Transsellar Transsphenoidal Rhino-Oral Encephalocele

Transsellar Transsfenoidal Rino-Oral Ensefalosel

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ABSTRACT

Transsphenoidal encephaloceles are rare and the transsellar variety is the least common. We present a 1-year-old male patient with transsellar transsphenoidal encephalocele which herniated into the oral cavity through the congenital split palate. The patient was operated on using a combined transcranial and transpalatal approach without mortality or permanent morbidity. Clinical findings, imaging reviews, surgical repair techniques and postoperative morbidity are discussed with the relevant literature. We conclude that repair of a transsphenoidal encephalocele should be coordinated between a team of neurosurgeons and otorhinolaryngologists. Our surgical outcome supports a transcranial approach for the treatment of these difficult lesions, with transpalatal dissection and exposure.

KEY WORDS: Encephalocele, Transcranial approach, Transpalatal approach, Transsellar meningoencephalocele

ÖZ

Transsfenodial ensefalosel nadir görülür ve transsellar tipi en nadir görülenidir. Bu raporda doğumsal yarık damaktan ağız boşluğuna fıtıklaşmış transsellar transsfenoidal ensefaloselli bir yaşında erkek olgu sunuyoruz. Hasta mortalite ve kalıcı morbidite olmaksızın kombine transkranyal ve transpalatal yaklaşımla opere edildi. Klinik bulgular, görüntüleme çalışmaları, cerrahi tamir teknikleri ve ameliyat sonrası görülen morbidite ile ilgili literatür tartışıldı. Transsfenoidal ensefalosel tamiri nöroşirürji ve kulak burun boğaz uzmanlarından oluşan bir takım tarafından yapılmalıdır. Cerrahi sonucumuz tedavisi zor olan bu lezyonların transkranyal yaklaşım ile transpalatal disseksiyon ve ortaya koyma ile desteklenmesi gerektiğini ortaya koymaktadır.

ANAHTAR SÖZCÜKLER: Ensefalosel, Transkranyal yaklaşım, Transpalatal yaklaşım, Transsellar meningoensefalosel

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INTRODUCTION

Encephaloceles occur in approximately 1 in 3000 to 5000 live births (4,9). Basal meningoencephaloceles are rare anomalies, reportedly constituting 1% - 10% of all encephaloceles and originate from a congenital opening in the midline region of the skull base, which permits meninges, neural tissue or both to herniate from the intracranial space (8.12.14.15). Basal encephaloceles, occur with an estimated incidence of one in every 35,000 live births and have been further subdivided, depending on the location of the bone defect, into transethmoidal, sphenomaxillary, sphenoorbital and transsphenoidal (18). Transsphenoidal encephaloceles are rare and the transsellar variety is the least common.

A CASE REPORT

A 1-year-old male with a diagnosed transsphenoidal encephalocele was referred to us in 2005. There was a history of a slowly progressive mass in the mouth since birth and his attending pediatrician advised neurosurgical repair after cranial MRI. He had been born at the term with a body weight of 2750 g. He was the first child of his parents and there was no consanguinity. His mother had no history of drug or another chemical agent using during pregnancy. On examination the child was alert and playful. His body weight was 8500 g, body height was 67 cm and the head circumference was 44 cm. He had no neurological deficit. His mental milestones were normal. He had a mass, which was partially compressible, in the mouth 4 cm in diameter and herniating from the nose through the split soft and hard palate. A transsellar transsphenoidal rhino-oral wide encephalocele was diagnosed on the MRI (Figure 1). The CT scan with bone window settings, including a paranasal sinus view, revealed a large defect in the planum sphenoidale and sella turcica (Figure 2). There was no other cerebral malformation. The results of laboratory examinations including hematological, endocrinological and chromosomal studies were normal.

Prophylactic antibiotics were given and the patient was operated via a transcranial and transpalatal approach. A bicoronal skin incision was used to perform a bifrontal craniotomy and a large pericranial flap was obtained. On microscopic and endoscopic observation, the encephalocele was tightly adherent to the rim of the bony defect within

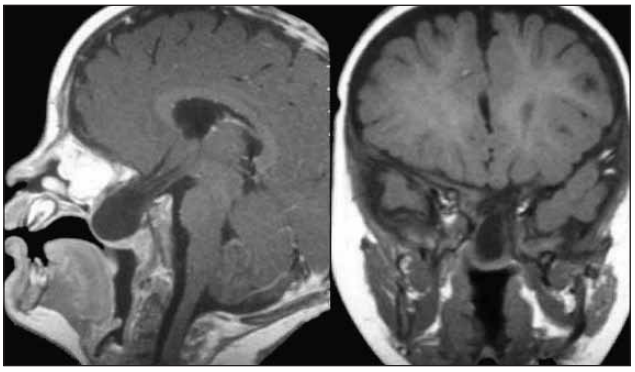

Figure 1: Preoperative sagittal (left) and coronal (right) T1-weighted MR images demonstrating a transsphenoidal cephalocele.

