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Endonasal Management of Congenital Transethmoidal Meningocele in a 3-Year-Old

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Abstract | **Case Report**

Meningoencephaloceles pose a complex challenge in both diagnosis and treatment, requiring a multidisciplinary approach for optimal management. This case report presents the clinical presentation, diagnostic workup, and management of a 3-year-old patient with a left ethmoidal meningoencephalocele. The patient exhibited symptoms of snoring and unilateral left nasal obstruction since early childhood. Imaging studies, particularly CT scan, revealed a left-sided brain herniation through a defect in the cribriform plate. Subsequent endonasal surgical repair was performed, resulting in a successful outcome with no postoperative complications. The discussion delves into diverse skull base reconstruction strategies and graft materials, underscoring the significance of meticulous surgical planning and comprehensive postoperative care.

Keywords: Meningoencephaloceles, Congenital, Pediatric Management.

INTRODUCTION

Meningoencephaloceles, characterized by protrusion of intracranial contents through a cranial defect [1], present a complex clinical scenario requiring careful diagnosis and management. These anomalies, while rare, necessitate a multidisciplinary approach involving neurosurgeons, otolaryngologists, and other specialists. They often present with symptoms such as nasal obstruction, rhinorrhea, and recurrent meningitis. Diagnosis typically involves imaging studies, including CT and MRI, to assess the extent of the lesion and associated anatomical abnormalities [2]. Historically, surgical treatment primarily involved craniotomy with anterior fossa defect reconstruction [3]. However, advancements in endoscopic techniques have revolutionized treatment options, offering reduced morbidity and improved outcomes [1].

This case report highlights the clinical presentation, diagnostic workup, and successful management of a pediatric patient with a congenital left ethmoidal meningoencephalocele.

CASE REPORT

A 3-year-old patient with no previous medical history presented with snoring and unilateral left nasal obstruction since early childhood. The obstruction was accompanied by ipsilateral clear rhinorrhea, without associated headaches, epistaxis, or otologic symptoms. Clinical examination revealed a negative Dandy sign, and rhinoscopy demonstrated a whitish mass filling the entire left nasal cavity figure (1). Ophthalmologic examination was unremarkable.

Initial CT imaging showed complete filling of the left nasal cavity by an oval mass with soft tissue density, measuring 25 x 14.5 mm. A bony defect at the cribriform plate, measuring 4 mm in width, allowed for herniation of brain parenchyma surrounded by a cerebrospinal fluid pocket, with displacement of the nasal septum to the right, confirming the diagnosis of left ethmoid osteomeningeal breach with meningoencephalocele figure (2).

Subsequent MRI confirmed a left ethmoid bone defect measuring approximately 4 mm, through which meningeal and cerebral contents filled the nasal cavity

and extended to the inferior meatus, consistent with a left ethmoidal meningoencephalocele. No other cranio-cerebral malformations were observed figure (3).

Surgical intervention was performed via an endonasal approach, involving cauterization and reduction of the meningocele, excision of the encephalocele, and closure of the defect using septal cartilage in underlay, followed by overlay with fascia lata and fixation with biological glue and Surgicel figure (4).

Postoperative follow-up was uneventful, with no ophthalmic or neuromeningeal complications. The patient underwent a depletive lumbar puncture on postoperative day 1 and removal of nasal packing on day 5.

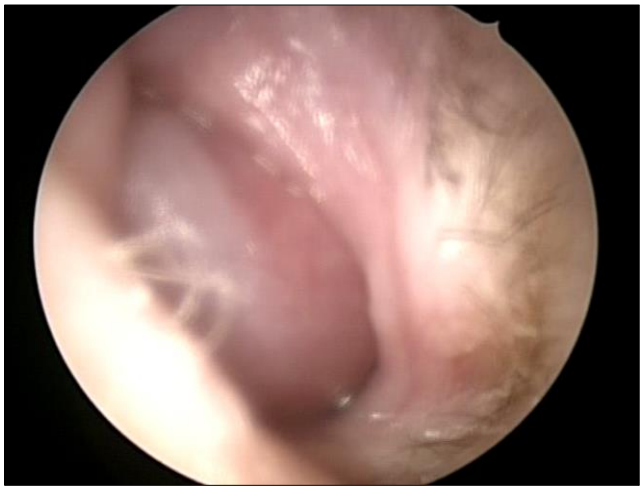


Figure 1: Endoscopic examination revealing the mass filling the entire left nasal cavity

