Intraorbital Encephalocele in an Adult Patient Presenting With Pulsatile Exophthalmos
—Case Report—

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Abstract
A 25-year-old man presented with an intraorbital encephalocele manifesting as progressive left pulsatile exophthalmos. He had a history of frontal lobe contusion from a motorbike accident 10 years before the onset of the symptom. Computed tomography and magnetic resonance imaging revealed an oval-shaped defect in the left orbital roof with an underlying intracranial cystic lesion, herniated into the orbit. Intraoperative findings included disruption of the dura mater around the bony defect. The loculated arachnoid membrane and protruding brain tissue were excised with primary dural closure and reconstructive cranioplasty with a titanium mesh. The postoperative course was uneventful, and the pulsatile proptosis disappeared immediately after the procedure. Intracranial cyst may be important in the development of progressive pulsatile exophthalmos and intraorbital encephalocele.

Key words: intraorbital encephalocele, pulsatile exophthalmos, intracranial cyst, traumatic encephalocele, orbital roof fracture

Introduction
Classification of encephaloceles is based on the location within the cranium.7) Encephaloceles commonly occur in the occipital regions (70%), with fewer in the parietal regions (10%), whereas basal encephaloceles account for only 1.5% of all cases.7) Intraorbital encephaloceles, a type of basal encephalocele, are very rare and occur mostly in patients who have experienced head injuries, especially those resulting in orbital roof fractures.2,4,5,8) The most common causes of intraorbital encephaloceles are trauma, tumors, and congenital skull malformations.6,11) Treatment choices for intraorbital encephaloceles include surgical reconstruction of the skull base and duraplasty.3,12,13)

Here, we describe a patient who presented with pulsatile exophthalmos with intraorbital encephalocele and was successfully treated by skull base reconstruction with titanium mesh plate and duraplasty.

Case Report
A 25-year-old man presented with progressive exophthalmos in the 6 months prior to admission to our hospital. He had a history of intracerebral contusion after a motorbike accident 10 years previously. He had no medical history of congenital anomalies, bone dysplasia, or neurofibromatosis. The patient complained of oscillopsia and pulsatile proptosis. These symptoms became particularly apparent after drinking alcohol.

On admission, pulsation of the eyeball could be observed in synchrony with the arterial pressure pulse. Examination by an ophthalmologist revealed asymmetric protrusion of 3 mm in the left eye compared with the right eye by Hertel exophthalmometry. The pupils were equal in diameter and there was no limitation of ocular rotation. Visual acuity and both fundi were completely normal.

Three-dimensional computed tomography (CT) revealed an oval-shaped bony defect 24 mm long and 20 mm wide (Fig. 1). Magnetic resonance (MR) imaging demonstrated a left frontal intracranial cyst just above the bony defect, and an intraorbital encephalocele communicating with the intracranial cyst (Fig. 2).

Under general anesthesia, a lumbar drainage tube was introduced to reduce intracranial pressure prior to craniotomy. A bifrontal skin incision was made inside the hairline. A pericranial flap was used to seal the well-pneumatized frontal sinus, which was opened during craniotomy. The dural defect, adherent to the smooth margin of the bony defect, was observed intraoperatively. An old linear fracture running outward from the lateral margin of the bony defect was also detected. The herniated intraorbital contents and gliotic brain were excised, which revealed the clear margin of the bony defect. (Fig. 3). Primary watertight dural closure was achieved with 5–0 Prolene sutures. The orbital roof was reconstructed with a titanium mesh plate, reinforced with fibrin glue

The postoperative course was uneventful. Three-dimensional CT after the procedure demonstrated that the titanium plate covered the bony defect (Fig. 4) and the intraorbital encephalocele was no longer present. He has shown no recurrence of the symptom for 3 years after surgery.

Discussion

Intraorbital encephaloceles have mostly occurred as an important complication of orbital roof fractures, especially in pediatric patients.  

