

Coexistence of occipital and infratorcular meningocele with cervical split cord anomaly

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ABSTRACT In this case report, we present an 18-year-old girl with occipital and infratorcular meningocele and co-existing split cervical cord. She had been admitted into the hospital for cosmetic considerations due to an inborn swelling in the back of the head and neck. Her mental status and neurologic examination were surprisingly intact. We discuss a very rare case of meningocele associated with split cord anomaly in the light of the current literature.

Keywords: anomaly, meningocele, occipital bone defect, spinal cord
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INTRODUCTION

Bony defects of the cranium with no protruding soft tissue is defined as occult cranial bifida. These bony defects are generally placed in the middle of the frontal or occipital bones, and the size of these defects may vary from a small window to a very large opening that may extend down to the posterior arch of the upper cervical vertebrae. The protrusion of only the leptomeninx and cerebrospinal fluid (CSF) through a bony window is defined as meningocele.⁽¹⁾

CASE REPORT

An 18-year-old girl was admitted to the hospital for cosmetic considerations due to an inborn swelling in the back of her head and neck. She was extremely concerned with the cosmetic deformity caused by this lesion before her wedding ceremony. Physical examination revealed a lesion protruding from the mid-occipital region down to the neck. The lesion was soft in consistency and covered by skin folds. We did not detect any fluctuations and pulsations over the lesion. Neurologic examination was within normal range and there was no mental abnormality based on IQ and mini-mental tests. The patient was one of dizygotic twins. Her birth had been delayed due to breech presentation. However, she had never experienced any difficulties after birth. None of her family members had such lesions, and there was no history of a consanguineous marriage between her parents. There was no history of maternal drug use, trauma, exposure to radiation and threatened abortion during her mother's gestational period.

Computed tomography (CT) was first ordered to evaluate the bone and surrounding soft tissue at the same time. As the CT images showed a herniated sac in heterogeneous density through the occipital bone defect which extended to the midline (Fig. 1), magnetic resonance (MR) imaging was performed to evaluate the soft tissue components in detail. We found that the cervical spinal cord was split proximally in a fork-

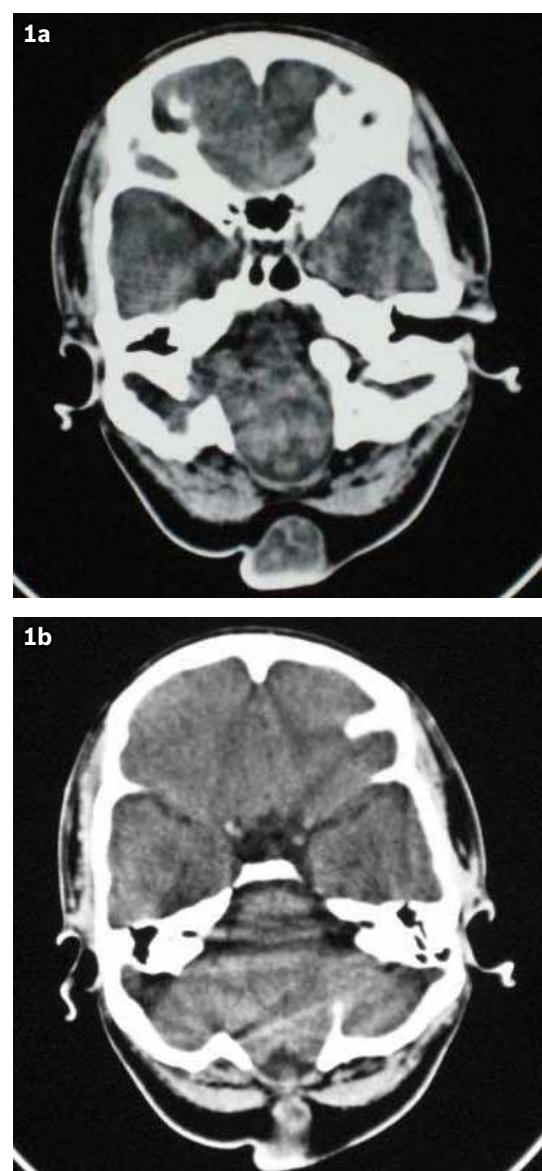


Fig. 1 Sequential axial CT images through the base of the occipital bone show (a) the midline bony defect and (b) the herniated sac containing soft tissue extending subcutaneously.

