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Case Report

Trans-Sellar Trans-Sphenoidal Herniation of Third Ventricle with Cleft Palate and Microophthalmia: Report of a Case and Review of Literature

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Abstract

Trans -sellar trans-sphenoidal encephalocele is an extremely rare entity. We present the case of an 18-month old boy who presented with a trans-sellar, trans-sphenoidal encephalocele associated with cleft lip, cleft palate and microphthalmia. This patient was treated successfully by a trans-cranial extra-dural route. In this paper, we discuss the clinico-radiological findings as well as various surgical options in managing these rare lesions and briefly review the literature.

Keywords: *Basal encephalocele, cleft lip, craniopharyngeal canal, transcranial surgery, trans-sellar, trans-sphenoidal*

Introduction

The incidence of congenital encephaloceles in general is very low (approximately 1 in 3000–5000 live births).[1] Basal encephaloceles are the least common (1.5%) of all types of encephaloceles (with an incidence of 1 in 35,000 live births).[2] Basal encephaloceles are traditionally classified into five anatomic types: Sphenoethmoidal, trans-sphenoidal, spheno-orbital, transethmoidal, and spheno-maxillary.[3,4] Trans-sphenoidal variety is the rarest of all; only 19 cases has been reported in literature. We report a case of trans-sellar, trans-sphenoidal herniation of third ventricle floor, which was associated with cleft palate and microphthalamia in this report and present a review of the literature.

Case Report

An 18-month-old male child with right microphthalmia and complete corneal opacity, midline cleft palate and a repaired cleft lip presented to us with a cystic swelling protruding into his oral cavity. The child had undergone repair of his cleft lip at 10 months of age. Examination of the oral cavity demonstrated a wide complete cleft palate that allowed direct visualization of a completely compressible cystic swelling showing cough impulse.

Computed tomography (CT) head showed a defect in the floor of sella and sphenoid sinus through which the floor of third ventricle was herniating like a cerebrospinal fluid (CSF)-filled sac [Figure 1a and b]. Magnetic resonance imaging (MRI) of the head demonstrated the CSF-containing sac extending through a bony defect at the base of skull passing through the sella and sphenoid sinus into the oral cavity. This CSF-filled sac was actually the floor of the third ventricle which was herniating through the defect, probably containing the structures of the lateral wall of third ventricle, i.e., thalamus or hypothalamus proximally. In addition, both medial orbital walls were seen to be widely separated. There was agenesis of the corpus callosum [Figure 2a and b]. As the sella was affected, we carried out a complete battery of endocrine evaluation that revealed hypothyroidism and necessitated initiating oral thyroxine replacement.

After ensuring that thyroid functions were optimized, the child was taken up for transcranial repair of the encephalocele. Bi-frontal craniotomy with an extradural approach was used to access the third ventricle herniating into the sphenoid and posterior ethmoidal sinuses. The sac was opened to inspect its contents. The cavity was empty and seemed to be lined by ependyma indicating herniation of the infundibular recess and third ventricular floor through the bony defect; proximal part of the sac also contained part of the hypothalamus and thalamus. Excision of

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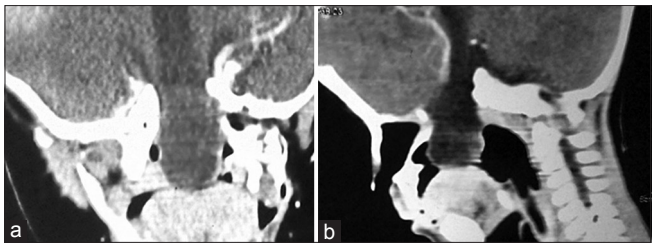

Figure 1: (a and b) Coronal and sagittal images of computed tomography head showing defect in the sellar floor and sphenoid sinus with herniation of the third ventricular floor into the oral cavity

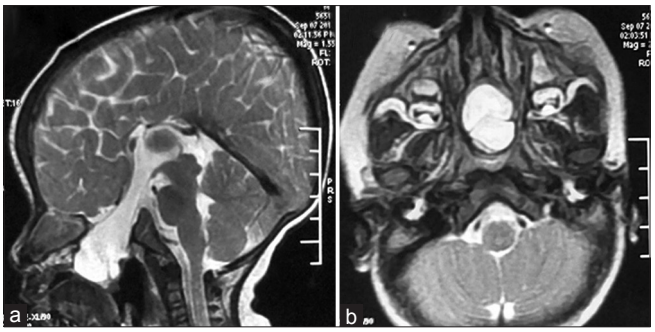

Figure 2: (a and b) Sagittal and axial images of magnetic resonance imaging brain showing herniation of cerebrospinal fluid filled sac through the defect in sellar floor and sphenoid sinus

the distal part of the sac with re-positioning of normal brain tissue was done and the remaining part of the sac was plicated. The cavity was obliterated with fat and the dural defect was sealed with fascia lata overlay graft, reinforced with small bone piece and fibrin glue. Postoperative CT scan showed complete obliteration of the sac. The patient was discharged on the 7th postoperative day and referred to plastic surgery for repair of the cleft palate. At 6 months follow-up, the child was doing well without any CSF leak.