Orbital roof fractures were found in 7.1% of patients who suffered head injuries, and 13% of those patients developed intraorbital encephaloceles in the pediatric group. All patients who developed intraorbital encephalocele after head injury showed pulsatile exophthalmos in this series. Strabismus and supraocular masses were also observed as complications. Symptoms developed within 1 month after head injury, with the average period between head injury and onset of pulsatile exophthalmos was 18.7 days.

Pulsatile exophthalmos may develop at least 1 year after head injury. Disruption of the dura mater was observed in all patients with delayed onset of trauma-induced pulsatile exophthalmos. A 65-year-old man with delayed onset of traumatic pulsatile encephalocele complained of progressive pulsatile exophthalmos 20 years after craniofacial blunt injury, and autologous bone repair and fascia lata grafts relieved the symptoms. Our adult patient had suffered a motorbike accident 10 years prior to the onset of pulsatile proptosis, and had been hospitalized...
for 1 month. Intracranial contusion was noted at the time of the accident but further details were not available. However, we detected an old linear fracture and tear of the dura mater intraoperatively, which are consistent with traumatic intraorbital encephalocele associated with orbital roof fracture. Trauma-induced intraorbital encephalocele appears to be the most likely cause of the symptoms in our patient but we have no additional clinical evidence (initial CT, continuous image assessment, medical records). It is unlikely that symptoms originated from a neoplastic lesion considering the patient’s medical history, and neuroimaging and intraoperative findings. We did not perform histological examination because his past history and intraoperative findings strongly suggested a trauma-induced etiology, but the cause of the intraorbital encephalocele should be identified by pathological examination. In addition to traumatic encephaloceles, congenital and idiopathic intraorbital encephaloceles have also been reported to cause pulsatile exophthalmos. A 25-year-old patient suffering from neurofibromatosis localized in the cranio-orbital region presented with the main symptom of right pulsating exophthalmos caused by herniation of the fronto-temporal lobe through a defect of the greater wing of the sphenoid.

The natural history of the evolution of skull base defects is controversial. Gradual herniation of the arachnoid into a pre-existing fracture or congenital bony defect, physiologic growth of the cranium and the brain, continuous pulsation of the cerebrospinal fluid, and absence of the bony counter compression and dural defects appear to be the accepted mechanisms for growth and expansion of bony defects, especially in vault encephaloceles. Disruption of the dura mater appears to be essential in the formation of intraorbital encephaloceles, through which the pulsation of the brain gradually erodes bony structures. A gradient between the pressure in the subarachnoid space and the intraorbital compartment may also be important, particularly in intraorbital encephaloceles. Children with orbital roof fractures associated with frontal cerebral contusions are at greater risk for developing intraorbital encephaloceles. This is in agreement with our intraoperative findings.

Three-dimensional CT is recommended as the first step to detect orbital roof bony defects. MR imaging is essential to evaluate herniated intraorbital meningeal or cerebral tissue and to differentiate from intraorbital tumors and vessel malformations. In our case, MR imaging with contrast medium showed no enhanced mass lesion and T2-weighted turbo spin echo MR imaging clearly demonstrated cyst formation in the left frontal lobe.

Treatment of encephaloceles requires that all gliotic brain tissues and arachnoid loculations protruding into the orbital cavity should be exposed completely and all herniated gliotic tissues should be removed, followed by watertight primary closure of the opened dura mater and reconstructive cranioplasty. In our case, we used a titanium mesh for repair of the bony defect. In general, cranioplasty is not always performed, but duraplasty was performed in all previously reported cases. There were no reported surgical complications, and improvement in preoperative ocular symptoms was achieved in all patients.

In conclusion, surgery is recommended for patients with late onset of traumatic intraorbital encephaloceles to achieve favorable outcome without postoperative neurological complications. Neurosurgeons consider the possibility that intraorbital encephaloceles may develop years after head injury.

References


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Neurol Med Chir (Tokyo) 50, December, 2010