Fronto-Nasal Encephalocele: Case Report

Frontonazal Ensefalosel

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ABSTRACT Encephaloceles are congenital malformations and consist of glial components. Nasal encephaloceles are often observed on the sincipital region. Nasofrontal encephalocele was diagnosed in a 1.5 month old female patient who was referred to our clinic due to a swelling that was observed on the nasal dorsum since his birth. Intracranial connection was cut via a bifrontal approach. Encephalocele sac have been excised by extracranial approach. In these cases, the bone defect is usually between the frontal and ethmoid bones and usually on the glabella. Multidiciplined approach is required for treatment. Transcranial approach is required to administer dura mending of situations where bone defect was observed on the cranial magnetic resonance imaging and cranial computer tomography. In the same session as the above mentioned, extracranial mending can also be done.

Key Words: Encephalocele; craniotomy; duraplasty

ÖZET Ensefalosel glial dokular içerebilen konjenital malformasyonlardır. Nazal ensefalosel sıklıkla sinsiput bölgesinden gelişir. Doğumundan bu yana burun sırtında gözlenen şişlik nedeniyle kliniğimize sevk edilen 1,5 aylık bir kız hasta nazofrontal ensefalosel tanısı ile ameliyat edildi. Ensefaloselin kafa içi bağlantısı bifrontal yaklaşım ile kesildi. Ensefalosel kesesi ekstrakranial yaklaşımla eksize edildi. Bu vakalarda, frontal ve etmoid kemikler arasında ve glabella üzerinde genellikle kemik defekti mevcuttur. Tedavi için multidisipliner yaklaşım gereklidir. Transkraniyal yaklaşım, kranial manyetik rezonans görüntüleme ve kranial bilgisayarlı tomografide gözlenen kemik defekti ve dura tamirini yönetmek için gereklidir. Aynı seansta transkraniyal yaklaşımla beraber rekonstriksiyon amaçlı ekstrakraniyal yaklaşım ile cerrahi işlem gerçekleştirilebilir.

Anahtar Kelimeler: Ensefalosel; kraniyotomi; duraplasti

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Herniation of cranial contents through a defect in the skull is called as an encephalocele that is a congenital deformity.[1] Encephaloceles are classified as occipital, parietal, basal and syncipital types. Occipital type is the most common representing. The syncipital group is included the frontonasal, frontoethmoidal, frontoorbital, interfrontal and craniofacial types.[2] The frontonasal type is rare and is presented as a mass over the nose, glabella, or forehead. A late neurulation defect during the fourth gestational week is accused in pathogenesis of an encephalocele. In the early childhood especially the neonatal period, the encephaloceles are often time defined.

We report a neonatal childhood with frontonasal meningoencephalocele which we had operated.

CASE REPORT

She was born at term after an uncomplicated gestation. At delivery, her mother was a 20 year-old, healthy, gravida 1, para 1 woman who denied illnesses and use of drugs or medications during pregnancy. Both parents were normal. At birth, the infant was noted to have a skin-covered mass bulging from the right frontonasal suture extending externally to the tip of her nose and marked hypertelorism. The skin on the mass was intact. The lesion was measured 30x15x18mm. She was weighed 3560 g (50-75p), was 49 cm (50-75 p) long (Figure 1) and had a head circumference (OFC) of 34.5 cm (50 p). A cranial CT scan showed a large defect in the frontal bone, a right frontonasal encephalocele and dilatation of the right lateral cerebral ventricle. A cranial MRI showed intracranial connection of malformation (Figure 2, 3). At age 6 weeks, one week after the *v*entriculoperitoneal shunt surgery, part of the frontal mass was removed with bifrontal craniotomy and the defect was repaired with duraplasty and a cranial bone graft was inserted to reconstruct the fronto-basal bony defect. The patient was evaluated with MRI after surgery (Figure 4, 5). She did well and was discharged from the hospital with normal neurologic function and growth. The histopathologic report of the removed cystic mass was "a cystic structure containing mature brain tissue."

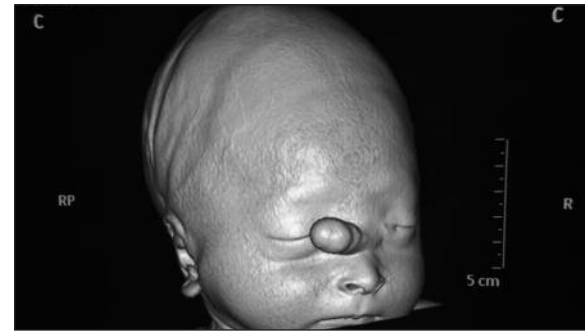

FIGURE 1: Swelling on the nasal dorsum.