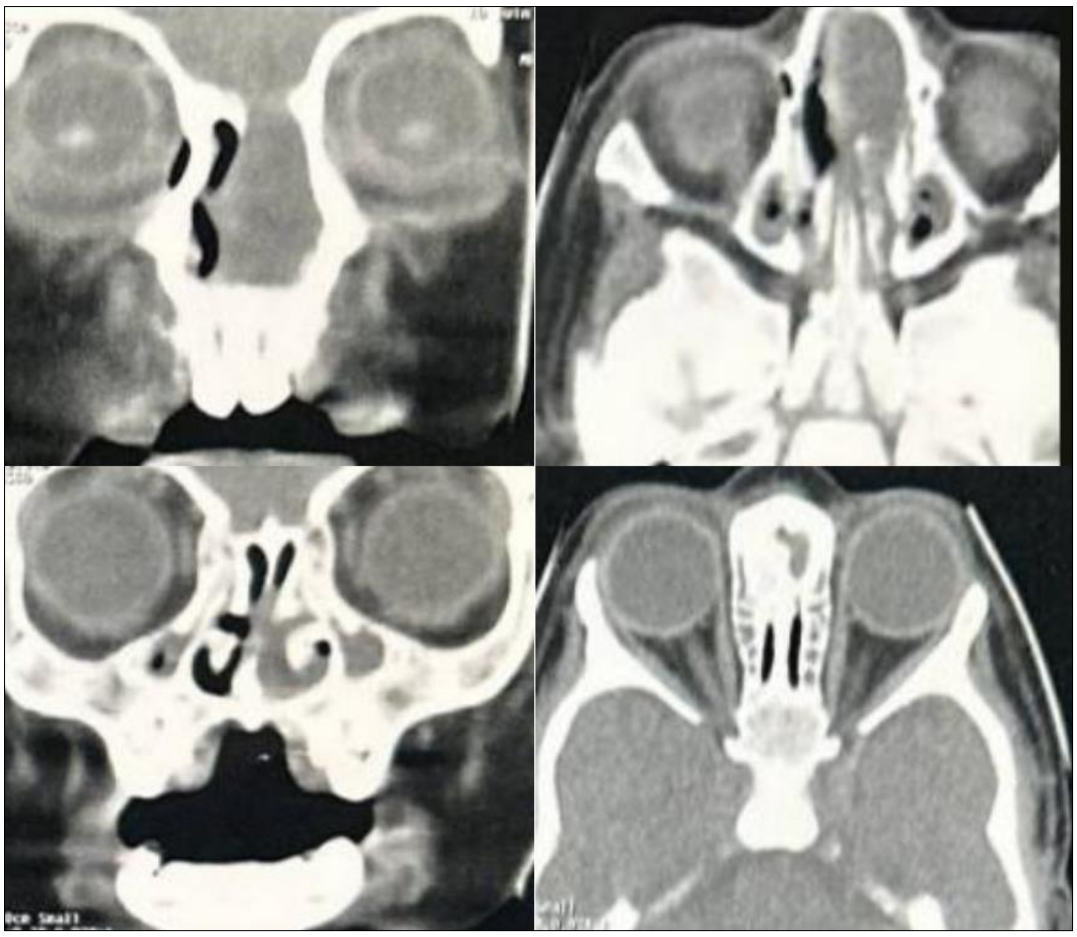


Figure 2: CT scan showing left ethmoidal meningoencephalocele and associated anatomical features

