

Case Report

Proptosis with Growing Skull Fracture of Orbit: A Lesser Known Entity

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Abstract: Growing skull fracture is a well known entity in neurosurgical literature. However growing skull fracture of the orbit is relatively rare and hence remains under diagnosed. Orbital fracture is more common in children. This fracture can even expand several months or years later by evertting its edges. This can result in herniation of brain into orbit which can produce variable symptoms. We present a 3 year old girl with history of fall from height. Ophthalmological examination revealed left inferior displacement of the globe with proptosis and restriction of movement in left upper outer field. Sequential radiological examination revealed a growing skull fracture of orbit. Repair of the orbital roof was done with Titanium miniplates and screws and follow up examination showed complete disappearance of proptosis. The exact pathophysiology of growing fractures is still debated in the literature, but a dural laceration along a fracture line is noted in all cases, and frontobasal brain injury seems to play an important role in the pathogenesis of the fracture growth. Growing skull fracture of the orbital roof should be considered in the differential diagnosis in cases of persistent ocular symptoms. Early diagnosis and management is extremely important in the optimum management and good long-term prognosis of the patient.

Keywords: Growing Skull Fracture, Proptosis, Orbital Roof

1. Introduction

Fractures of the skull are easily diagnosed and are seen by most practicing doctors. Fracture of the orbit mostly remains underdiagnosed. They can present with loss of vision and are referred to ophthalmology for treatment [1]. We present a rare entity known as growing fracture of the orbit in a child. We also analyze the published literature of this rare entity, its presentation and management.

2. Case History

A 3year old girl presented with history of fall from a height of 10 feet, two years ago. She had a brief loss of consciousness without any episode of seizures. There was no redness, pain or swelling in the eyes and no neurological deficit. NCCT brain showed bi- frontal basal contusions and fracture of left orbital

roof and right frontal bone. She was discharged after 48 hrs on symptomatic treatment.

She presented later with progressive proptosis of the left eye with deviation of the eyeball inferiorly and to the left for the last 2 months. On examination she was alert and active. Her vitals and neurological examination were normal except for a lateral and downward displacement of the left eye globe. Ophthalmological examination revealed left inferior displacement of the globe (dystopia) with restriction of movement in left upper outer field. There was left upper eyelid swelling and venous engorgement. NCCT scan revealed bilateral frontal contusions with comminuted fractures of bilateral frontal bones on either side of midline. There was also evidence of fracture of left orbital roof with intraorbital displacement of fractured fragments pushing the left globe inferolaterally (Figure 1).

MRI revealed a left frontobasal cystic lesion hypointense on

T1W images, hyperintense on T2W images extending into the orbit through a defect in the orbital roof. Associated gliotic and cystic changes in the frontobasal region were also present (Figure 2).



Figure 1. Coronal CT scan showing fracture of left orbital roof with displacement of fracture fragment in the globe (arrow).

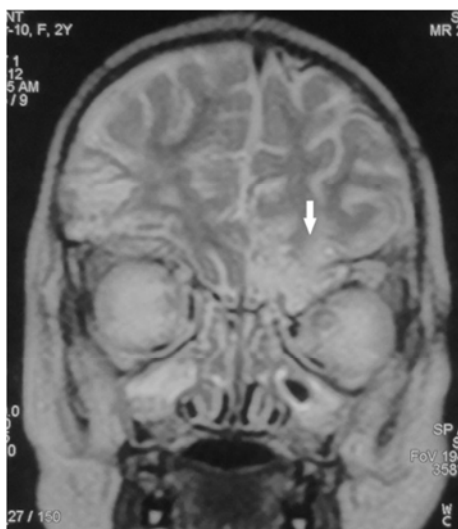


Figure 2. Coronal section of T2 weighted MRI scan showing a cystic lesion in left frontobasal region extending into the left orbit (arrow).

An elective left frontal craniotomy was performed. There was a fracture on the orbital roof and fractured fragment had displaced the periorbital inferiorly laterally. New bone formation was noticed along the fracture margins. The fronto-orbital gyrus showed gliotic changes along with adhesions between the dura and the brain. Multiple dural defects were present with encephalomalacic changes in the brain. There was a cystic lesion in the gliotic brain herniating through the defect into the orbit displacing the globe.

