Meningoencephalocele: a late complication of Le Fort III midfacial advancement in a patient with Crouzon syndrome

Case report

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Complications of Le Fort III midfacial advancement include CSF rhinorrhea, meningitis, and ocular and cerebral injury. This report reviews the anatomy of the Le Fort III osteotomies and their relevance to the unusual complication of meningoencephalocele.

In this report, a young male patient with Crouzon syndrome underwent subcranial midfacial advancement at the age of 10 years for obstructive sleep apnea and ocular exposure. He presented 4 years later complaining of nasal obstruction. On physical examination, a mucous-covered mass was noted in the left upper nasal vault medial to the turbinates. Computed tomography scanning and MR imaging confirmed the diagnosis of frontoethmoidal meningoencephalocele. Repair of the meningoencephalocele was accomplished using a combined neurosurgery and plastic surgery approach.

Meningoencephalocele is a rare complication of subcranial midfacial advancement. The abnormal anatomy of the anterior cranial base in patients with syndromic craniosynostosis places them at greater risk for fracture of the cribiform plate and dural tears during this procedure. Unrecognized dural injury is the etiology of this complication in this young patient; however, elevated intracranial pressure may have been a confounding factor. Attention to the anatomy of the anterior cranial base, as seen on sagittal CT images, will aid in preventing this complication.

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Key Words • meningoencephalocele • encephalocele • Le Fort III • Crouzon syndrome • midfacial advancement

Le Fort III midfacial advancement is often necessary in patients who have the well-known eponymous craniosynostoses. The indications for this procedure are obstructive sleep apnea, exorbitism, and Class III malocclusion as well as psychosocial issues. Rare but grave complications of this procedure include CSF rhinorrhea, meningitis, blindness, brain damage, and hemorrhage. The high-level dysjunction at the Le Fort III nasofrontal osteotomy is associated with fractures of the cribiform plates and tearing of the tightly adherent dura at the cranial base. This result may be CSF rhinorrhea, meningitis, meningoencephalocele, or encephalocele.17 Meningoencephalocele is defined as a herniation of meninges and brain from the cranial cavity. Most commonly, these anomalies are congenital, and rarely do they present as a complication of a cranial procedure.2

Case Report

History and Examination. This 14-year-old Caucassian boy with Crouzon syndrome and a C-to-G (cysteine-to-tryptophan) transversion mutation of FGFR2 has been monitored since infancy at our craniofacial center. As an infant, he had sagittal and coronal synostosis, and a sagittal strip craniectomy was performed. Hydrocephalus was successfully managed by a ventriculoperitoneal shunt placed at 1 year of age. Chiari malformation with tonsillar herniation to the C-2 level and significant cervicomedullary compression required decompression of the posterior fossa at 4 years of age. As a child, his cranial index was balanced because of the combined sagittal and coronal synostosis, a condition known as balanced dysmorphism.6 The patient demonstrated minor exorbitism with Class III malocclusion, and it was expected that midfacial advancement would be necessary in the future. At 10 years of age, he began to exhibit symptoms of obstructive sleep apnea, ocular exposure, and corneal irritation. The decision was made to proceed with Le Fort III advancement with a halo device for distraction. A preoperative CT scan was obtained showing abnormal anatomy (Fig. 1).
Meningoencephalocele as a complication of midfacial advancement

First Operation. The pterygomaxillofacial osteotomies were performed via bilateral gingivobuccal incisions and a coronal incision. The zygomatic arches were cut using a sagittal saw. The sphenozygomatic osteotomies were made from the infraorbital fissure to the level of the zygomaticofrontal junctions. The medial orbital osteotomies were made behind the posterior lacrimal crest and anterior to the foramen of the anterior ethmoidal artery. The medial and lateral orbital cuts in the floor were connected under direct visualization. The nasofrontal osteotomy was planned based on the sagittal CT images. A sagittal saw was used to begin the osteotomy at the nasofrontal suture, that continued inferiorly to the superomedial orbital osteotomies. A curved 10-mm osteotome was used to extend the midline osteotomy through the perpendicular plate and vomer. Rowe forceps were used to mobilize the midfacial complex. Lateral canthopexies were completed, and the scalp was closed over a drain. No dural tears or CSF was seen. The rigid external distraction device was secured to the midface with an initial advancement of approximately 10 mm.

