Median Facial Cleft with a Frontoethmoidal Encephalocele Treated with Craniofacial Bipartition and Free Radial Forearm Flap: A Case Report

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ABSTRACT

We describe a patient with a median facial cleft with a frontoethmoidal encephalocele, hypertelorism, hydrocephalus, and cerebrospinal fluid (CSF) leakage referred to our department due to numerous complications after previous surgical treatments. An 8-year-old girl, born with median cleft syndrome, underwent neurosurgical repair of the encephalocele at another hospital and cleft lip/palate repair later in the same year. Her hydrocephalus was treated with a ventriculoperitoneal shunt, but she underwent numerous shunt revisions due to recurrent intracerebral infections. In 2008, she was rehospitalized due to a gram-negative meningitis and cerebral abscess. She underwent surgery where part of her frontal bone was removed due to osteomyelitis. She was referred to our department due to persistent CSF leakage, recurrent infections, and significant dura defect. In addition, she had hypertelorism and a strongly reduced vision. We performed a monobloc and facial bipartition osteotomy where 15 mm of her frontal and nasal bone was removed after facial bipartition. The dura defect was closed using a free fasciocutaneous flap. The patient had no CSF leakage or infections postoperatively, and her hypertelorism was reduced. The case represents the first monobloc and facial bipartition osteotomy performed in Norway as a part of the treatment of median cleft syndrome with a nasoethmoidal encephalocele.

KEYWORDS: Congenital deformity, encephalocele, median cleft syndrome, craniofacial surgery, facial bipartition

An encephalocele is defined as a protrusion of cranial contents beyond the normal confines of the skull.¹ They are generally thought to arise from neural tube defects, leading to malformations of the skull base. Although 80 to 90% of encephaloceles in the Western hemisphere are occipital, anterior localization occurs

much more commonly in the Eastern hemisphere.² In Southeast Asia and parts of India, Russia, and Central Africa, the highest incidence reported is 1 in 3500. This is 10 times more frequent than countries such as Australia, North America, Japan, Hong Kong, and Western Europe, where the incidence is as low as 1 in

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35,000 live births.³⁻¹⁵ The frontoethmoidal localization is the most frequent variety of anterior meningoencephalocele.^{6,8,12,13,16}

Frontoethmoidal meningoencephalocele is a congenital herniation of intracranial contents, including meninges and brain, through a bony defect in the skull at the junction of the frontal and ethmoid bones. A cephalocele can consist of meninges (meningocele), meninx and nervous tissue (meningoencephalocele), or meninx, nervous tissue, and a part of a ventricle (hydroencephalomeningocele). They are further classified into nasofrontal, nasoethmoidal, and naso-orbital types according to localization.¹⁷

The frontoethmoidal encephalocele manifests itself clinically as a congenital mass near the root of the nose. Frontoethmoidal encephaloceles are covered with skin, although the skin may be affected by pressure necrosis or severe distension of the cele.¹⁸

Frontoethmoidal encephaloceles are often accompanied by an increased distance between the medial orbital walls, but not the distance between the lateral orbital walls. This is called interorbital hypertelorism.¹⁹ The cele may cause nasal visual field defects and nasal airway obstruction leading to obligatory oral respiration. Ocular movements may be disturbed.

Median facial clefts (with or without encephalocele) are associated with true hypertelorism, leading to numerous symptoms. This can be best treated with facial bipartition and translocation of the orbits medially.

MANAGEMENT

Management involves meticulous preoperative assessment using computed tomography (CT) scans and

magnetic resonance imaging, followed by surgical repair of the central nervous malformation, skeletal deformities of the orbit, and nasal deformities. The aims of surgical treatment are to prevent complications of an encephalocele and to achieve cosmetic improvement.

The goals of surgical treatment are reduction of the encephalocele, watertight closure of the dura mater, closure of the skull defect, and reconstruction of the midface together with the medial orbital walls. The operation uses a craniofacial approach. A two-stage reconstruction can be performed, with neurosurgical repair during the first procedure and craniofacial reconstruction during the second procedure. However, a single-stage approach is preferable, unless there are extenuating circumstances.

CASE REPORT

Clinical Presentation

An 8-year-old girl was born with median cleft syndrome including nasoethmoidal encephalocele, cleft lip, and cleft palate. She had hypopituitarism with diabetes insipidus, treated with hormone replacement. In addition, she had secondary hydrocephalus, hypertelorism, and a strongly reduced vision due to repeated shunt failures and meningitis.

Shortly after birth, she underwent her first surgical treatment for her encephalocele in another hospital, where artificial duragraft (TachoSil Nycomed Pharma, Drammensveien, Asker, Norway) was used to close the dura mater defect. Her cleft lip and palate were operated in a second hospital the same year. Her secondary hydrocephalus was treated with a bilateral ventriculoperitoneal

Figure 1 Preoperative magnetic resonance imaging showing herniation of intracranial contents through a congenital malformation of the bony structures of the midface and excessive dural defect. Axial (A) and sagittal (B) views.

(VP) shunt. The VP shunt had to be revised first in 2002, and she underwent further revisions in 2008 due to recurrent intracerebral infections. She was hospitalized in 2007 due to a shunt malfunction, and a herniation of the brain through a defect in the frontal bone was discovered (Fig. 1A,B). For this, she underwent another attempt at frontobasal reconstruction, using a pericranial flap. The patient had a generalized seizure 2 weeks later due to an intracerebral abscess. She was operated twice in March 2008: first to remove her abscess and an infected frontal bone flap (Fig. 2), and then to seal a cerebrospinal fluid (CSF) leak. She was admitted to the hospital again in May 2008 due to general fatigue, nausea, leakage, and stiff neck, and a diagnosis of gram-negative meningitis was made. Due to persistent CSF leakage, the patient was referred to the Neurosurgical Department of Rikshospitalet Oslo for salvage surgery.

