Trans Sellar- Trans Sphenoidal Encephalocele Presenting as Nasal Obstruction in an Infant: a Rare Case Report

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Case Report

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Abstract

Trans sellar trans sphenoidal encephalocele is a rare congenital anomaly, with only around 10 cases having been documented in literature around the world. Surgical repair of these defects in the pediatric population commonly uses either the trans cranial or the trans palatal approach, with the choice of approach being individualized based on the clinical features, age, and associated defects present in the patient. Here we document a case of a 4-month-old child who presented to us with nasal obstruction, who was diagnosed with this rare entity and successfully underwent a transcranial repair for the same.

Introduction

The incidence of encephalocele ranges from 1 in 3000 to 1 in 5000 thousand live births [1]. The incidence of basal cephaloceles is thought to be 1 in 35,000 live births, making them rare anomalies. Based on the location of the defect in the bone, they have been further sub-classified as Trans ethmoidal, sphenoorbital, sphenomaxillary, and transsphenoidal encephaloceles. Trans Sphenoidal encephaloceles themselves are a rare entity, accounting for 5% of all basal encephaloceles (1 in 70,000 live births), with only a handful of cases having been documented around the world [2]. Trans sellar Trans sphenoidal encephalocele is the least common subtype, where less than 10 cases have been reported worldwide [3,4]. We hereby present a case of a 4-month-old child with this rare presentation, who was successfully operated on using the transcranial approach.

Case Report

A 4-month-old child, born at term as the first child of a non-consanguinous marriage. There was no history of maternal exposure to drugs or radiation. An antenatal scan at 5 months of intrauterine gestation revealed polyhydramnios and a cleft palate. He was born via Cesarean section at term with a birth weight of 3.35 kg, with midline facial defects including a cleft lip, submucous cleft palate, and nasal bone hypoplasia. Anterior fontanelle and cranial sutures were within normal limits. The child was born with normal male external genitalia. There were no focal neurological deficits at birth. An endocrine assessment revealed the child had congenital hypopituitarism and he was started on replacement steroid therapy.

The child now presented at 4 months, with complaints of persistent noisy breathing and obstructive apnea. Magnetic Resonance Imaging (MRI) brain showed a congenital absence of septum pellucidum and corpus callosum with herniation of meninges with CSF through a defect in the anterior cranial fossa, reaching up to the nasopharynx with a defect in the hard palate(Figure 1).

Computed Tomography (CT) Scan showed a bony defect from crista galli up to clivus with the absence of sellar floor (Figure 2).

A diagnosis of trans sellar transsphenoidal encephalocele was thus made and surgical repair was planned for the same. A transcranial approach was chosen and a bifrontal craniotomy was done. Sub-
frontal dissection was carried out extradurally to reach the defect in the skull base. The bony defect was exclusively in the sellar region. The dura was opened, the gliotic brain in the cavity was resected and the basal defect was closed with a bony graft which was harvested from the skull (Figure 3). The postoperative period was uneventful. There was no development of CSF leak, meningitis, endocrine dysfunction, or other complications. Cleft lip repair was performed later. Palatal repair is planned to be done at an appropriate age.

Discussion

The persistence of a craniopharyngeal or transsphenoidal channel is hypothesized to cause transsphenoidal encephalocele. It is a vertical midline skull base defect that extends from the sellar floor to the nasopharynx and has a diameter of typically less than 1.5 mm. The diagnosis is made in the first year of life for the vast majority of patients. However, in the absence of distinctive facial features, the diagnosis may not be made until adolescence or maturity [2].

Most transsphenoidal meningoencephaloceles are unearthed in the first year of life as a result of symptoms like respiratory distress brought on by pharyngeal or nasal obstruction, feeding issues, hypertelorism, a mass in the oral cavity, and endocrine anomalies [1,5]. A third of the patients have congenital anomalies such as hypertelorism, median nasal fissure, broad nasal root, and cleft lip or palate. Anophthalmia or microphthalmia, colobomas, retinal abnormalities, and morning glory syndrome, are among the optic malformations commonly observed. Corpus callosum agenesis occurs in approximately half of the afflicted individuals, as does hydrocephalus and pituitary hypoplasia. An association with Wolf-Hirschhorn syndrome has also been described in the literature [6,7,8].

Neuroimaging with high-resolution CT and MRI usually confirms the diagnosis and provides a clear picture of the contents in the herniated sac. An endocrine assessment is also essential, with the most common observed deficiencies being a deficiency of anti-diuretic hormone and growth hormone deficiency [2].

The surgical approach for the repair of this defect includes transcranial and trans palatal approaches and an endoscopic repair via the nasal cavity.

In adults, the endoscopic approach via the nasal cavity is the preferred route for lesion repair. This approach has been used on older children as well however, in the neonatal population, there are no documented cases of endoscopic management. Anatomical distortions of cleft lip and palate may however pose challenges in the trans nasal approach.

Other documented cases of neonatal cephalocele have been corrected using the transpalatal approach, especially if facilitated by a sufficiently large cleft palate. Its benefits include improved access (especially in cases of the associated cleft palate), improved cosmesis, and lower recurrence rates. By using the transpalatal approach, there is a lower risk of damaging the functional tissue in the encephalocele wall. The sac is easily dissected and reinserted into the cranium. This method, however, makes repairing the
skull base difficult. If a sufficiently large cleft palate is not present, palatal osteotomies must be performed and the hard palate removed. The closure of the mucosal layer and skull base reconstruction is difficult, often requiring skin and bone graft or silicone plate.

The transcranial route is the standard method of repair, as was chosen for our patient. The transcranial approach gives a more detailed view of defects and thus facilitates better repair. The repair of basal defect can be done with either an autologous graft or titanium mesh. However, transcranial repair of these lesions necessitates a craniotomy and carries a mortality rate of more than 50%. The most common complications are associated with the removal of functioning neural tissue, anosmia, intracerebral hemorrhage, and frontal lobe dysfunction.\[1,2,4\]. In our patient, the immediate postoperative period was uneventful, and the child continues to be under regular endocrine evaluation for hormonal therapy, including thyroxine supplementation.

**Conclusion**

Meticulous repair of defects and a multidisciplinary approach to repair other facial defects may help the child to lead a normal life. The positive outcome of our surgery leads us to support a transcranial approach for the repair of such lesions in the infantile age group. Imaging is an essential modality for diagnosis and planning of the surgical approach to be considered.

**Declarations**

- **ETHICS APPROVAL**

Ethics approval and consent to participate

Ethical approval was waived by the local Ethics Committee of Suez Canal University in view of the retrospective nature of the study, and all the procedures being performed were part of the routine care. The study was conducted in accordance with the declaration of Helsinki.

Written Informed Consent was obtained from the parent.

- The authors have no relevant financial or non-financial interests to disclose.
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- All authors certify that they have no affiliations with or involvement in any organization or entity with any financial interest or non-financial interest in the subject matter or materials discussed in this manuscript.
- The authors have no financial or proprietary interests in any material discussed in this article.
- Written informed consent for publication of their clinical details and/or clinical images was obtained from the parent of the patient. A copy of the consent form is available for review by the Editor of this journal.
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References


Figures
Figure 1

MRI BRAIN IMAGE SHOWING HERNIATION AND AGENESIS OF THE CORPUS CALLOSUM
Figure 2

CT IMAGING SHOWING BASAL DEFECT WITH ABSENT SELLAR FLOOR
Figure 3

POST OPERATIVE CT IMAGE SHOWING SUCCESSFUL REPAIR OF THE DEFECT