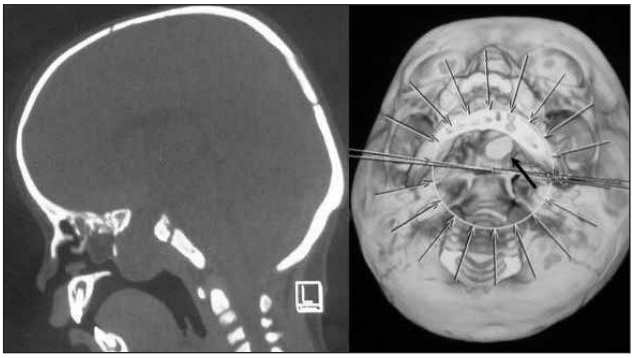

Figure 2: Preoperative sagittal CT scan (left) demonstrating a large defect in body of the sphenoid with a transsphenoidal encephalocele, and a cleft palate. Preoperative 3D reconstruction of CT scan (right) demonstrating a bone defect (black arrow) in the skull base.

the planum sphenoidale and the sella turcica, displacing the optic nerves and the chiasm anteriolaterally. Under microscopic and endoscopic vision, strenuous efforts were made to separate the wall of the encephalocele from the underlying nasal mucosa. First, the anterior wall of the sac was partially incised, permitting CSF outflow from the encephalocele. Separation proceeded from the anterior toward the posterior wall. With partial resection of the anterior wall of the sac, the encephalocele could be separated from the underlying tissue without sacrificing the posterior portion because of the fragility of the nasal mucosa. Then the sac was seperated from the surrounding tissue. The herniated neural elements were pulled up kindly from the nasal cavity. Resection of anomalous neural elements were not performed. Then the herniated wall of the sac was amputated at the lowest level possible, intraoperatively thought to be

near the floor of the sella turcica, and was closed by suturing and with fibrin glue. An ample amount of pericranial flap was placed in the floor of the planum sphenoidale and sella turcica. Then fibrin glue was used again. The second step of operation was performed by otorhinolaryngologists via the trans-palatal approach. The remaining wall of the sac was separated and released from the hard and the soft palate and excised. The dead space under the sella turcica was filled with a free fat graft and the repair was further enforced by a nasopharyngeal mucosal flap. The palatal defect was closed by mobilising the nearby soft tissue and mucosal flaps (Figure 3A,B,C,D,E).

There was no development of CSF leak, endocrine problems, or other complications. The patient received postoperative parenteral antibiotics for a period of seven days and he was discharged from the hospital. Follow-up MR imaging was performed after 12 months postoperatively. Postoperative MRI showed reduction of the encephalocele (Figure 4). For the past two years after discharge, he has remained well without meningitis or other complications.

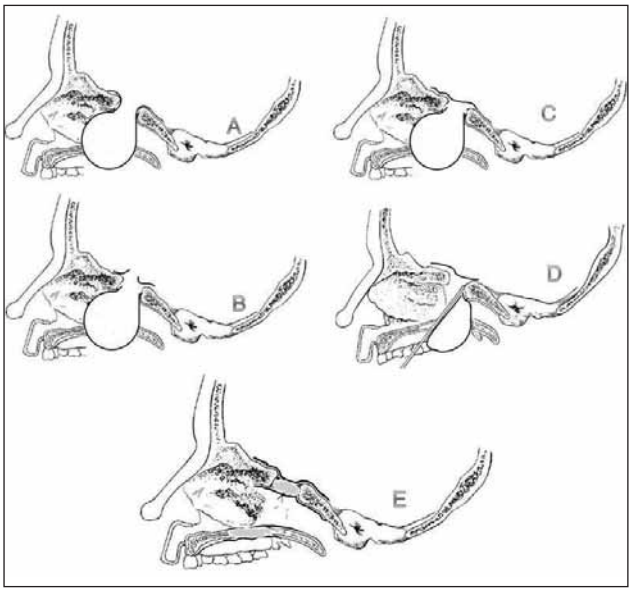

Figure 3: Schematic drawings. **A.** Schematic drawing of transsphenoidal cephalocele, **B.** Under microscopic and endoscopic vision, the wall of the sac was incised, and the herniated neural elements were pulled up gently from the nasal cavity. **C.** The dural sac was closed by suturing and with fibrin glue. **D.** The remaining wall of the sac was separated and released from the hard and soft palate and excised. **E.** The dead space under the sella turcica was filled with free fat graft and then the repair was further enforced by nasopharyngeal mucosal flap. The palatal defect was closed by mobilising the nearby soft tissue and mucosal flaps.