First Postoperative Course. The patient’s postoperative course was uncomplicated; no CSF rhinorrhea was noted. After 25 mm of advancement, all symptoms of obstructive sleep apnea resolved, and after a 3-month period of consolidation, the device was removed without difficulty.

For 3.5 years, the patient did well without complaints. On a routine annual visit, 4 years after the midfacial advancement, the patient reported a 6-month history of progressive sleep disturbance, snoring, nasal obstruction, and nasal drainage. He did not have any symptoms suggestive of shunt malfunction. On physical examination, he was noted to have a gray mass medial to the middle turbinate in the left nostril. Computed tomography and MR imaging revealed a 12-mm dehiscence of the ethmoidal roof and a 19-mm soft-tissue density in the frontoethmoidal recess consistent with meningoencephalocele (Fig. 2). Brain MR imaging demonstrated gliotic frontal lobe tissue that herniated through the anterior cranial floor within the meningoencephalocele (Fig. 2). Given the high risk of continued herniation and meningitis or encephalitis, operative repair was necessary.

Second Operation. The patient was brought to the operating room for craniofacial reconstruction through a combined approach between the neurosurgery and plastic surgery teams. The goals of the procedure were to clearly define the planes of the meningoencephalocele, resect the encephalocele, and prevent recurrence and minimize the risk of CSF rhinorrhea by reconstructing the floor of the anterior cranial fossa. In addition, the bandeau and frontal bones were to be advanced to improve his sagittal supraorbital cranial-globe relationship. The coronal scar from the previous procedure was reincised, and a bifrontal craniotomy was performed. The orbital bandeau was removed in the standard fashion. The dura surrounding the region of the left-sided ethmoidal herniated meningoencephalocele was gently dissected from the anterior cranial floor (Fig. 3). To dissect the posterior portion of the meningoencephalocele, a durotomy was made for exposure of the gliotic herniated frontal lobe. All tissue descending below the cranial base was amputated. The remaining brain surface was examined, and hemostasis was achieved. The posterior portion of the herniated dura could then be retrieved and elevated from within the 2 × 1.75–cm anterior cranial defect. The dural edges were then closed primarily using 4-0 nylon suture. Split-thickness calvarial bone graft was taken from the frontal cranial base defect. Fibrin glue and DuraGen (Integra LifeSciences Corp.) were used to reinforce the dural repair. Frontoorbital advancement was then performed, the frontal bone segments were replaced, and endocortical particulate bone graft was applied to fill the coronal gap created by the advancement. The scalp was closed over a bulb-suction drain, and a lumbar drain was inserted to aid in prevention of CSF rhinorrhea and to allow proper healing of the dural repair.

Second Postoperative Course. The patient stayed in...
the hospital for observation for CSF leakage, and he was discharged from the hospital without complication after removal of the lumbar drain 1 week postoperatively. No changes to his ventriculoperitoneal shunt were made; he has done well without recurrence for 6 months. Additional CT scanning demonstrated correction of the meningoencephalocele (Fig. 4).

Discussion

Le Fort III advancement osteotomy is indicated to correct midfacial hypoplasia, upper airway obstruction, tracheostomy dependency, and ocular problems including exposure keratitis, corneal ulceration, globe herniation, and blindness.6,12,17 The principles of distraction osteogenesis were first applied to lengthening long bones, and later to craniofacial bones. Using Le Fort III distraction, the effects of soft-tissue traction can be minimized, allowing for an increased distance of midfacial advancement with less relapse.1,4,10

Documented complications of the Le Fort III osteotomy include cerebral injury, CSF leakage, and meningitis. In his original description, Tessier17 reported 2 cases of CSF rhinorrhea after Le Fort III osteotomy and advancement in 3 patients. In a review of all intracranial craniofacial procedures, Whitaker et al.18 reported an incidence of 2.2% of CSF leakage that required either an internal or external shunt and possible dural patch. They noted that there were more frequent cases of CSF leakage that resolved spontaneously within 48 hours, although these were not included in this statistic. Multiple meningoencephaloceles were observed as being encountered during primary intracranial dissection by Whitaker et al.18 Phillips et al.19 found no instances of meningitis or CSF rhinorrhea in 14 patients undergoing midfacial advancement. Using distraction osteogenesis, Cedars et al.3 described 1 case (7.1%) of CSF leakage and meningitis due to violation of the cribriform plate. No case of CSF leakage was reported by Fearon5 in 51 cases or by Chin and Toth6 in 9 patients.