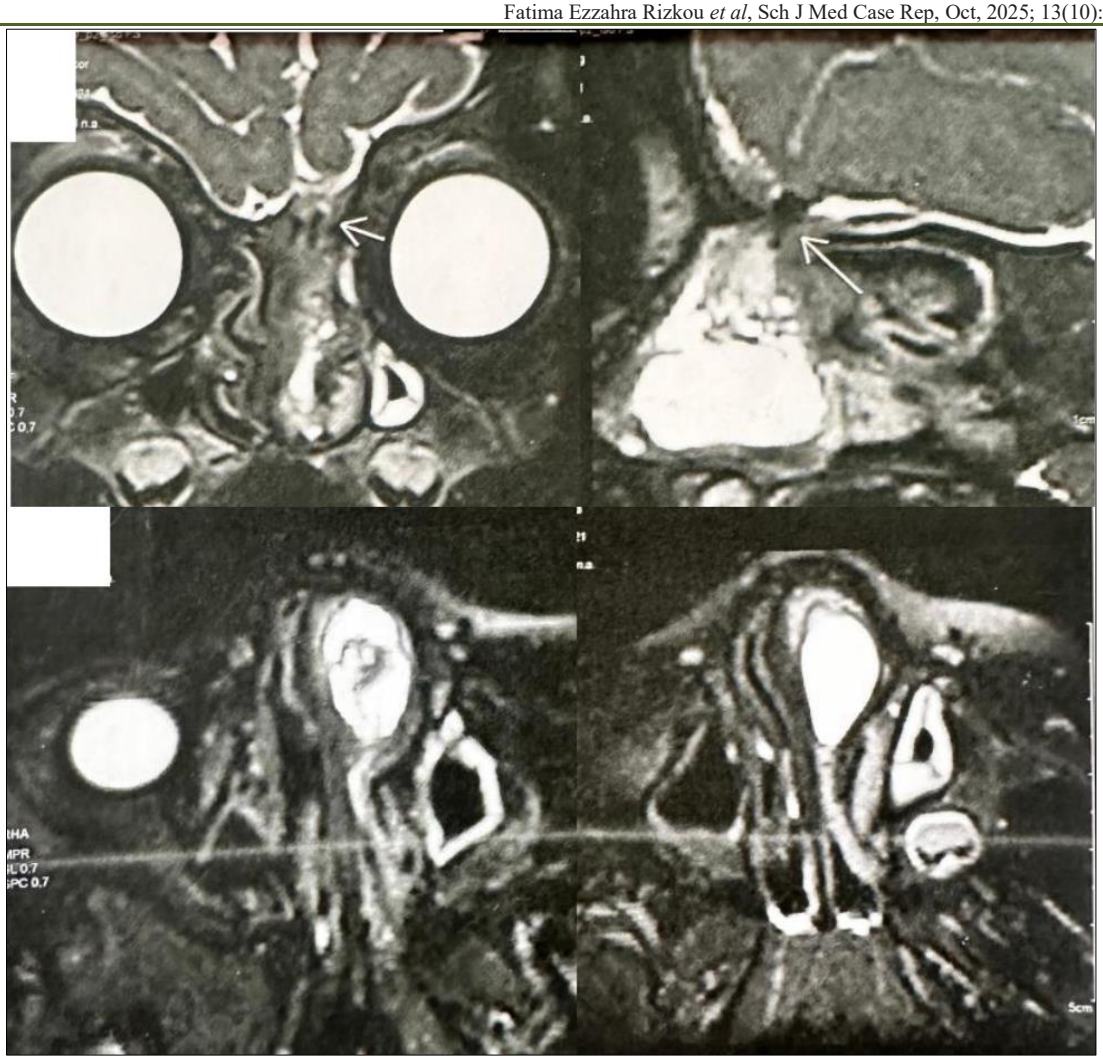
