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CASE REPORT

Partial thickness autologus calvarial bone orbitocranioplasty for a sphenorbital encephalocele presenting as pulsatile exophthalmos

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ABSTRACT

Basal encephalocele accounts only 1.5% of all encephaloceles. But Sphenorbital encephalocele is the rarest cause of herniation of brain into orbit leading to pulsatile exphothalmos. Authors presenting a case of sphenorbital encephalocele in a 16 yrs old girl successsfully managed by orbitcranioplasty by partilal thickness autologus calvarial bone graft.

Key words: Cranioplasty, pulsatile exophthalmos, spenorbital encephalocele

Introduction

Herniation of the brain or meninges through congenital skull defect is classified on location of cranium.[1] Occipital encephalocele is the commonest, seen in 70% cases and parietal in 10% and basal encephalocele account, only 1.5% of cases.[2] We describe a rare type of basal encephalocele. Sphenorbital encephalocele presenting as pulsatile exophthalmos is successfully repaired by partial thickness autologus calvarial bone graft.

Case Report

When this patient was 10 years old, her parents noticed pulsation in her right eye. Right eye was gradually coming out when she bends forward. X-ray skull and CT scan obtained at a neighborhood hospital showed bone defect in the posterior wall of right orbit. She was followed up with CT examinations but without treatment and her symptoms remained for 6 years. She was referred to us at the age of 16 years. She was conscious, oriented, and her right eye showed pulsatile exophthalmos [Figure 1].

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An ophthalmological examination revealed punctate keratitis. Her visual acuity was 20/40 in right eye and 20/25 (no change) in the left eye. Movements of both eyes were full and the visual field was intact. Light reflex was prompt bilaterally and no neurological deficits were noted.

She had multiple subcutaneous nodules in arm, forearm, and trunk with axillary freckling. Her family history revealed that her father has multiple subcutaneous nodules in face, neck, and arm and diagnosed to have neurofibromatosis type 1. Genetic screening was not performed.

CT Scan Head demonstrated agenesis of right sphenoid wing and bone defect in posterolateral wall of Right orbit [Figure 2].

Magnetic resonance imaging revealed tip of right anterior temporal lobe protruding toward the right orbit with hypointense sac anterior to right temporal lobe in T1W image and hyperintense in T2W image indicative of meningocele [Figure 3].

Operation - After investigations and pre-anesthetic check up, patient was taken up for surgery with due counseling. The patient was placed supine with head turn to left side approximately 45° and lumbar drainage tube was introduced under general anesthesia.

A curved incision was made inside the hair line from the midline to the level of upper border of zygomatic arch. Sub periosteal dissection along the orbital rim freed the periorbita from the orbital wall. Standard frantotemporal craniotomy performed. The tip of temporal lobe protruded forward through bone defect to the orbit. As there were adhesions between periorbita and duramater of temporal lobe, anterior temporal lobectomy, and adhesiotomy done. The defect in

