

# Fronto-ethmoidal meningoencephalocele: A case report

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## ÖZET

Burun kökünde büyük bir kitle (frontoetmoidal meningoensefalosel) ile doğan altı yaşında bir erkek çocuğu olgusu sunulmaktadır. Radyolojik incelemeleri takiben, protrüzyon gösteren kitle ekstrakraniyal olarak bikoronal ve fasiyal insizyonlarla çıkarılmış ve fronto-bazal bölgedeki kemik defekti rekonstrüksiyonu yapılmıştır. Bu yazıda, hastaya uygulanan cerrahi tedavi belirtilmekte ve literatür gözden geçirilmektedir.

Anahtar kelimeler: Ensefalosel, nazoetmoidal meningoensefalosel, frontoetmoidal meningoensefalosel

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## SUMMARY

A six-year-old boy is demonstrated who was born with a large swelling (fronto-ethmoidal meningoencephalocele) at the root of the nose. Following radiographic investigations, he underwent surgery by extracranial approach utilizing bicoronal and facial incisions for removal of the protruding mass and reconstruction of the fronto-basal defect. In this paper, the surgical treatment of the patient is presented and the literature is reviewed.

Key words: Encephalocele, naso-ethmoidal meningoencephalocele, fronto-ethmoidal meningoencephalocele

## INTRODUCTION

Encephaloceles result from congenital openings in the midline region of the skull, often at the junction of the chondro- and desmocranium, which permit meninges, brain substance or both from the cranial cavity. Data on the prevalence of encephaloceles range from 1 in 2500 to 1 in 25000 normal births. A meningocele is a cerebrospinal fluid (CSF)-filled hernial sac that is lined and covered by meninges. A sac that additionally contains brain tissue or other glial matter is referred to as meningoencephalocele. An encephalocystocele is formed by the herniation

of brain and fluid-filled parts of the ventricles through the osseous defect. The severest grade of herniation is the encephalocystomeningocele, in which portions of the brain and ventricles are accompanied by a large collection of CSF in the meningeal space (1,8).

Meningoencephaloceles may be subdivided into occipital, parietal, basal and syncipital (6). The syncipital group has been divided into three types which comprise fronto-ethmoidal (subdivided by facial skeleton exit site into naso-frontal, naso-ethmoidal and naso-orbital), interfrontal and those associated with craniofacial clefts (11). Naso-ethmoidal me-

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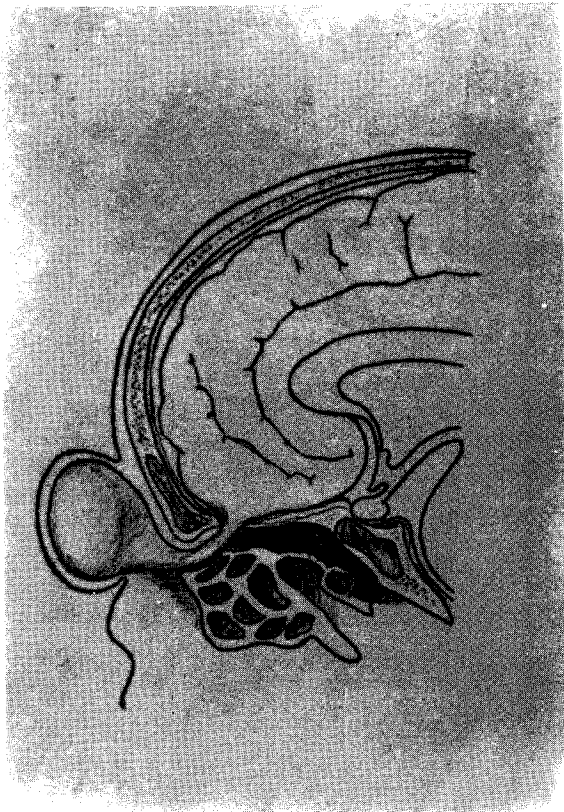


Figure 1. Nasoethmoidal encephalocele. The hernial opening is situated more laterally between the frontal and nasal bones.

ningocele is the herniation of meninges with or without brain tissue through the anterior cranial base in the region of the foramen caecum which displaces the cribriform plate and crista galli posteriorly and protrudes through the skull at between the nasal bones and nasal cartilage (Figure 1). In this paper, we report a case with fronto-ethmoidal meningoencephalocele which we had operated.