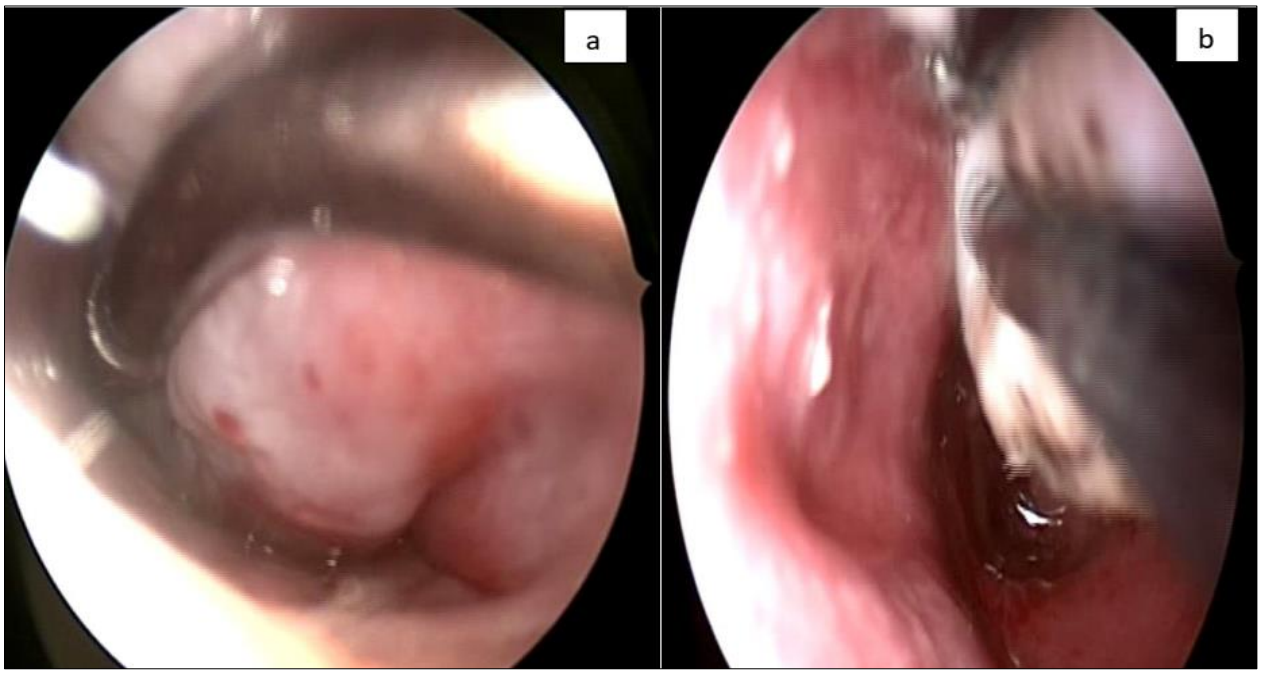


