Case Report

Posttraumatic giant fronto-orbito encephalocele causing cosmetic disfiguring forehead swelling with proptosis: Management

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ABSTRACT

Fracture of the anterior skull base can occur following head injury. Growing skull fracture is usually observed in children under age of 3-years. It commonly involves frontal and parietal regions. However, orbit involvement is extremely uncommon. Authors report a case of giant orbital encephalocele with a forehead disfiguring swelling in a 4-years boy, who sustained head injury about 3½ years back. However, such delayed presentation of traumatic encephalocele is extremely uncommon and represents the first case in the Western literature, who had a successful postoperative outcome.

Key words: Fronto-orbital growing skull fracture, management forehead swelling, orbital extension

Introduction

Growing skull fracture is unusual complication of head injury. Growing fractures rarely develop in the skull base. It commonly involves frontal or parietal bones. Involvements of orbit are extremely rare. In the most of reported orbital growing fracture, defect in such cases is usually small and often limited to the orbital roof only. These cases commonly present with proptosis and visual symptoms. However, such fracture also extending through orbit and frontal region causing forehead swelling and proptosis is not reported previously. Authors report the first case in the world literature. The pathophysiology of growing skull fracture is still debated. A dural tear underlying a skull fracture is widely believed to be the initiating event. Suri and Mahapatra observed the presence of orbital fracture and associated dural tear was observed in both the cases. Suri and Mahapatra also concluded frontonasal brain injury play an important role in the growing skull fracture pathogenesis.

Case Report

A 4-year-old boy presented with progressive swelling over the right frontal region and right upper eyelid. He has a history of fall from bed at the age of 6 months. Since then the swelling was noticed. There was no history of cerebrospinal fluid (CSF) leak. The swelling was tender, soft, and compressible. It was pulsatile and not covered with bone. The bony defect was palpable around the swelling which was irregular and

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extended down to the roof of the orbit. The lower margin could not be palpated. Neurological examination was normal. Although there was ptosis in the right eye, the vision was intact in his right eye. All this was suggestive of a growing skull fracture of the right fronto-orbital region.

Computed tomography (CT) scan showed a large bony defect in the right frontal region extending to the roof of orbit [Figure 1a and b] with underlying gliotic area without any evidence of hydrocephalus. He underwent surgical repair under general anesthesia. The intraoperative findings revealed a large bony defect of 12 cm × 8 cm with anteroposterior diameter being the longer. The defect was crossing the midline with posterior extension up to the coronal suture. The exposed brain was gliosed and extending through the fracture to the right frontal sinus and adjoining right orbital roof. The bony defect was extending inferiorly up to the optic canal. A large craniotomy was made, and the defect was closed with split thickness bone graft harvested from the skull adjacent craniotomy [Figure 2a and b] after primary watertight dural closure. He had an uneventful postoperative course with subsidence of orbital proptosis and forehead.

**Discussion**

Fracture of the anterior skull base can occur following head injury. The incidence of leptomeningeal cysts ranges from 0.05% to 0.6% of all skull fractures. The majority of these lesions occur in children, and the parietal skull is the most common location. However, delayed presentation of this with a traumatic encephalocele is uncommon. Such patients commonly present with proptosis and visual symptoms. It is commonly found in adult patients following orbital roof fracture with underlying frontal contusion. Pediatric patients with orbital roof fractures that exhibit more than 2 mm diastasis and associated frontal cerebral contusion may be at greater risk for developing intraorbital encephalocele. The defect in such cases is usually small and often limited to the orbital roof. Repair of the orbital roof is the key step in the management of such cases, and the repair should be done in a rigid manner to avoid the transmission of intracranial pressure into the orbit.

In the current case, there was a large defect which involved the right frontal region and also extended downward to the roof of the orbit and frontal sinus and posteriorly involved the optic canal with brain matter herniating through defect. Such a large defect in a traumatic encephalocele which is extremely rare. Moreover, in spite of this large defect, the patient had no visual acuity reduction.

Orbital growing skull fracture can have a spectrum of clinical presentation. The children present with a tender, pulsatile mass, or can be incidental picked up while adult counterpart present with nonpulsatile and nontender swelling having history of the previous head injury. When the orbital roof manifestation include diplopia, proptosis, and restriction of extraocular movements.

Naim-Ur-Rahman et al. classified growing skull fracture depending on the basis of content including into three types. The Type I being those containing herniation of arachnoid tissue only and Type II also additionally containing brain matter herniation and finally type III is associated the presence of porencephalic cyst of underlying brain. These stages correspond to leptomeningeal cyst represented by Stage I and encephalocele as Type II respectively.

For clinical diagnosis in addition to clinical history, examination, the CT provides excellent view bony defects

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**Figure 1:** (a) Computed tomography showing large defect of orbit roof. (b) Computed tomography showing large defect of frontal bone causing forehead swelling

**Figure 2:** (a) Postoperative computed tomography showing large defect of orbital roof reconstructed using autologous bone graft. (b) Postoperative computed tomography showing repair of frontal bone reconstructed using autologous bone graft
in the orbital roof and three dimensional reconstruction. The magnetic resonance imaging shows the intraorbital extension and additional associated intracranial porencephalic cyst, gliosis and helpful in distinguishing the contents as herniated brain parenchyma or CSF or rarely prolapsing venous sinuses.\cite{9,10}

Management of such cases is challenging, and surgical repair is considered as mainstay of treatment; however, cases of spontaneous stabilization are reported in the literature.\cite{5,9,11}

However, aim of the surgical repair to prevent the development of rupture, visual disturbance, cosmetic deformity. Suri and Mahapatra reported two cases of growing orbital roof fracture in children. Both cases were operated with disappearance of proptosis in the postoperative period. Authors recommended child developing ocular symptoms, who sustained a head injury several months or years before should be suspected for possible development of the growing fractures of the orbital roof.\cite{3}

**Conclusion**

In every child presenting with swelling of forehead and orbit with history of head injury even after many years after head injury, possibility of orbital growing skull fracture should always be considered and early surgical management can yield good cosmesis and visual outcome.

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**Conflicts of interest**

There are no conflicts of interest.

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