Discussion

Trans-sphenoidal encephalocele is a rare variety of basal encephalocele. Trans-sphenoidal encephalocele can be congenital, iatrogenic, posttraumatic, or spontaneous in origin.[4,5] These encephaloceles are usually divided into two variants, namely, intra-sphenoidal and trans-sphenoidal depending on the relationship of the fundus of the sac with the floor of the sphenoid sinus.[5] In congenital varieties, this herniation of CSF-filled sac with or without cranial contents occurs through a defect in the sellar floor in trans-sellar type and through the persistent lateral craniopharyngeal canal (Sternberg canal) in the lateral type of trans-sphenoidal encephaloceles.[4,6,7]

Embryologically, multiple theories have been put forward to explain the formation of trans-sphenoidal encephaloceles, such as incomplete closure of neural tube, persistence of the craniopharyngeal canal, anomalies in the development of sphenoid bone and failure of the neuroectoderm to separate from the surface ectoderm during development of the neural tube.[5,8] Etiologies of the acquired type of sphenoidal encephaloceles include trauma, tumor, raised intracranial pressure due to any cause, intracranial infection and surgical procedures in and around the sella.[6]

Most patients present during childhood and these patients usually have associated craniofacial anomalies such as cleft lip and palate, facial hypoplasia, ocular deformities, craniosynostosis, and hypertelorism. At times the diagnosis can be delayed into adolescence and adulthood when these patients can present with CSF rhinorrhea, meningitis, endocrine dysfunction, or progressive visual deficits. In a child who presents with recurrent nasal obstruction this lesion can be confused with nasal polyps, in which case any intervention without proper evaluation can be disastrous. To the best of our knowledge, only 19 other cases of trans-sellar, trans-sphenoidal encephalocele have been reported in pediatric patients [Table 1]. Endocrine evaluation and neuro-ophthalmic evaluation are essential before any intervention. Hypothalamic–pituitary axis dysfunction is common, especially in trans-sellar type; growth hormone and antidiuretic hormone are most commonly deficient.[18] In our patient, hypothyroidism was detected and needed thyroxine replacement.

MRI of the brain is the best imaging modality for evaluating trans-sphenoidal encephalocele to determine their extent as well as their content. Determining the extent helps classify these lesions into trans-sellar and trans-sphenoidal varieties. MR angiography can supplement in identifying intracranial vasculature which may be herniating into the sac, which has important surgical implications. Trans-sphenoidal encephaloceles are more likely to contain viable neural structures such as hypothalamus, optic chiasm, pituitary gland and stalk and vessels of the circle of Willis.[19] CT scan head can help in identifying the bony defect in the skull base. As we found out during surgery, the infundibular recess of the third ventricle with the adjoining third ventricular floor were herniated, while the proximal part contained some hypothalamic tissue as well.

Multidisciplinary approach is required for the management of trans-sphenoidal encephaloceles.[11,16] Surgical repair can be done by transcranial, trans-palatal and trans-sphenoidal approaches. The surgical approach also depends on the size of the defect, degree of pneumatization of the sphenoid sinus and presence or absence of neurovascular tissue herniating through the defect. We preferred the transcranial approach for this patient because, with this approach, we were able to excise the sac and reposition the normal tissue and plicate the sac. For plication of the sac and repair of large bony defects, the transcranial approach is preferred; as without this there will be risk of CSF leak and sagging of brain tissue through the defect, which has potential risk of infection.[20] Some surgeons prefer the endoscopic trans-sphenoidal approach, but this approach has limitations in cases with large defects requiring skull base repair or with presence of neurovascular structures in

Table 1: Cases of trans-sellar trans-sphenoidal encephaloceles in pediatric patients reported in the literature

Author	Patient age	Presentation	Treatment
Mylanus *et al*.[9]	1 day	Respiratory distress	Trans-cranial and trans-oral
Narasimhan and Coticchia[10]	4 day	Respiratory distress	Trans-oral
Formica *et al*.[4]	8 months	Breathing difficulty, cleft lip and palate	Trans-palatal
Kahyaoglu *et al*.[11]	1 year	Progressive intraoral mass	Trans-cranial and trans-palatal
Franco *et al*.[12] (series of six patients)	6 months to 15 years	Intraoral mass with cleft lip and palate	Nasal endoscopy
Nishi *et al*.[13]	12 years	Hypopituitarism with diabetes insipidus and growth hormone deficiency	NA
Kumar *et al*.[14]	1 month	Intraoral mass with Respiratory distress	Trans-palatal
Steven *el al*.[15] (two cases)	5 months to 14 years	Nasal obstruction with intraoral mass	Trans-palatal
		Nasal obstuction	Trans-nasal
Raman Sharma *et al*.[16]	8 days	Respiratory distress	Trans-palatal
Rathore *et al*.[17] (series of 4 cases)	4 months to 14 years	Respratory difficulty	Trans-nasal
		Intraoral mass	Trans-palatal
Present case	18 months	Cleft lip and palate with intraoral mass	Trans-cranial

NA – Not available

the sac. Trans-sphenoidal approach also has the potential risk of CSF leak. Treatment is basically directed toward reduction or excision of the sac with preservation of vital neurovascular tissue contained within it.[21] Repair of the skull base defect and prevention of CSF leak are goals to be attained during the surgical repair. In our case, transcranial repair was performed in which the sac was decompressed and the cavity was packed with fat and fibrin glue. However, as reported earlier, trans-palatal bipolar shrinkage of the herniating sac could have been an alternative, considering that the patient had a complete cleft palate. The repair of the latter could have been planned in the same sitting.

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Conflicts of interest

There are no conflicts of interest.

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