The gliotic brain with cystic part was excised under microscope. The dural edges were released from the bony defect. It was reconstructed by using the pericranium. Repair of the orbital roof was done with 5mm Titanium miniplates and screws. Patient had good cosmetic recovery (Figure 3). Follow up examination at 6 months showed disappearance of

proptosis.



Figure 3. Pre [3a] and immediate postoperative [3b] photographs showing the reduction in the proptosis with widening of the palpebral fissure postoperatively with minimal postoperative edema of left eyelid.

3. Discussion

Growing skull fracture is a well known entity in neurosurgical literature. Orbital fracture is more common in children. The pediatric preponderance of orbital roof fractures reflects the high craniofacial ratio of the young child, the vulnerability of the large cranium to trauma and the lack of pneumatization of the frontal sinus, which is protective of the orbital roof [2].

However, growing skull fracture of the orbit is a relatively rare and hence remains under diagnosed [3]. Orbital roof fractures are generally not taken seriously unless symptomatic. This fracture can even expand several months later by everting its edges. This can result in herniation of brain into orbit which can produce variable symptoms.

Growing skull fracture usually occurs in children less than 3 years of age. The majority of these lesions occur in parietal or frontal bone. The incidence of growing skull fractures ranges from 0.05 to 0.6% of all skull fractures [4]. Though described in the 19th century, the anatomy, pathogenesis, and natural evolution of these fractures still remains obscure. For this reason, these lesions were called by various names including growing skull fracture, cephalohydrocele, leptomeningeal cyst, and posttraumatic porencephaly [5]. Orbital roof fractures were found in 7.1% of patients who suffered head injuries and 13% of those patients developed intraorbital encephalocele [6]. It is also a common practice that many of these fractures are treated conservatively, particularly if there is no loss of vision.

The most accepted theory on the development of these lesions is of Taveras and Ransohoff [7]. Trauma produces a skull fracture and underlying dural tear. At the same time, there is probably sufficient sub-arachnoid hemorrhage to hinder the local circulation of cerebrospinal fluid. The arachnoid membrane projects out through the dural tear into the fracture site. This trapped arachnoid hernia, aided by the normal pulsations of the brain, gradually erodes the edges of the bone, and compresses the underlying cortex. A ball valve mechanism is at work also, with the cerebrospinal fluid having easier ingress than egress from the cyst. Arachnoid adhesions about the margin of the brain also play a part in trapping the fluid locally. A gradient between the pressure in the subarachnoid space and the intraorbital compartment through which the pulsations of the brain gradually erode the bony structures [8]. Such expansion can occur over a variable period of time ranging from few months to several years.

Sequential plain skull radiographs are sufficient to diagnose

such a condition. However, orbital roof growing fractures may require CT scan or MRI, as plain radiographs may not reveal the separated, sclerosed fracture margins of orbital roof clearly [9]. Reconstructed images of CT scan may help further in delineating the extent of bony defect. MRI may differentiate intra-orbital fluid collection (pseudomeningocele) from herniated neural tissue [10]. Delineation of bony architecture is better defined on coronal sections of CT scan, whereas MRI is better for intracranial neural tissue, which herniates into orbit [13].

Treatment of growing fracture of orbit requires that all herniated gliotic tissue should be removed, followed by watertight primary closure of the dural tear and reconstructive cranioplasty [11].

In order to have better results, it is mandatory to perform a water tight dural repair. The bony defect can be covered by various means using natural or synthetic materials [12]. However many authors do not recommend bony reconstruction. Natural bony healing seems to be superior to cranioplasty, which is not always required; especially in a cosmetic region like supraorbital ridge. We repaired the bony defect with titanium plates as the defect was large. It may be rational to avoid bony repair for a small defect however the defects are often enlarged by the surgeon to get proper access to the herniated tissue for its excision [14, 15].

4. Conclusion

Growing skull fractures of the orbital roof are extremely rare. Such lesions should be included in the differential diagnosis when a patient presents with proptosis or orbital pain and a history of head trauma during childhood. It is also essential to do close follow up of orbital fracture in to detect such complication later.

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