Intracranial or dural injuries resulting in CSF leakage, meningitis, or in our case meningoencephalocele likely occur at the midline frontonasal suture osteotomy. As emphasized by Tessier,17 this osteotomy should begin just above the frontonasal suture and cut inferiorly in a plane parallel to the cranial base, as seen on radiographs. This infrabasal osteotomy meets the medial orbital wall osteotomies that are placed 15 mm behind the posterior lacrimal crest. During this osteotomy, the goal is to stay below the anterior cranial base avoiding communication with the epidural space.

Although there is a risk of damaging the cribiform plate in all Le Fort III fractures, traumatic or surgical, in patients with Apert and Crouzon syndromes, the risk is increased due to the anomalous craniofacial morphology. Craniosynostosis affects the position of the cribiform plate and anterior base in relation to external cranial anatomical markers. Craniosynostosis and elevated intracranial pressure can result in an inferior position of the olfactory groove and/or cribiform plate and thinning of the anterior cranial base. In several reports, erosion of the ethmoid has been reported to be so extensive as to result in a nasal meningoencephalocele prior to any intervention.7,8 This occurred in 1 of the 2 cases of CSF rhinorrhea in the original review by Tessier.17 In addition, coronal synostosis can result in abnormal sloping of the frontal bone and anterior cranial base. The results are the well-documented flattening of frontal processes of the maxilla, increased vertical sloping and reduction of orbital depth, shallowing of the medial orbital wall, and expansion of the “ethmoid labyrinth” by ballooning of the medial orbital walls. The combination results in an increased risk of injury to the cribiform plate during osteotomy of the nasofrontal suture. Tessier advocated a low keel-shaped osteotomy to reduce this danger. In the 2 cases in which he reported a CSF leak complication, this technical modification had not yet been introduced.

Coronal and sagittal CT images demonstrate these cranial basilar differences and may help in the safe planning of this osteotomy. Computed tomography scanning is instrumental in guiding the surgeon’s understanding of the relationship between the cranial base and the nasofrontal suture. When the cranial base is high, Le Fort III osteotomies can be done extracranially and blindly using the landmark of the anterior ethmoidal artery foramen and cutting anterior to this point, as was done in this patient. When CT scanning demonstrates a low-lying cra-
nial base, an intracranial approach is recommended to allow for better visualization of the anterior cranial fossa during the osteotomy.

In our center, the standard practice has been to perform blind osteotomies after careful review of the CT findings. It is possible that in the future, intraoperative image guidance could be applied to assist with the intracranial approach. Image guidance has become a mainstay in the resection of brain tumors and has wide applications in both neurosurgery and otolaryngology.11,13 Neuronavigation has been used with preoperative MR imaging and CT imaging to better delineate lesions and surrounding structures. In patients with anterior cranial base lesions, an image-guided approach with CT would improve the safety of the procedure.

There are alternative approaches to frontal craniotomy for repair of congenital and iatrogenic encephaloceles. Among these is an extracranial approach through a single midline forehead incision. The endoscopic technique is commonly used as a minimally invasive approach to repair encephaloceles, and pedicled nasal flaps can be harvested and placed endoscopically to assist with the repair of larger defects.14,15 We elected to perform a bifrontal craniotomy in this patient because he had multiple previous operations, and we anticipated significant scarring. Craniotomy allowed for visualization of the edges of healthy dura as well as the option to harvest a temporoparietal fascial flap and split calvarial bone grafts.

Conclusions

Rare but serious complications of Le Fort III distraction osteogenesis include CSF rhinorrea, meningitis, and ocular or cerebral injury. The etiology of the abnormal anatomy of the anterior cranial base in patients with craniosynostosis predisposes to these problems. We believe that the likelihood of these complications can be reduced by careful preoperative inspection of the sagittal CT images. We review the anatomy of the Le Fort III nasofrontal osteotomy and its relevance to the late complication of meningoencephalocele. We also describe a safe and effective repair technique of an iatrogenic meningoencephalocele.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: all authors. Acquisition of data: Ridgway, Mulliken, Padwa. Analysis and interpretation of data: Ropper, Ridgway, Guummerova. Drafting the article: Ropper, Ridgway, Mulliken. Critically revising the article: all authors. Reviewed final version of the manuscript and approved it for submission: all authors. Administrative/technical/material support: Ropper. Study supervision: Guummerova.

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