis & CSF rhinorrhea	Traumatic	Cribriform plate	Right	No	-	Single layer mucosal free graft	5
6	14 Y	Male	Recurrent meningitis & CSF rhinorrhea	Traumatic	Postero-superior wall of the frontal sinus	Right	No	-	Single overlay layer	3
7	8 Y	Female	CSF rhinorrhea	Traumatic	Cribriform plate	Right	No	-	Single layer mucosal free graft	1
8	10 Y	Male	CSF rhinorrhea	Idiopathic	Anterior Ethmoid roof	Right	No	MC	Excision, Single layer overlay free graft	2
9	15Y	Male	CSF rhinorrhea	Traumatic	Anterior Ethmoid roof	Right	Yes	MC	Excision, multiple layers underlay & overlay	2

M: Months; Y: Years; CSF: Cerebrospinal fluid; MCM eningocele, MEC: Meningoencephalocele, FU: Follow-up (years)

Table 2: Classification of encephaloceles based on the location of the skull defect.

I. Occipital	
II. Cranial vault	1. Interfrontal
	2. Anterior fontanelle
	3. Interparietal
	4. Posterior fontanelle
	5. Temporal
III. Frontoethmoidal/Sincipital	1. Nasofrontal
	2. Nasoethmoidal
	3. Naso-orbital
IV. Basal	1. Transethmoidal
	2. Sphenoethmoidal
	3. Transsphenoidal
	4. Frontosphenoidal / Spheno-orbital

[8,12]. Basal and frontoethmoidal encephaloceles collectively known as anterior encephaloceles [8]. The most common site is occipital which accounts for 75% of the cases [9].

Previous literature indicate that majority of pediatric encephaloceles are congenital and may present as nasal masses [4,8,9]. In our study, 2 out of 3 encephalocele patients had nasal masses associated with nasal obstruction symptoms. Traumatic lesions occur due to iatrogenic causes or accidental head injury [4,9]. Some patients are asymptomatic and diagnosed incidentally on imaging [1,8]. Others may present with nasal obstruction, mouth breathing, snoring, failure to thrive and CSF leak [8,9]. Delayed presentation of encephalocele includes meningitis, which appear on 1 out of 3 encephalocele patients in our series [8]. Frontoethmoidal and fronto-sphenoidal encephaloceles are frequently associated with external facial deformity and hypertelorism [3,12]. Basal encephaloceles can herniate into nasal structures causing airway obstruction [1,12].

The clinical manifestation of pediatric CSF leak include; clear watery nasal discharge, headache, and meningitis [10,11]. In the current study, all patients with traumatic CSF leak presented with clear rhinorrhea, and recurrent meningitis was a common presentation (3 out of 4 cases).

Endoscopic repair has many advantages over the traditional trans-cranial approach including; minimally invasive, faster recovery, short hospital stay, less recurrence rate, and avoiding permanent anosmia [8]. Based on the literature, the endonasal endoscopic approach has become the preferred modality of skull defects repair in adults with a very high success rate [1]. However, this cannot be simply implemented in pediatric patients as they have a narrow nasal cavity. Our study shows an excellent outcome in repairing pediatrics skull defects using endoscopic approach, which is in agreement with the emerging reports [1,4].

Conclusion

Skull base defects are rarely noted in pediatric patients. Diagnosis of these defects require high index of clinical suspicion, endonasal endoscopic examination, laboratory test and imaging studies. Preoperative CT and MRI permits excellent localization of the skull base defect. The definitive management is the endonasal endoscopic surgical repair. The endonasal endoscopic approach is safe, direct and efficient approach of re-pairing skull base defects in children with the advantages fast recovery, minimal morbidities and short hospital stay and avoidance of the external craniotomy approach with its morbidities.

References

1. Ma JH, Huang Q, Li X, Huang D, Xian J, Cui S, et al. Endoscopic transnasal repair of cerebrospinal fluid leaks with and without an encephalocele in pediatric patients: From infants to children. Childs Nerv Sys. 2015;31(9):1493-8.
2. Locatelli D, Rampa F, Acchiardi I, Bignami M, Pistochini A, Castelnuovo P. Endoscopic endonasal approaches to anterior skull base defects in pediatric patients. Childs Nerv Syst. 2006;22(11):1411-8.
3. Pianta L, Pinelli L, Nicolai P, Maroldi R. Cerebrospinal Fluid Leak, Meningocele and Meningoencephalocele. In: Maroldi R, Nicolai P, editors. Imaging in treatment planning for sinonasal diseases. New York, United States: Springer Publishing. 2015. p. 93-106.
4. Keshri A, Shah S, Patadia S, Sahu R, Behari S. Transnasal endoscopic repair of pediatric meningoencephalocele. J Pediatr Neurosci. 2016;11(1):42.

5. Mahapatra AK, Suri A. Anterior encephaloceles: A study of 92 cases. Pediatric NeuroSurg. 2002;36(3):113-8.

6. Seidahmed MZ, Abdelbasit OB, Shaheed MM, Alhussein KA, Miqdad AM, Khalil MI, et al. Epidemiology of neural tube defects. Saudi Medical J. 2014;35(Suppl 1):S29-35.

7. El-Awad M EH, Sivasankaran S. Neural tube defects in southwestern region of Saudi Arabia. Ann Saudi Med. 1992;12(5):449-50.

8. Marfatia HK, Parelkar KA, Chakraborty A, Mishra S. Pediatric meningoencephaloceles endoscopic endonasal repair: Our experience. Allergy Rhinol. 2018;9:215265671880240.

9. Stavrakas M, Karkos PD, Triaridis S, Constantinidis J. Endoscopic management of paediatric meningoencephaloceles: A case series. Eur Arch Otorhinolaryngol. 2018;275(11):2727-31.

10. Peng A, Li Y, Xiao Z, Wu W. Exploration of endonasal endoscopic repair of pediatric cerebrospinal fluid rhinorrhea. Int J Pediatr Otorhinolaryngol. 2011;75(3):308-15.

11. Psaltis AJ, Schlosser RJ, Banks CA, Yawn J, Soler ZM. A systematic review of the endoscopic repair of cerebrospinal fluid leaks. Otolaryngol Head Neck Surg. 2012;147(2):196-203.

12. Hoving EW. Nasal encephaloceles. Childs Nerv Syst. 2000;16(10-11):702-6.