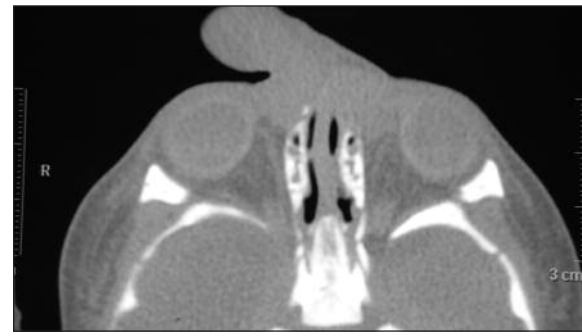

FIGURE 2: Cranial CT showed bone defect.

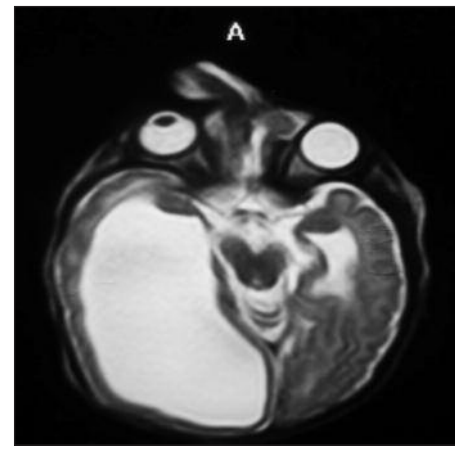

FIGURE 3: A cranial MRI showed Intracranial connection of malformation.

DISCUSSION

Encephaloceles are herniations of the meninges and/or brain which maintain a subarachnoid connection from sutura of the cranium. Frontonasal encephalocele is very rare congenital anomalies. There are a few large series published on the world literature. In one study, 133 case was reported and another study, 187 in 1157 patient with craniospinal congenital malformation were reported to be encephaloceles malformation. Encephaloceles may be included in the fourth groups which comprise occipital, parietal, basal and syncipital groups. The sincipital group is divided into subtypes as follows: the nasofrontal, which exits the cranium between the nasal and frontal bones; the nasoethmoidal, which exits between the nasal bones and nasal cartilages; and the nasoorbital, which exits through a defect in the maxilla frontal process.[3] A frontonasal encephalocele may be mistaken for a nasal teratoma or glioma. Frontonasal

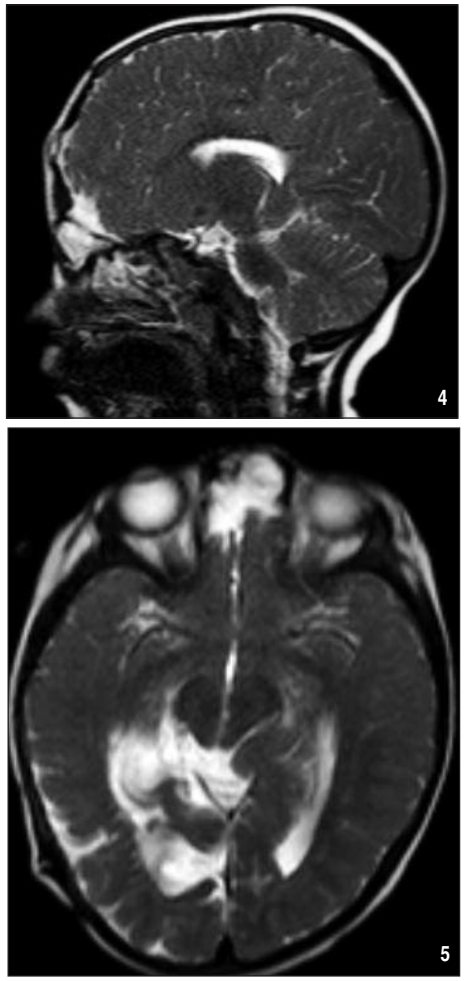

FIGURE 4, 5: Postoperative MRI showed reconstruction of the crania and dura.

encephalocele is showed a solid mass with components. It is anticipated that the prenatal sonographic finding may be useful in diagnose the congenital frontonasal encephalocele.[4] In our patient, frontonasal encephalocele was diagnosed after birth. CT imaging is showed the bony defect while MRI is identified an intracranial connection.

Encephaloceles may associated congenital anomalies, and may presented with facial cleft and hypertelorism. Nasal masses also can lead to cerebrospinal fluid rhinorrhea. Hydrocephalus is rare in anterior encephaloceles. Only 10-15% do have associated hydrocephalus.[3] In this case, encephalocele is associated with hydrocephalus therefore the *v*entriculoperitoneal shunt was placed in the right lateral cerebral ventricle before the surgery.

Biopsy is strongly contraindicated due to risk of infection and meningitis. A bifrontal craniotomy is often used in the surgical procedure of encephalocele. Resection of the pathologic tissue, duraplasty, repair of the osseous defect and reconstruction of the facial deformities are the basic surgical approach.[5] In conclusion, both transcranial and extracranial approach will be useful in the same session in the encephalocele patients with bone defect and its connection to the brain.

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