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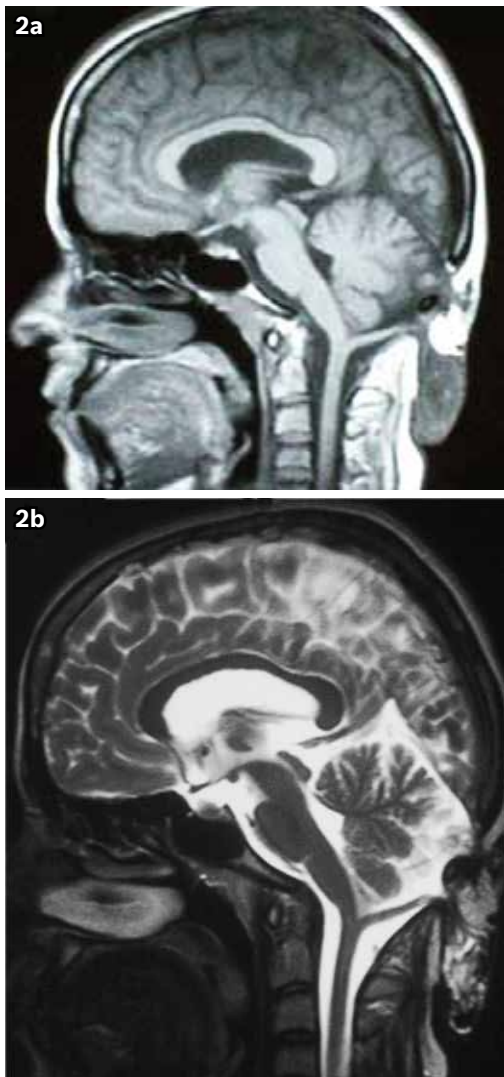


Fig. 2 (a) Non-enhanced sagittal T1-W MR image and (b) sagittal T2-W MR image show the occipital meningocele and superior spinal cord anomaly.

like fashion and extended posteriorly toward the meningocele in the occipital region (Fig. 2). We also detected adipose tissue, leptomeningeal layers and CSF in the protruding sac cavity. Although there was no cerebral or cerebellar tissue in the herniated sac, for practical anatomical considerations, we preferred to use the term meningocele for this herniated sac through the occipital bony defect.

Cerebral MR venography was unremarkable, with normal configuration of the sinuses. No displacement or any pathological appearance and encasement of the great sinuses was seen within the herniated sac cavity (Fig. 3). The patient and her family were informed in detail regarding the benefits and potential risks of the operative intervention, and the patient refused operation at this time.

DISCUSSION

Bony defects of the cranium with no protruding soft tissue is defined as cranium bifida or occult cranial bifida. These bony defects are generally located in the middle of the frontal or occipital bones, and the size of these defects may vary from a

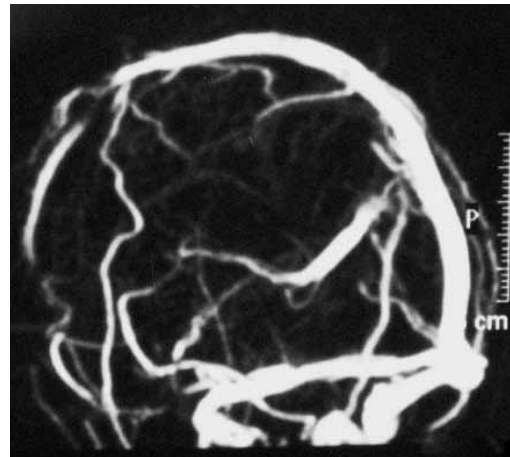


Fig. 3 Reconstructed 3D MR venography of the patient shows no vascular anomaly and occlusion.

small window to a very large opening that may extend down to the posterior arch of the upper cervical vertebrae. Protrusion of only the leptomeningeal and CSF through a bony window is defined as meningocele.⁽¹⁾ Occipital meningoceles are generally detected at birth and are mostly localised under the torcula; rarely, the sac is covered with normal skin layer.⁽²⁾ In our case, the sac was located in the midline where there was a bony defect, and MR imaging signals were in accordance with the existence of leptomeningeal and CSF within the herniated sac cavity.

The dorsal aspect of the split cervical cord extended to the cranial bony defect and was in close proximity with the herniated sac. This appearance is unique, as the coexistence of a split cervical cord with meningocele has not yet been reported; however, split cord malformations may be associated with other anomalies. In this regard, the investigation of 1p35-pter deletion and 14q32-qter duplication inherited from a maternal balanced translocation would have added valuable information to our findings, but this analysis could not be carried out due to lack of informed consent from the patient.

In general terms, encephaloceles are included in neural tube closure defects; however, it is believed that these congenital anomalies occur as a result of a developmental impairment in tissues of mesenchymal origin during weeks 8–12 of embryogenesis.^(1,2) Unlike meningoceles, encephaloceles are defined as a post-neurulation defect during primary neurulation, with a prevalence of 1/5,000–10,000.⁽²⁾ They are 5–10 times less frequent than myelomeningocele, and occipital localisation is more frequent in females and Western societies.^(2,3) Our patient was also a female with a posterior cranial defect. Localisation of the encephalocele with regard to the torcula determines the frequency of neurologic deficits and mental/cognitive development.^(4,5) Although supra-torcular encephaloceles consist of fluid and dysplastic nodules, chances of normal mental development have been reported to be 60%–80%, and the frequency of other associated anomalies, including heterotopias, pachygyria and corpus callosum agenesis, is reported to be approximately 20%.^(6–9) On the contrary, sub-torcular encephaloceles, even with no dysplastic tissue and other

associated anomalies, have only around 30% chance of normal motor-mental development.⁽¹⁰⁻¹²⁾ Surprisingly, our patient's neurologic examination and mental status were completely normal, although she was harbouring a sub-torcular meningocele.

