Late Surgical Management Of A Rare Case Of Post-traumatic Orbital Encephalocele

Orbital roof fractures after blunt head trauma are not uncommon. Herniation of the brain tissues into the orbit through a bony defect, so-called post-traumatic orbital encephalocele, is a rare entity. The herniated brain tissue causes compression of the intraorbital contents, particularly the optic nerve, extraocular nerves and muscles and compromise the vascular supply. Raised intraorbital pressure may lead to irreversible damage to the optic nerve. This can be prevented by early diagnosis and timely management. Repair of the orbital roof needs to be performed to avoid transmission of intracranial pressure into the orbit. Early intervention is needed in order to prevent visual loss. We present a case of post-traumatic orbital encephalocele who underwent late surgical treatment with direct repair of dura opening, reinforcement with temporalis fascia and reconstruction of orbital roof using skull bone graft. Complete resolution of the pulsatile proptosis with excellent cosmetic result was observed at follow up. However, the visual diminution did not recover significantly due to late diagnosis and intervention. Early diagnosis and surgical management of this rare condition can prevent permanent visual loss and also achieve good cosmetic results.

Key words: Orbital encephalocele, Traumatic encephalocele
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Case Report

A 6-year-old boy presented to the Neurosurgical outpatient clinic, Tribhuvan University Teaching Hospital, Kathmandu, Nepal, with chief complaints of pulsatile proptosis of right eye with visual diminution in both eyes. According to his past history, six months back, he had sustained injury to right side of head and eye following fall while riding a bicycle. During the fall, the bicycle’s handle had caused blunt injury to the right side of the forehead and the right eye. Following the incident, he had a brief loss of consciousness and had multiple episodes of vomiting. However, he had no seizure. Initially, he sustained injury to right side of head and eye following fall while riding a bicycle. During the fall, the bicycle’s handle had caused blunt injury to the right side of the forehead and the right eye. Following the incident, he had a brief loss of consciousness and had multiple episodes of vomiting. However, he had no seizure. Initially, he received first aid care in the local hospital and was then referred to Universal College of Medical Sciences, Bhairahawa, Nepal, where he underwent CT scan of head. The CT report showed right frontal lobe bleed of 41 x 12 mm with minimal perilesional edema, intraventricular extension, communities and depressed fracture of the superior medial wall of right orbit with a displaced fragment in the right frontal lobe. There was presence of pneumocephalus, right orbital hematoma, pneumococcus and extension of intraventricular blood in the lateral, third and fourth ventricles. He was managed in the Intensive Care Unit (ICU) for seven days and was then discharged without any surgical treatment. According to the hospital records, he was seen by an ophthalmologist during his hospital stay on June 25, 2018; and his visual acuity was noted as normal (6/6) in both eyes.

Soon after discharge, his mother noticed bulging of the right eye that progressed gradually. She also noticed the pulsatile nature of the right eye bulge. The patient was taken back to the ophthalmology clinic after two months of trauma. Ophthalmology out-patient clinic note revealed that his visual acuity in the right eye and left eye were 6/60 and 6/6 on September 9, 2018. On fundoscopic examination, papilledema and macular edema was seen in the right eye. On ophthalmic examination, his visual acuity was noted to be 2/60 in the right eye and 6/6 in the left eye with diplopia. He was then referred to our Neurosurgical service for further evaluation and management.

Neurological examination at our clinic revealed Glasgow Coma Scale (GCS) score of 15 (E4V5M6). Bilateral pupils were round in shape, regular and reactive to light. He had downward and outward pulsatile proptosis of the right eye (Figure 1A - B). Relative afferent pupillary defect (RAPD) was present in the right eye. His visual acuity was perception to light in the right eye and 1/60 in the left eye.

