

Congenital Nasal Encephalocele - A Review of Surgical Techniques

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Summary

We report a case of a 6 month old baby boy who had congenital nasal encephalocele, repaired via the traditional staging procedure. The surgical techniques and procedures are described and discussed.

Key Words: Nasal encephalocele, Operative procedure, Operative technique.

Introduction

Congenital nasal encephalocele is a complex anatomical abnormality involving various facial and intracranial structures. Its correction ideally requires the expertise of neurosurgical and maxillofacial teams working together. The preliminary investigations for this condition are two and three dimensional CT scans which can give adequate information on the extent of bony and soft tissue pathology. We describe an operative technique for the correction of nasal encephalocele in a child who had undergone a two-stage repair of the condition successfully and discuss other advocated operative techniques.

Case Report

A 6 month old boy was first seen in the surgical clinic at the age of two months with a history of congenital midline upper nose swelling. It was located at the bridge of the nose between the two orbits. The swelling did not increase in size, even during crying and was not associated with rhinorrhea, visual impairment or abnormality in eye movements. The baby did not have any symptoms or signs of increased intracranial pressure. He had been well, tolerating

feeding and had no disturbance in normal developmental milestones.

On general examination, he had no particular dysmorphic features, appeared active with a notable swelling at the bridge of the nose lying between two orbits measuring about 4 x 4cm, soft in consistency, non tender, fixed to inside deeper tissues and non-pulsatile with an obvious palpable bony defect at the base of the nose. The cranial nerves were intact and no other systemic abnormalities were noted.

Initial CT scans reported a mass located between the orbits, which was as isodense as the brain tissues and appeared connected to the brain tissues with an obvious bony defect at that region. In view of the CT findings, he was planned for first stage operation, reduction in herniated brain tissues at the fronto-ethmoidal region with autograft closure of bony defect at age of 6 months.

Operative Techniques

Patient was laid supine with the head above the heart at 30 degrees. The scalp was shaved thoroughly and a marking made for skin incision. The area was then cleaned and draped. A bicoronal skin incision was

This article was accepted: 3 January 2004

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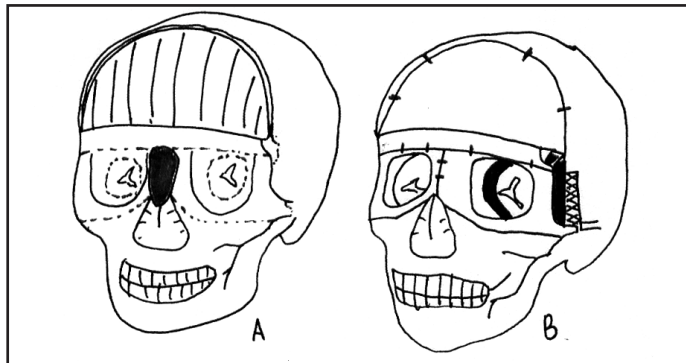
made from the right temporal, anterior to the tragus to the same point on the opposite side after local infiltration of subcutaneous tissues with bupivacaine 2% mixed with adrenaline. Rinnies' clips were used to secure the bleeding at the skin edges. Dissection of the scalp was made to the lower limit of the defect or abnormality. The galea layer was incised with monopolar diathermy and periosteum separated from the bony layer by using periosteal elevator. Bifrontal craniotomy was made with the Smith craniotomes and dura spatulas. The encephalocele was approached via the extradural route without injuring the dural protective layer. The nasal bone defect was identified and the herniated portion of abnormality was opened and noted to contain brain tissues. The surgical reduction of herniated brain was made and then the dura layer was resutured and supported with galea tissues and 1ml of fibrinogen product was injected onto the area. Then the bony defect was reconstructed with the use of autogenous bone graft taken from part of the Burr hole bony fragments. A temporary CSF (Cerebrospinal Fluid) diversion from the operated area was done to ensure good healing and no CSF leak occurred post-operatively. The extraventricular CSF drainage system was inserted and connected to the set. The layers were then closed with Dexon 3/0 for subcutaneous layer and Dafilon 3/0 for skin. He recovered well from the first neurosurgical reduction of herniated brain tissues and reconstruction of bony defect. He was discharged on the eleventh post operative day.

Following the success of the first stage surgical correction, he was then referred to the maxillofacial team. The second stage surgical intervention was the reduction of orbital hypertelorism. This was again successfully accomplished by the Neurosurgical and Maxillofacial teams working together. At the age of 14 months, his second stage operation was completed.