The most sensitive and reliable imaging modality in the diagnosis of meningocele is MR imaging, which is also of paramount importance in the planning of surgical interventions.⁽¹³⁾ The nature of the components in the herniated sac, the configuration of the neck of the sac and its association with the venous sinuses should all be evaluated by MR imaging and MR venography pre-operatively, as the occipital meningocele and its content, as well as rarely encountered lesions or a split cervical cord can be demonstrated clearly on MR imaging. Additionally, MR venography had enabled us to evaluate the proximity of the big venous sinuses with the encephalocele, and has clearly shown that there was no association between the venous sinuses and the neck of the herniated encephalocele sac in our patient.

The basic purpose of surgical treatment of encephaloceles is to excise the herniated sac and its content.^(6,14) Surgical interventions are usually satisfactory when the sac size is small and when they are performed at referral centres where sufficient equipment and experienced staff are available.⁽¹⁴⁾ As the sac size increases and its contents become heterogenic, surgical treatment becomes more challenging, thus increasing the mortality and morbidity rates. In this case, we explained the pathology, surgical treatment options and the potential postoperative risks to our patient, and she had decided not to be operated on at that time.

In conclusion, the prognosis of encephaloceles may vary depending on the size of the bony opening, the herniated sac and its content, the severity of neurologic sequels, other associated genetic anomalies and developmental disorders of brain tissue.^(5,7,15,16) Lastly, as the size of the bony defect and the soft

tissue content in the herniated sac increases, the chance of neurologic dysfunction rises.

REFERENCES

1. Ba MC, Ly Ba A, Hossini A, et al. [The occipital encephaloceles: report of 16 cases]. *Mali Med* 2007; 22:54-7. French.
2. Raja RA, Qureshi AA, Memon AR, Ali H, Dev V. Pattern of encephaloceles: a case series. *J Ayub Med Coll Abbottabad* 2008; 20:125-8.
3. Onrat ST, Seyman H, Konuk M. Incidence of neural tube defects in Afyonkarahisar, Western Turkey. *Genet Mol Res* 2009; 8:154-61.
4. Soumaya SG, Aida M, Sami M, et al. [Encephalocele: 26 retrospective cases at the Maternal and Neonatal Center of La Rabta, Tunis]. *Tunis Med* 2001; 79:51-3. French.
5. Dadmehr M, Nejat F, El Khashab M, et al. Risk factors associated with occipital encephalocele: a case-control study. *Clinical article. J Neurosurg Pediatr* 2009; 3:534-7.
6. Kotil K, Kilinc B, Bilge T. Diagnosis and management of large occipito-cervical cephaloceles: a 10-year experience. *Pediatr Neurosurg* 2008; 44:193-8.
7. Agrawal D, Mahapatra AK. Giant occipital encephalocele with microcephaly and micrognathia. *Pediatr Neurosurg* 2004; 40:205-6.
8. Oruçkaptan HH, Senmevsim O, Akalan N. Occipital encephalocele associated with mediastinal enteric duplication cyst. *Pediatr Neurosurg* 1997; 27:214-7.
9. Suri M, Brueton LA, Venkatraman N, Cox PM. MURCS association with encephalocele: report of a second case. *Clin Dysmorphol* 2000; 9:31-3.
10. Agrawal A, Lakhkar BB, Lakhkar B, Grover A. Giant occipital encephalocele associated with microcephaly and micrognathia. *Pediatr Neurosurg* 2008; 44:515-6.
11. Fatnassi R, Memi A, Bibi M, Khairi H. [Prenatal diagnosis of encephaloceles]. *Tunis Med* 2008; 86:858. French.
12. Lin HJ, Cornford ME, Hu B, et al. Occipital encephalocele and MURCS association: case report and review of central nervous system anomalies in MURCS patients. *Am J Med Genet* 1996; 61:59-62.
13. Carvalho CH, Acioly MA, Will B, Tatagiba M. Magnetic resonance angiography of a complex occipital meningoencephalocele involving the confluence of sinuses. *Case illustration. J Neurosurg Pediatr* 2008; 1:261.
14. Vergara G, Vergara P. Occipital mega encephalocele. *Case report. Minerva Pediatr* 2007; 59:53-6.
15. Ozturk E, Sonmez G, Mutlu H, et al. Split-cord malformation and accompanying anomalies. *J Neuroradiol* 2008; 35:150-6.
16. Muroi A, Fleming KL, McComb JG. Split medulla in association with multiple closed neural tube defects. *Childs Nerv Syst* 2010; 26:967-71.