Figure 3: MRI imaging demonstrating left ethmoidal meningoencephalocele and adjacent structures



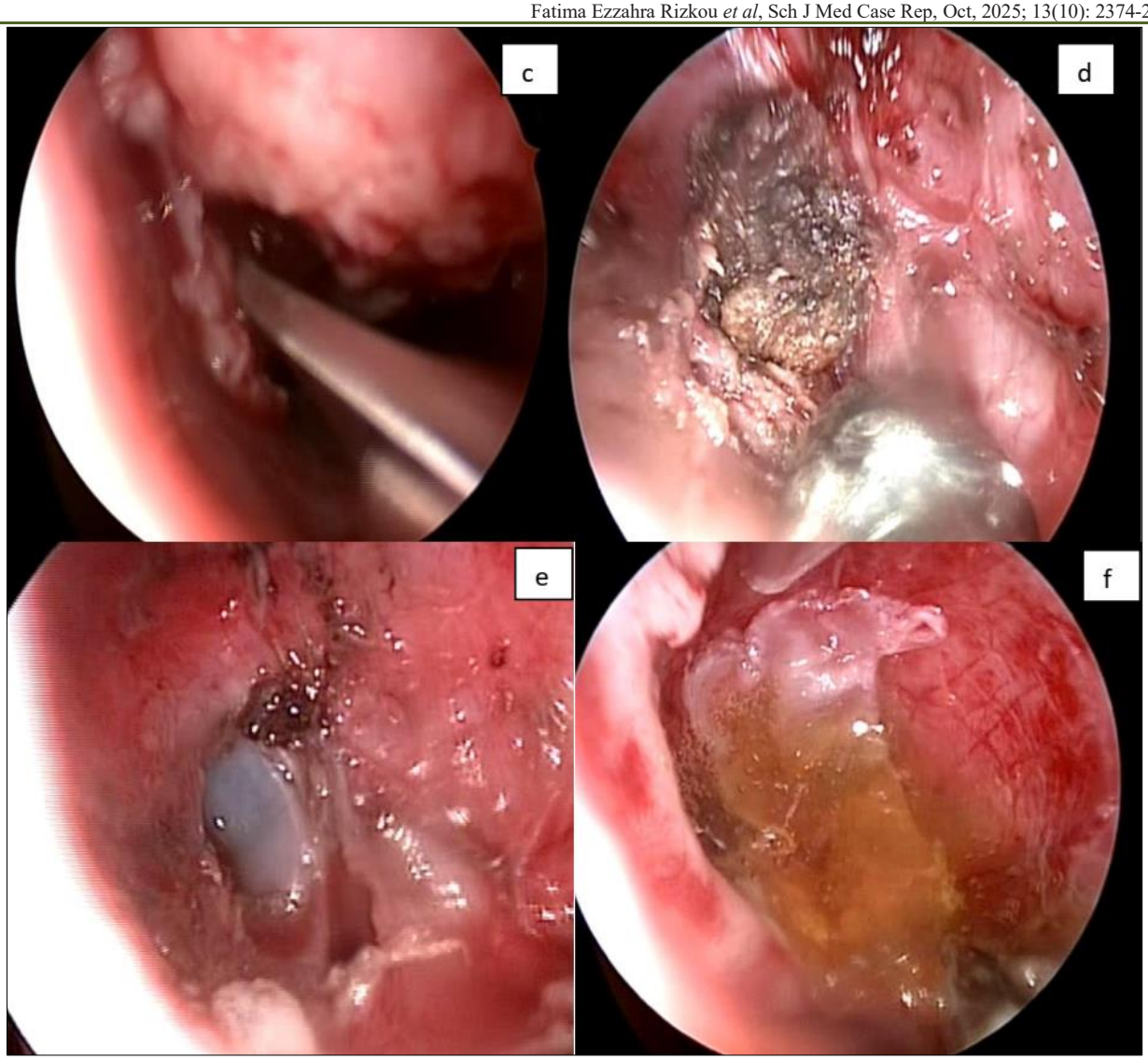


Figure 4: Intraoperative image depicting surgical repair of left ethmoidal meningoencephalocele: a: Intraoperative view showing the surgical the left ethmoidal meningoencephalocele. b: Cauterization of the surgical site during repair of left ethmoidal meningoencephalocele. c: Removal of surrounding mucosa. d: Peroperative view of the defect. e: Positioning of cartilage graft in underlay technique. f: Final view after application of glue, fascia lata

DISCUSSION

A congenital encephalocele is defined as a herniation of cranial contents through a cranial defect in patients without secondary causes [1]. Pediatric encephaloceles occur at an overall incidence of approximately 0.8–3.0 per 10,000 live births [2, 3].

Anterior encephaloceles are divided into frontoethmoidal and basal encephaloceles based on the location of the protrusion relative to the cribriform ethmoidal plate [2]. Among these, basal encephaloceles are particularly rare, accounting for only 1.5% of all encephaloceles, with an estimated incidence of 1 in 35,000 live births [4, 5]. Basal encephaloceles are further classified into five anatomical types: spheno-ethmoidal, trans-sphenoidal, trans-sphenoidal trans-sellar, spheno-orbital, spheno-maxillary, and trans-ethmoidal [4]. The trans-ethmoidal type constitutes only 8% of all anterior encephaloceles [5].

A congenital encephalocele is a late neurulation defect that results from disturbances in the separation of surface ectoderm and neuroectoderm in the midline around the 4th week of gestation [5]. This condition can be associated with other midline anomalies such as hypertelorism, broad nasal root, cleft lip, and cleft palate [5]. Numerous theories have been proposed to explain its etiology, defining it as a multifactorial condition influenced by both genetic and environmental factors that prevent the neural tube from fusing at the anterior neuropore [1, 2]. Potential causes include neural tissue overgrowth, viral infections, radiation, hyperthermia, hypervitaminosis, salicylates, trypan blue, hypoxia, and other agents [5]. Although the age at presentation varies

from early infancy to adolescence, very few cases are reported in the neonatal and infantile period [1].

