Case Report

A rare case of huge frontoethmoidal encephalocele projecting through mouth, with cleft palate

ABSTRACT

Frontoethmoidal encephalocele represents protrusion of meninges and brain in a sac through a defect in the anterior skull base, mostly as swelling over the nose. Rarely it is associated with facial dysmorphism and palatal cleft. There are various perioperative concerns like airway difficulties, leaking from the swelling causing fluid and electrolyte disturbances, risk of infection, compression of the swelling causing a rise in intracranial pressure, bleeding, hypothermia, etc., In neonates, these challenges rise exponentially because of the inherent difficulties in dealing with this group of patients. Frontoethmoidal encephaloceles are common in low socioeconomic strata and are often missed in the antenatal period. We are reporting a rare case of frontoethmoidal encephalocele, with a huge swelling protruding through the cleft palate and occupying more than 50% of the face and oral cavity, making mask ventilation impossible.

Key words: Encephalocele, frontoethmoidal encephalocele, neonatal difficult airway

Introduction

Encephalocele is a type of neural tube defect where a fluid-containing sac with meninges and brain tissue extends through a defect in the calvarium. Its incidence is 1 in 5,000 to 1 in 10,000 live births. It can be occipital, sincipital/frontoethmoidal, or basal encephalocele.[1] Frontoethmoidal lesions are more common in the South and Southeast Asian populations.[2] Anesthetic management of occipital meningoencephalocele has been reported several times, along with a few reports of frontoethmoidal encephalocele. We report this case of a rare frontoethmoidal encephalocele arising through the cleft palate, occupying more than 50% of the face and oral cavity, causing partial airway obstruction.

Access this article online	
Website: https://journals.lww.com/sjan	Quick Response Code
DOI: 10.4103/sja.sja_748_23	

Case

A seven-day-old male child weighing 2.1 kg, born at 36 weeks through normal vaginal delivery at a rural healthcare center in Rajasthan, with a huge frontoethmoidal encephalocele measuring approximately 7 × 7 cm from the cleft palate and occupying almost 50% of the face and oral cavity [Figure 1a-c]. The mother had lost follow-ups in antenatal clinics, and the antenatal scans were unavailable. The child had stable vitals and oxygen saturation of 97% at room air. The child had assumed a hyperextended neck position and left lateral head tilt. Systemic examination was normal, with no neurological deficit. Magnetic resonance imaging (MRI) brain revealed an anterior skull base defect at the level of the cribriform plate

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How to cite this article: Rathod D, Goyal N, Sharma A, Kumari K, Meshram T. A rare case of huge frontoethmoidal encephalocele projecting through mouth, with cleft palate. Saudi J Anaesth 2024;18:283-5.

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Submitted: 06-Sep-2023, **Revised:** 07-Sep-2023, **Accepted:** 07-Sep-2023, **Published:** 14-Mar-2024

through which bilateral frontal lobes appeared to herniate out along with cerebrospinal fluid (CSF), covered with meninges [Figure 1d]. The child had a cleft palate through which the sac protruded intraorally. All blood investigations were normal.

Informed consent was obtained from the parents. Inside the operating room, the American Society of Anesthesiologists recommended monitors were attached. Inj Atropine 20 mcg/kg was given intravenously for premedication. A difficult airway cart was kept ready, and high-flow oxygen was supplemented through a nasal cannula. As the mask placement was impossible because of the huge swelling, we planned for awake intubation in the lateral position, using TuoRen Videolarygoscope (Henan Tuoren Kingtaek Medical Device) blade size 0. Because of the difficulty in visualizing even epiglottis, the child was turned supine with neck extension and a shoulder roll. One resident supported and displaced the swelling. Using TuoRen video-laryngoscope, epiglottis was visualized, and posterior arytenoids were visible with backward, upward, rightward pressure. The intubation attempt failed as the child started desaturating, and it was difficult to negotiate the tube through the glottis. After oxygenation, a video-laryngoscopy with fiberoptic-guided intubation was tried. However, the bronchoscope could not be advanced through the glottis due to an active child. On the third attempt, with C-MAC video-laryngoscope (Kart Storz) blade 2 (because of the unavailability of neonatal sizes at our place), Cormack-Lehane grade 2 was achieved, and a stylleted uncuffed endotracheal tube, size 3, was positioned in the trachea.