Again under general anaesthesia, the patient was laid supine with head supported at 30 degrees with extended neck. The scalp was shaved and the area was cleaned and draped. Local anaesthesia was infiltrated subcutaneously. Then the frontal region was exposed with bilateral coronal skin flap and bilateral craniotomy was made by the neurosurgeon, followed by exposure of bilateral zygomatic arches and freeing of orbital content from bone by the maxillofacial surgeon. Then a blepharoplasty incision was made bilaterally exposing the infraorbital rim and enabling

identification of the infraorbital nerves and anterior edge of the inferior orbital fissure. This was immediately followed by creation of the orbital frame with bilateral orbitotomy with square margin incision of about 5mm above the supraorbital rim, 5mm below the infraorbital rim (above infraorbital canal), medially at the lateral nasal wall, laterally at the temporal region and posteriorly at the anterior edge of the inferior orbital fissure. Elliptical incision of the fibrous scar at the nasal region and removal of redundant skin and underlying fibrous lesion were made prior to reduction of orbital hypertelorism. The reduction of hypertelorism via mobilization of both orbital frames medially was done for a reduction length of 1.5cm. The upper frontal bar was left after the bifrontal craniotomy and used for plating and screwing the bilateral orbital frames. This was approximated closely as mentioned above by excision of fibrous tissues and some bones at the nasal region. Canthopexy to fix the medial canthal ligament bilaterally to the nasal bone was then made. The fractured bone chips from the nasal region were placed bilaterally at the temporal region with additional support of the new located orbital frame by a split right 6th rib autogenous bone graft. This additional strut allows for stabilization of the advanced segments. A piece of rib measuring 8 cm in length was taken after a small oblique skin incision over the area. It was freed from the periosteum by using a periosteal elevator. Wound closure was made in 2 layers, muscle and skin for blepharoplasty incision as well as incision at the nasal intercanthal region. The neurosurgical team then proceeded with plating of the frontal craniotomy piece and closure of the bicoronal flap after resuturing of temporalis muscles bilaterally. Then the right Rickham's catheter was inserted into the right ventricle via the old Burr hole for previous extraventricular drainage system and the layers closed after dome connection was made.

He was well after operation. The antiepileptic medication and broad spectrum antibiotics were prescribed prophylactically, but on the seventh post-operative day, a colourless fluid was noted to drip out via his nasal intercanthal incision and this tested positive for CSF. Tapping of CSF was made and later the definitive treatment of CSF diversion was made with ventriculo-peritoneal shunt. He made a good recovery thereafter. His third reconstructive surgery will involve correction of the nose at a later age and at a later date if required.



**Fig. 1: A: Central fronto-nasoethmoidal defect shown. Orbital medialization through the intracranial route made via bifrontal craniotomy and the resection of bone between the orbits as shown, leaving behind the supraorbital bar.
B: The orbits are displaced medially and fixed with absorbable miniplates and screws to each other and the supraorbital bar. The autogenous rib bone piece used to support the defect after medialization of the orbits at the bilateral temporal fossa.**

Discussion

Encephalocele is a severe malformation with the involvement of many anatomical areas, treatment of which requires a multidisciplinary approach. In the growing patient, it is necessary to restore as soon as possible the physiological morphology of the craniofacial complex. Many authors^{1,2} have described the various operative techniques in repairing the fronto-nasoethmoidal encephalocele. In the past, operations for encephaloceles were performed in two sessions, but since the development of craniofacial surgery, an intracranial approach combined with extracranial operations in a single session is the choice.

This case was treated with a two-stage operation in order to avoid perioperative complications in a young child like excessive haemorrhage and hypothermia. The two-stage procedure is considered appropriate when:

- 1) Vital brain structures herniate via the defects.
- 2) herniated tissues that lead to respiratory embarrassment, as all neonates are dependent on nasal breathing.
- 3) CSF fistula is present.

- 4) the risk of perioperative morbidity and even mortality is high in a single session.

Except in cases with the above reasons, the current trend in treating this condition is by the single stage operation. Details of the surgical techniques are described above. Figure 1 shows the corrective technique which was used in this case. The single stage operation is similar in terms of surgical techniques as the two-stage operation but has the disadvantage of longer operative time and therefore a possibility of anaesthetic complications such as hypothermia, hypovolaemic shock and acidosis.

Conclusion

Congenital Nasal Encephalocele is a severe disorder that requires a multidisciplinary approach. The neurosurgeon and maxillofacial teams nowadays work together in one session of repair. This is believed to be superior to the historic 2 or 3 stage repair. The staging procedure is indicated in some cases due to the urgency of correcting the cranial defect and to prevent the perioperative complications in a young child.

References

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