Transethmoidal encephaloceles are challenging to diagnose early because basal encephaloceles herniate posterior to the cribriform plate, are present in the nasal cavity, and are not externally visible [1]. The age at clinical diagnosis is largely determined by the size of the encephalocele, related anomalies, and the presence or absence of respiratory difficulties [1]. Symptoms often include upper airway obstruction, snoring, feeding difficulties, pulsatile mass, endocrine abnormalities, recurrent meningitis, cerebrospinal fluid (CSF) rhinorrhea, or purulent discharge in the nasopharynx [1-4].

Consequently, these malformations are typically diagnosed during investigations for upper airway obstruction [3-5]. Endoscopic findings frequently reveal an intranasal mass, often unilateral and located over the middle meatus, with displacement of the nasal septum. Associated craniofacial anomalies can include cleft lip and palate, ocular anomalies, and craniosynostosis [6].

The differential diagnosis for these symptoms includes nasal polyp, allergic rhinitis, lacrimal duct cyst, nasal glioma, and nasal meningocele [1-5]. Biopsy is strongly contraindicated due to the risk of infection and meningitis [7].

Preoperative MRI and CT scans are crucial diagnostic tools, as they not only aid in evaluating associated brain abnormalities but also in delineating vital structures within the herniated sac, such as the pituitary gland, hypothalamus, optic pathways, and vascular anatomy. Given that diagnosis heavily relies on these imaging modalities, their role in surgical planning and postoperative monitoring becomes even more significant [5].

Multidisciplinary management is recommended for the diagnosis and treatment of meningoencephalocele. Currently, surgical interventions are the primary treatment approach, which may involve craniotomy with anterior fossa defect reconstruction or more recently; endonasal endoscopic resection and repair, often in combination [7]. Endoscopic methods, such as transpalatal and transnasal approaches, are increasingly preferred due to their lower morbidity and shorter hospital stays, without impacting facial development, allowing for early intervention with minimal risk of meningitis or CSF leaks [6].

During surgery, meticulous techniques involving bipolar cauterization are used to remove the meningoencephalocele up to the level of the skull base. The surrounding mucosa is then excised, and various skull base reconstruction techniques, including underlay and overlay methods, are employed [6, 7]. Multilayer repairs are preferred to address CSF leaks, reinforce the thin skull base, and prevent recurrence [1-6]. Various graft materials, both autologous (mucoperichondrial, fascia, cartilage, pericranial-galeal, and bone grafts) and heterologous (collagen matrix and acellular dermis), have been successfully utilized, with recent advancements including vascularized pedicled flaps; such as nasoseptal or posterior pedicle inferior turbinate flaps; promoting optimal healing and preventing complications from persistent communication between the cranial cavity and sinonasal tract [6, 7].

The choice of graft material is tailored to factors such as defect size, configuration, underlying pathophysiology, and surgeon preference [7]. Endoscopic repair has demonstrated success even in very young patients, with cases reported in infants as young as 21 days old [7]. A systematic review highlighted differences in intraoperative and postoperative outcomes based on patient age, with younger children (<2 years) showing higher rates of postoperative CSF leaks and meningoencephalocele recurrence, necessitating more frequent surgical reinterventions compared to older children [6].

Follow-up care is crucial as growth can alter skull-base geometry, emphasizing the importance of long-term monitoring and surveillance [6].

Conclusion

meningoencephaloceles remain a challenging entity, requiring a comprehensive management approach involving surgical expertise and careful postoperative surveillance. The evolution of endoscopic techniques has expanded treatment options, offering improved outcomes and reduced morbidity. However, differences in outcomes based on patient age underscore the importance of individualized care strategies. Continued research and collaboration among specialties are essential to further refine diagnostic and treatment modalities for meningoencephaloceles, ultimately improving patient outcomes and quality of life.

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