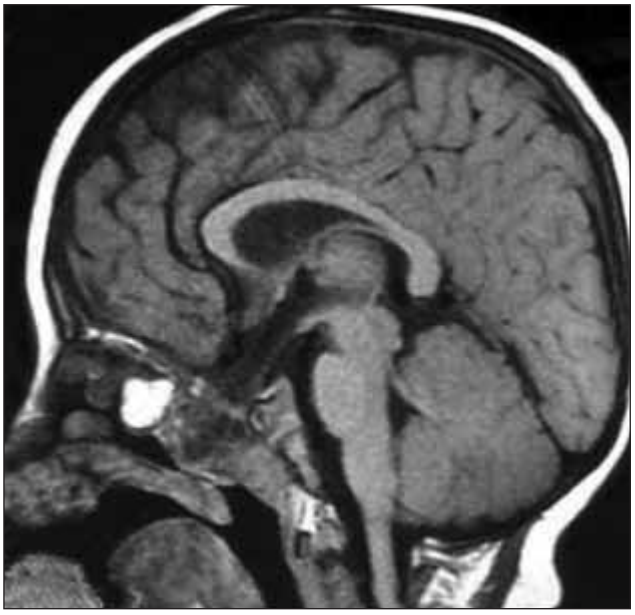

Figure 4: Postoperative MR images demonstrating reduction of encephalocele at 1 year.

DISCUSSION

Encephaloceles are classified as anterior (frontal, sincipital and basal) and posterior (infra- and supratorcular) (7). Posterior encephaloceles are most common (75%) and basal ones least common (1,5%). Basal encephalocele are classified as transethmoidal, sphenoorbital, sphenomaxillary and transsphenoidal (7). The transsphenoidal variant represents approximately 5% basal lesions (6,9) and has an estimated incidence of 1 in 700,000 live births (9). It may be divided into intrasphenoidal, extending into the sphenoid sinus, and true transsphenoidal, transversing the floor of the sinus and protruding into the nasal cavity or nasopharynx (9).

The majority of transsphenoidal meningoencephaloceles are diagnosed during the first year of life due to manifestations such as respiratory distress caused by epipharyngeal obstruction, feeding difficulties, cranial midline defects with cleft lip or cleft palate, hypertelorism, optic malformations with anophthalmia, retinal abnormalities, optic nerve hypoplasia, unexplained bouts of recurrent meningitis or endocrine abnormalities (3.5.10.11.16). Associated congenital anomalies have been noted in one third of the cases of sphenoidal encephalocele

(14.16.17). However, if there are no considerable difficulties and no distinctive facial anomalies during childhood, the diagnosis of the disease may be delayed up to adulthood, when distinctive symptoms such as rhinorrhea, visual defect or endocrine dysfunction occur. Our case had only split palate. There was a history of a slowly progressive mass in the mouth since birth. Endocrine evaluation showed normal levels of hormones and visual functions were also normal.

Advanced imaging studies are necessary to confirm the diagnosis of transsphenoidal encephalocele and to define any neural or vascular elements that may be included in the herniation. CT scan and MRI are the most useful modalities for diagnosing meningoencephalocele (19). In the present case, CT scan including 3D reconstruction allowed visualization of bone defects in the skull base and a well-circumscribed expansile mass lesion in the extracranial area communicating with the intracranial space. MRI with gadolinium enhancement evaluated the content of the encephalocele and eliminated other brain anomalies. MR angiography may be needed to evaluate intracranial vasculature before surgical repair is performed. CT scan including 3D reconstruction and MRI have helped us in planning our surgical approach in the present case.

Management of the transsphenoidal encephalocele certainly requires a multidisciplinary approach. The contents of the sac need to be preserved as the sac invariably contains vital structures. Transsphenoidal encephalocele has been treated by either the transcranial (1,2,13) or the transpalatal approach (13,16,20). The optimal mode of treatment has not yet been established. In reviews by Yokota et al. (20) in 1986 and David (4) in 1993, mortality rates, mainly through transcranial repair, approached 50% and morbidity or long-term severe disability 70%. Transsphenoidal encephalocele is frequently accompanied by a split palate, and these conditions can be operated upon at the same sitting via the transpalatal approach. There is also less risk of damaging the functioning tissues within the wall of the encephalocele. However, the transpalatal approach is feasible only in those cases that have a sufficiently large cleft palate. Otherwise, palatal osteotomies must be performed and the hard palate must be removed. The closure of the mucosal layer and reconstruction of the skull base is then difficult, often requiring the use of skin and bone graft or silicone plate.

CONCLUSION

We conclude that repair of the transsphenoidal encephalocele should be coordinated between a team of neurosurgeons and otorhinolaryngologists. Our surgical outcome supports a transcranial approach for the treatment of these difficult lesions, with transpalatal dissection and exposure. Preoperative evaluation of transsphenoidal encephalocele by fine-cut CT scan including 3D reconstruction and MRI is essential to confirm the extension of the lesion and any associated abnormalities.

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