**CASE**

B G, a six-year-old male child was referred to our clinic complaining with a large swelling between his eyes (Figure 2,3). The little boy had been born with this round and soft subcutaneous mass. The covering skin was normal in appearance with slight hyperpigmentation over the distal part. The lesion was not compressible, but was increasing slightly in size during crying. Radiographic studies showed the mass to be cystic with the transverse diameter of 45 mm and there was an osseous defect in the fronto-basal region. The sac seemed to project laterally between



Figure 2,3. The smooth and rounded swelling seen over the nose of the patient.

the frontal and nasal bones (Figure 4). A bicoronal scalp incision was done to reach to the mass from above for removal. In addition, the overlying skin of the lesion was incised on the midlateral line and the lesion was progressively isolated from the nose towards the cranial base. The hernial opening was lo-





Figure 4. The radiologic appearance of the lesion and the cranial base defect.



Figure 5,6. The appearance of the patient seen post-operatively.

cated more laterally between the frontal and nasal bones. When the cranial defect had been reached through the bony nasal framework, the hernial sac was removed and the meningeal defect was carefully closed through the bicoronal scalp incision. Duraplasty was not required. A cranial bone graft was inserted to reconstruct the fronto-basal bony defect. Bilateral medial canthopexy was accomplished. A Z-plasty skin closure was performed to prevent subsequent contracture of the skin. The histopathologic report of the removed cystic mass was "a cystic structure containing mature brain tissue. The features are those of an encephalocele". The post-operative period was uneventful. Figures 5 and 6 show the post-operative appearance, There has not been any relapse and the patient has not requested a secondary surgical procedure to improve his aesthetic appearance.

**DISCUSSION**

Smith et al. states that cysts containing brain tissue, even if a link with the brain can not be de-

monstrated, should be regarded as encephaloceles (10). As in the cases reported by various authors, our patient had a naso-ethmoidal meningoencephalocele of the fronto-ethmoidal type because the hernial opening was situated more laterally between the frontal and nasal bones (8,11). Because of their position and size, fronto-ethmoidal meningoceles and meningoencephaloceles cause alterations and distortions of the surrounding facial structures such as displacement of the medial orbital walls, the entire orbits, telecanthus and hypertelorism (2-5,9,12,13). Patients with this malformation demonstrate swellings

of varying size in the glabella-nasal region. These swellings may be sessile or pedunculated. On palpation the mass may be solid and firm or soft and cystic. The contents of the sac mostly consists of glial tissue, often infiltrated with fibrous trabeculae. The skin over the mass may be normal in appearance, thin and shiny or thick and wrinkled. Hyperpigmentation and hypertrichosis may be noted. Visual acuity may be decreased. Strabismus and lacrimal obstructions, resulting in epiphora and/or dacryocystitis can be observed (4,7,13).

Differential diagnosis should be made from traumatic encephalocele, ethmoid-frontal sinus mucocele, neurinoma, hemangioma and glioma. Diagnosis or clinical recognition might not be easy if the cerebral hernia is confined mainly within the nose. A common feature of encephaloceles that enter the nasal and nasopharyngeal space is impairment of nasal airway. These lesions are easily mistaken clinically for nasal polyps or tonsillar hyperplasia. Not infrequently, the diagnosis is made only after meningitis develops following an adenotomy or nasal polyp removal. In doubtful cases endoscopy of the nasopharynx will reveal the sacs as smooth and pulsating masses (2-8). Conventional radiography and three dimensional computerized tomographic (3D-CT) investigations demonstrates the cranial and facial exit holes of the encephaloceles and aid in treatment planning (2).

A surgeon treating a nasal fistula or cyst must be prepared for the necessity of opening the subarachnoid space and performing a duraplasty, as the lesion may communicate with the intracranial cavity. The same possibility exists in the patient who presents with a rounded, firm or tense swelling on the nasal dorsum, which may be a meningocele. It is recommended that large encephaloceles, especially those projecting into the nasopharynx, be exposed through a transfrontal intradural approach, reduced into the cranium and secured with a pericranial flap (8). Lello et al. describes one-stage correction of fronto-ethmoidal meningoencephaloceles and related stigmata, via an orbito-cranial approach. A bifrontal craniotomy is only required when simultaneous correction of hypertelorism is to be undertaken. The authors recommend a combined intra- and extracranial approach. The possibility of a

high relapse rate for repaired fronto-ethmoidal meningoencephaloceles, together with the possibility of prolonged post-surgical cerebrospinal fluid leakage, meningitis and other complications is invited when either a transcranial bifrontal craniotomy surgical approach or an extracranial approach via the facial lesion, is undertaken alone. They mention a modification of existing craniofacial surgical approaches in order to avoid a frontal craniotomy, to allow for good repair of the encephalocele, together with significant benefits in terms of simplification of the surgical procedure, operating time, blood loss, frontal lobe retraction and complications (4). In conclusion, comprehensive surgical treatment of patients with fronto-ethmoidal meningoencephaloceles involves resection of the pathologic tissue, meningo-plasty, repair of the osseous defect and reconstruction of the facial deformities (bone and soft tissues).

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