He was diagnosed as a rare post-traumatic orbital encephalocele with visual impairment and was subjected to bicornal craniotomy and extradural repair of orbital roof. First, a bicornal scalp incision, extending from right tragus to left frontal bossing, was made. An asymmetrical bifrontal (right more than left) craniotomy was made using Stryker electric craniotomy drill. After the removal of frontal bones across the midline, the dura was meticulously dissected off the anterior skull base, exposing the right orbital roof and cribriform plate. There was a large dura defect with brain tissue herniating into the bony defect of the right orbital roof. A small bony fragment was also seen to be pointing towards the frontal lobe. The bony fragment dissected off the surrounding thickened dura and was nibbled away with a small double action rongeur. The bony edge was smoothened. The herniated brain tissue with CSF filled sac was amputated and brain tissue from intra-orbital space was removed. The circumferential thickened dura was meticulously dissected off the anterior skull base. The two leaflets of the dura opening was closed interruptedly using 4-0 polyglactin suture and reinforced with a strip of temporal fascia in a watertight fashion. The bone defect was reconstructed with the inner table split bone graft of the cranium and fixed with silk 4-0 (Figure 3A - C). His post-operative course was uneventful. Post-operative CT-scan of head showed the reconstructed right orbital floor with no communication with right orbital (Figure 4 A, B). His proptosis decreased and pulsation of right eye disappeared completely (Figure 5). The staples over the scalp incision were removed on post-operative tenth day and discharged. However, there was no improvement in vision in both eyes during discharge and even on two-month follow up.

Discussion

Encephalocele is defined as an extracranial herniation of brain parenchyma through a defect in the bony skull. They are classified according to the anatomic region of the bone defect as occipital, head-dome, fronto-ethmoidal and basal. When there is orbital extension, it is known as orbital encephalocele. This classification is important for surgical approach.

Post-traumatic encephaloceles can be iatrogenic or secondary to blunt craniofacial trauma. Encephaloceles following trauma may present clinically as CSF rhinorrhea within days to months following trauma and can be associated with risk of meningitis, which may develop up to 3-8 months post-trauma. Following trauma, there is edema of the retroocular tissues resulting in congestion of the veins which in turn further increases the edema. Edema and venous congestion push the eyeball forwards that stretches the nerves and extraocular muscles further compressing the draining

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Major complication of orbital encephalocele is optic nerve compression. This can arise either due to direct compression from herniation or secondary to raised intra-orbital pressure causing secondary congestion of draining veins including retinal vein, later retinal artery and subsequent blindness. In our case, according to past history, he was initially treated locally for the brain contusion and then discharged. He subsequently developed pulsatile proptosis in over months which meant that the early diagnosis of the large bony defect over the right orbital roof may have been missed, leading to the development of orbital encephalocele and progressive visual impairment in the right eye. However, the diminution of vision in the left eye is plausibly due to the sympathetic injury of the contralateral eye, so-called sympathetic ophthalmia. If proper and timely identification of a large bony defect in the orbital roof was possible, followed by appropriate surgical repair of this defect, may have prevented visual diminution of both eyes. In this case, after late repair, the pulsatile proptosis disappeared and extraocular eye movement in right eye improved substantially; however, the diminished visual sensation did not improve in both eyes.

A small defect may be treated conservatively but a larger one may require a surgical approach. The defect can be closed with a number of options including temporalis fascia, titanium mesh, bone powder, fibrin...
glue, etc. But in our case, we closed the defect using the inner table of the skull bone flap, dural opening was closed directly, reinforced with temporalis fascia and pericranial reflected flap.

### Conclusion

Orbital encephalocele should be suspected in cases of craniofacial trauma associated with orbital roof fractures and frontal contusions. Thin cuts and 3-D reconstruction of CT of the orbit could have revealed the defect so that surgical management could have been performed earlier which could have prevented the visual diminution in both eyes. Therefore, orbital fractures should be recognized initially and if orbital encephalocele has developed, early surgical treatment should be performed to prevent direct and indirect visual complications.

Thus, we emphasize on early diagnosis and surgical management of this rare condition to prevent permanent visual loss and to achieve good cosmetic results.

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