SKULL-BASE MENINGOECEPHALOCELE PRESENTING AS A LABIOGNATOPALATOSCHIZIS AND BILATERAL MACROSTOMIA ON IMPENDING PARTIAL AIRWAY OBSTRUCTION IN A NEONATE

Nyoman Golden

ABSTRACT

A unique skull-base meningoencephalocele presenting as a labiognatopalatoschizis and bilateral macrostomia in a neonate is reported, with impending partial airway obstruction. Surgical management requires neurosurgical intervention and plastic reconstruction. This case report presented a term neonate who had several episodes of apnea related to partial airway obstruction by the mass. Computed tomography (CT) scan showed a large complex cystic and solid mass on lamina cribrosa of ethmoid bone. Focal calcification was seen within the mass. The brain on CT scan was normal with no dysmorphic structures. A transcranial approach by the neurosurgeon was performed to excise the cephalocele and close the dura mater. The bone defect on lamina cribrosa of ethmoidal bone was closed using peristeum tissue. The procedure was followed by total excision of the prolapsed brain tissue and osteotomy on the left palate and then nasal airway reconstruction continued with gradual reconstruction with external compression for close loopholes of the palate. Three months after the initial surgery, a defect of the palate was narrowing and with cranial nerve deficits. Skull-base transepthmoidal meningoencephalocele with labiognatopalatoschizis and bilateral macrostomia is a rare congenital abnormality. Neurosurgical procedures through transcranial approach are safe and provide excellent results. Moreover, gradual reconstruction will improve a patient's quality of life and activity of daily living.

INTRODUCTION

Respiratory distress in a neonate is more commonly caused by pulmonary, cardiac, or neurological aetiologies. The obstructive lesion of the upper airway is rare, unsuspected, and a potentially treatable cause of neonatal respiratory distress. An oropharyngeal mass presenting as a cause of neonatal respiratory distress is rare and may include bronchogenic cyst, epidermoid cyst, congenital cyst, and the least common basal meningoencephalocele.

Encephaloceles result from congenital openings in the midline region of the skull, often at the junction of the chondro- and desmocranium, with permit meninges, brain substance or both from the cranial cavity. Meningoencephalocele is a cerebrospinal fluid (CSF), and brain tissue or other glial matter filled hernia sac that is lined and covered by meninges. Meningoencephalocele represents 1.5–10% of all encephaloceles. Meningoencephaloceles may be subdivided into occipital, parietal, basal, and syncipital. Our rare reports of encephaloceles herniating through defects in the soft and hard palate with the bilateral macrostomia do not classically fit into the above classification scheme. The major predisposing factors are intranatal disorders, folic acid deficiency, or teratogenic factors. The complicated defect of the palate from the cell requires a gradual reconstruction from plastic surgery to optimize the growth. The disease is also rare in Sanglah General Hospital Denpasar, and this case report is a skull-base meningoencephalocele presenting as a labiognatopalatoschizis and bilateral macrostomia on impending partial airway obstruction.

CASE REPORT

A term neonate was noted at birth to have a soft, fixed, 5 cm mass full of his mouth with the impending partial airway obstruction. The neonate, who was otherwise healthy, had several episodes of apnea related to partial airway obstruction by the mass (Figure 1). Computed tomography (CT) scan showed a large complex cystic and solid mass on lamina cribrosa os ethmoidale. Focal calcification was seen within the mass. The extended mass resulting in the complete cleft of lip and palate, and bilaterally macrostomia. The disease is also rare in Sanglah General Hospital Denpasar, and this case report is a skull-base meningoencephalocele presenting as a labiognatopalatoschizis and bilateral macrostomia on impending partial airway obstruction.
hemostasis tests were within normal limits. The brain on the head CT scan was normal with no dysmorphic structures (Figure 2).

A transcranial approach surgery was performed by the neurosurgeon in order to excise the cephalocele and close the dura mater (Figure 3). The bone defect on the cribiform plate of ethmoidal bone was closed using periosteum tissue. The procedure was followed by total excision of the prolapsed brain tissue, osteotomy on the left palate, nasal airway reconstruction, and gradual reconstruction with external compression for close loopholes of the palate. Three months after the initial surgery, the defect of the palate was narrowing, and the patient resolved without any cranial nerve deficits (Figure 4).

**DISCUSSION**

Cephaloceles usually protrude from the skull along the suture line or at the junction of several bones. The connection with the cranial fossa can be broad-based, or pedunculated. Encephaloceles may contain recognizable organized protruded brain tissue or, as in this case, with more disorganized neural tissue with remnants of ependyma, choroid plexus, or glial tissue. The aetiology of meningoencephaloceles is still unknown. Regardless, they are occasionally associated with malformation syndromes, cleft palate, craniosenosis, agenesis of the corpus callosum, and brain-stem deformities.

Meningoencephaloceles of the skull-base are generally classified into five major anatomic types: frontoethmoidal, sphenoorbital, sphenomaxillary, temporal, and nasopharyngeal. Basal meningoencephaloceles are the rarest form, occurring with an approximate frequency of 1 in 35 000 births. The basal nasopharyngeal encephaloceles are further subdivided into the transethmoid, sphenethmoid, sphenonasopharingeal, and...
basioccipital-nasopharyngeal subtypes.1 Our rare case reports of encephaloceles herniating through defects in the soft and hard palate with the bilateral macrostomia do not classically fit into the above classification scheme. The basal cephaloceles, in particular, are more likely to be associated with midline cerebral anomalies.2 In general, the cephaloceles with better prognoses are less than 5 cm in diameter, do not contain dysplastic tissue, and are not associated with hydrocephalus or anomalies.

Computed tomography (CT) scan showed a large complex cystic and solid mass on lamina cribrosa os ethmoidale. Focal calcification was seen within the mass. The mass extended resulting in a complete cleft of lip and palate, and bilaterally macrostomia. The brain on CT scan was normal with no dysmorphic structures. In treating cysts, surgeons must prepare for the necessity of the opening the subarachnoid space and performing a duraplasty, as the lesion may communicate with the intracranial cavity. The same possibility exists in the patient who presents with a rounded, firm or tense swelling on the palatal dorsum, which may be meningocele. Total excision of the mass can be exposed through a transpalatal intradural approach, reduced into the cranium and secured with a pericranial flap was performed with the debulking and osteotomy on the left palate and nasal airway reconstruction, continued with gradual reconstruction with external compression for close loopholes of the palate with the plastic surgery. Three months after the initial surgery, the defect of the palate was narrowing. The histopathological result showed highly vascularized fragments of glossy white brain tissue and arachnoid, assert as the meningoencephalocele.

Overall management of this case between neurosurgery and plastic surgery is the best combination to include resection and reconstruction. Palate reconstruction phases were beginning at an early age after the eligible of “rule of ten”, started with labioplasty at 10 weeks, palatoplasty at 9-18 months, speech therapy management at 3-4 years, odontologic approach at 6-7 years, later with secondary lip and nose repair at 12-13 years.

CONCLUSION

Skull-base tranethmoidal meningoencephalocele with labiognatopalatoschizis and bilateral macrostomia is a rare congenital abnormality. Neurosurgical procedures through transcranial abnormality are safe and provide good results. Also, gradual reconstruction will improve a patient’s quality of life and activity of daily living.

REFERENCES


This work is licensed under a Creative Commons Attribution