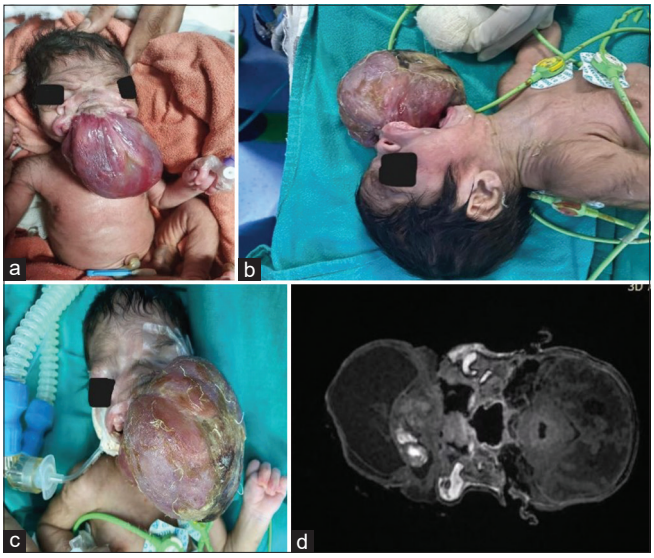

Figure 1: (a-c) Show the neonate with frontoethmoidal encephalocele with cleft palate. A swelling protruding through the mouth and obscuring half of the oral cavity. (d) MRI brain shows an anterior skull base defect at the level of the cribriform plate through which bilateral frontal lobes have herniated

The tube was fixed at the 10 cm mark after confirming bilateral equal air entry. The anesthesia plane was deepened with sevoflurane, fentanyl, and atracurium. There were no episodes of bradycardia during the procedure. Surgery was uneventful, with minimal blood loss. Postoperatively, the child was shifted to neonatal intensive care unit (NICU) intubated because of airway edema following multiple intubation attempts and intraoral surgery. On postoperative day 3, the child developed intestinal perforation for which he underwent exploratory laparotomy and eventually developed sepsis, and he succumbed on postoperative day 6 in NICU.

Discussion

A congenital encephalocele is a form of neural tube defect caused mainly by nutritional deficiency or use of teratogenic drugs and the presence of aflatoxins (ochratoxin A) in diet common in children of poor economic strata.[1] Around 85% are occipital encephalocele.[1] Frontoethmoidal encephalocele represents protrusion of meninges and brain in a sac through a defect in the anterior skull base, mostly as swelling over the nose.[2] Antenatally, neural tube defects can be diagnosed with maternal serum α fetoprotein levels, ultrasonography, and MRI.[2] An antenatal ultrasound in the second trimester detects 80% of the encephaloceles. MRI can provide a detailed description of the defect and presence of the brain, vasculature, their relationship, and any other central nervous system malformation.[2]

In developing countries like India, despite several ongoing health programs in rural areas, such congenital anomalies are common in low socioeconomic strata and often missed. The challenges for implementing national health programs include illiteracy, poverty, and poor medical infrastructure at primary and community healthcare centers, etc.[3]

Anesthesia management of occipital encephalocele has been described earlier. This is the first case of a huge frontoethmoidal encephalocele with a palatine defect leading to intraoral swelling with impossible mask-seal and ventilation. Leakage causing fluid and electrolyte disturbances, major blood loss, hypothermia, neonatal concerns, etc., are common perioperative concerns.[4] These patients pose challenges like airway difficulty, with nearly impossible mask ventilation because of the location of the swelling, difficult laryngoscopy and intubation, chances of encephalocele rupture, CSF leak, hemorrhage, and sudden hemodynamic collapse.[4]

We opted for awake intubation in the lateral position as the swelling could be displaced in this position. In neonates, the

airway reflexes are not developed like in bigger children.[5] Neonates respond differently to the stimulation of the airway. They tend to hold their breath and have central apnea or laryngospasm, leading to bradycardia. We premedicated the child with atropine to rectify the issue of bradycardia.

The second attempt was with TuoRen video-laryngoscope with FOB (hybrid method), and the last and most successful attempt was with blade size 2, C-MAC (Karl-Storz). This was not our first choice because of the nonavailability of neonatal blades. In the hybrid method, two different modalities are used so that the best of both methods can increase the chances of an attempt being successful.[6] However, our patient being neonate and awake, made those tiny movements for negotiating the bronchoscope through the glottis impossible.

We want to highlight that such cases are still common in developing countries with crippled healthcare systems in rural areas. This rare case requires a good preanesthetic examination and a set-up where proper planning and management of a difficult airway and perioperative challenges are possible.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Acknowledgments

We acknowledge Dr. Pradeep Bhatia, Professor and Head, and Dr. Nikhil Kothari, Additional professor, Department of Anesthesiology and Critical Care, AIIMS Jodhpur, for their constant guidance and encouragement.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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