Intervention and Technique

The aims of the surgical procedure were to repair the dura defect and stop the CSF leakage to the esophagus, repair the frontal bone defect, and correct the hypertelorism.

The operation was performed under general anesthesia with the patient's head placed in a Mayfield head holder in the neutral neck position. A bicoronal skin

incision was completed postauricularly. Interfacial dissection over the temporal muscles together with the exposure of the orbits, zygomatic arches, and the nasal region was performed. A bifrontal craniotomy was completed.

Zygomatic arch osteotomies were performed. The lateral orbital wall osteotomy was initiated through the inferior orbital fissure and then extended superiorly. The orbital roof osteotomy was continued laterally through the sphenoid wing. With the use of a thin chisel, the medial orbital wall osteotomy was completed posteriorly to the medial canthus and nasolacrimal apparatus. The orbital osteotomies were completed through the floor of the orbit.

The pterygomaxillary disjunction was performed from the cranial side, thus avoiding intraoral dissection. To identify the pterygomaxillary suture line, an image-guidance system was used (BrainLAB™ BrainLAB Scandinavia, Risskov, Denmark; Fig. 3). The sutures were marked out on the preoperative CT images, thus facilitating their identification intraoperatively. An image-guidance reference star was attached to the chisel, making the device trackable and allowing us to use the chisel as a navigation tool or pointer. Furthermore, the maxillary artery was also marked out on preoperative images, thus reducing the risk of accidental injury to the vessel.

The disimpaction of the midface could be performed with a pterygomaxillary spreader forceps and a nasomaxillary (Tessier) forceps. A midnasal osteotomy and sagittal splitting of the maxilla with a sagittal saw were performed to complete the facial bipartition. The original median cleft lip repair was preserved. The two midface halves were fused successfully and stabilized with microtitanium plates and screws. The midface was partially advanced and the zygomatic arches fixed with microtitanium plates and screws.

Figure 2 Preoperative computed tomography scan with three-dimensional reconstruction showing partial defect of the frontal bone after earlier osteotomy due to infection. Hypertelorism.

Figure 3 Intraoperative photograph of the free radical forearm fasciocutaneous flap.

Figure 4 Intraoperative neuronavigation image facilitating the identification of the pterygomaxillary suture.

After rigid fixation of the osteotomy lines, the defect in the anterior scala became more triangular. A 2.5×2.5 -cm free fasciocutaneous flap (Chinese flap) with a 15-cm pedicle was harvested from the left forearm and used to close the dural defect (Fig. 4). A graft including skin was chosen as this gives significantly reduced intranasal granulations compared with a fascial flap. Furthermore, the thickness of a fasciocutaneous flap makes it easier to handle, with enhanced strength of the anterior fossa repair. The vascularized graft was placed with dermis facing the nasal cavity. The fascial layer was sutured in a watertight fashion to the dural edge around the bony defect in the midline of the anterior cranial fossa, thereby creating a support for the brain tissue previously herniated into the rhinopharynx.

The radial artery and cephalic vein were carried from the anterior fossa to the infratemporal fossa via a tunnel drilled through the greater sphenoid wing and anastomosed to the superficial temporal artery and vein, respectively.

A new frontal bone was harvested from the left frontoparietal region, but the bone was too thin to allow

for a split graft. After copious irrigation of the operative field, the skin was closed with interrupted sutures, and subcutaneous suction drains were placed. The patient was treated with steroids and antibiotics as a prophylaxis in the postoperative phase.

Results

As a result of the surgical treatment, the skeletal morphology of our patient improved. Her hypertelorism was reduced by 15 to 16 mm and the interpupillary distance reduced from 74 mm to 61 mm (Fig. 5). There was no further CSF leakage postoperatively. The patient's vision did not improve.

DISCUSSION

Our case was particularly difficult, as the patient had undergone a primary neurosurgical encephalocele repair without attempted craniofacial reconstruction and then a primary median cleft lip repair elsewhere. Furthermore, she had had numerous shunt revisions due to recurrent intracerebral infections, leaving her almost blind. In

Figure 5 Postoperative computed tomography scan with three-dimensional reconstruction.

2008, she was rehospitalized with a gram-negative meningitis and cerebral abscess and underwent neurosurgical treatment, where a part of her frontal bone was removed due to osteomyelitis, and an attempted frontobasal reconstruction using a pericranial flap. She was referred to our department due to persistent CSF leakage at the age of 8.

To achieve our treatment goals (i.e., dura repair and correction of hypertelorism), we opted for a free fasciocutaneous flap to close the dura defect in the anterior cranial fossa floor and a facial bipartition osteotomy to reduce the interpupillary distance. Because of numerous previous surgeries, we used neuronavigation (BrainLABTM) to identify the pterygomaxillary suture lines prior to midface disjunction.

The surgical management of complex craniofacial malformations has evolved,²⁰ with monobloc²¹ and facial bipartition osteotomy¹⁹ representing perhaps the most sophisticated reconstructive approaches to correct the upper and midface deformities observed in median facial cleft syndromes.²² As this kind of surgical treatment involves many medical specialties, a centralization of patients and treatment should be considered to improve the effectiveness.

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