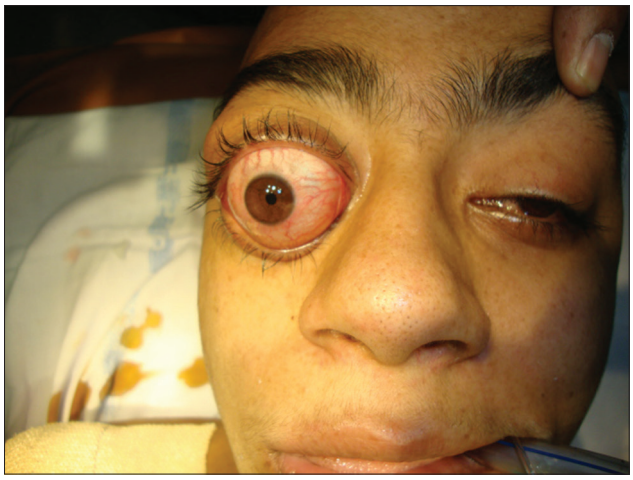

Figure 1: Preoperative photograph of the patients eye showing right exophthalmos

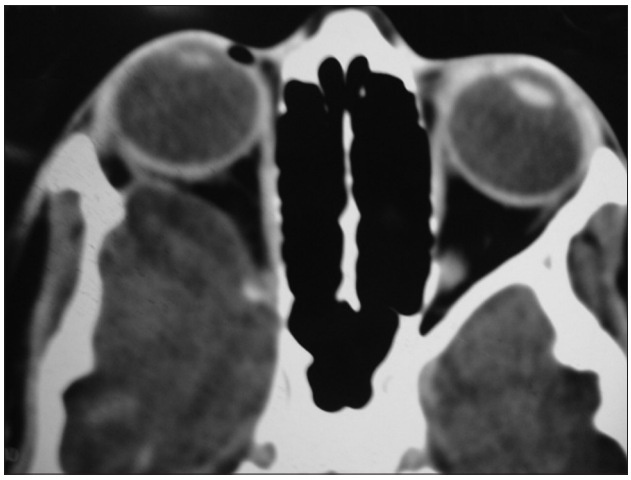

Figure 2: CT axial showing right temporal lobe protruding toward the right orbit

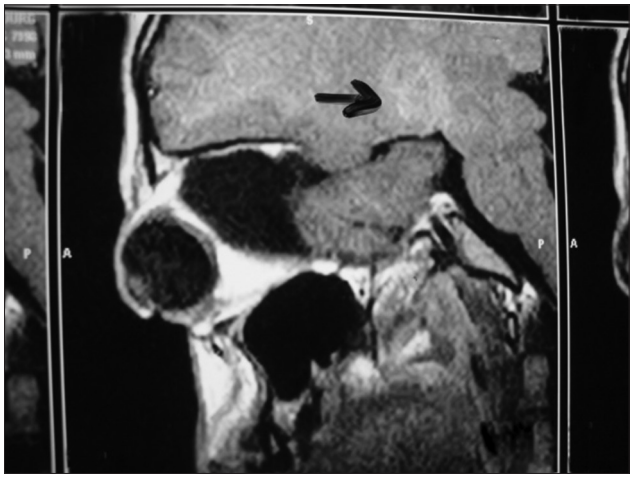

Figure 3: MR coronal T1 weighted showing tip of right temporal lobe protruding toward orbit with encephalocele

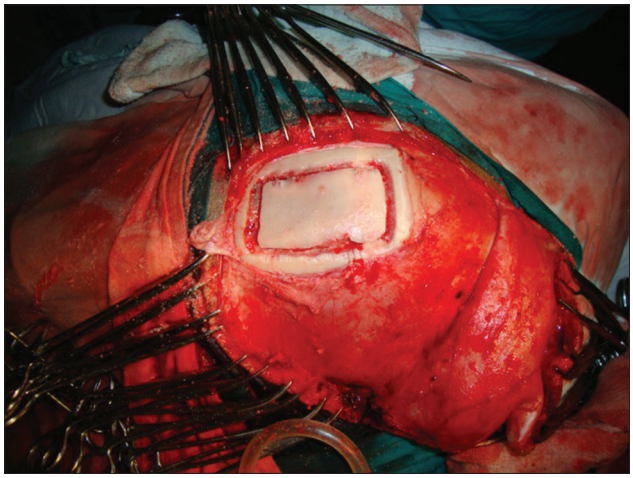

Figure 4: Operative photograph showing harvesting of partial thickness autologous bone graft

posterolateral wall of orbit was identified. Partial thickness bone graft was harvested from parietal region by drill and giggle saw [Figure 4]. The bone graft was inserted from posterolateral orbital margin above the periorbita and fixed with micro screw and plate with orbital wall. Thin facial graft was put behind the bone graft and fibrin glue was applied. Dura was closed, original bone flap reposited and scalp incision was closed in multi layer fashion. Postoperative period was uneventful without neurological deficit. The pulsatile exophthalmos disappeared after surgery with intact vision and no neurological deficit.

Post-operative 3D CT scan revealed the appropriate placement of implant covering the bone defect and sufficient space for the orbit. No compression of temporal lobe was seen [Figures 5-7].

Discussion

Consul and Kulshrestha[3] classify the orbital encephalocele into anterior and posterior group. Anterior ones arise from frontal and lacrimal bone, cribriform plate and nasal process of superior maxilla and posterior one arises (uncommon) from sphenoidal fissure, optic foramen. Posterior ethmoidal foramen and roof or wall of orbit.[4] Basal encephalocele comprises ethmoidal, sphenoethmoidal, transsphenoidal, and sphenorbital type.[1]

As described, we successfully repaired a bone defect causing sphenorbital encephalocele with partial thickness calvarial autologus bone graft. Preoperative planning with skull bone model useful in determining the appropriate size bone graft and its placement. There have been several reports of surgically treated sphenorbital encephalocele. Odake *et al*.[5] surgically repaired a defect from temporal bone to superior orbital fissure in 32 year old female with a small part of the frontal bone replaced by resin. Clauser *et al*.[6] described a 25-year-old male whose anterior skull base was reconstructed by splitting part of parietal bone we adopted our repair from this report. Sugawara *et al*.[7] reported a case of an 11-year-old boy treated using a two

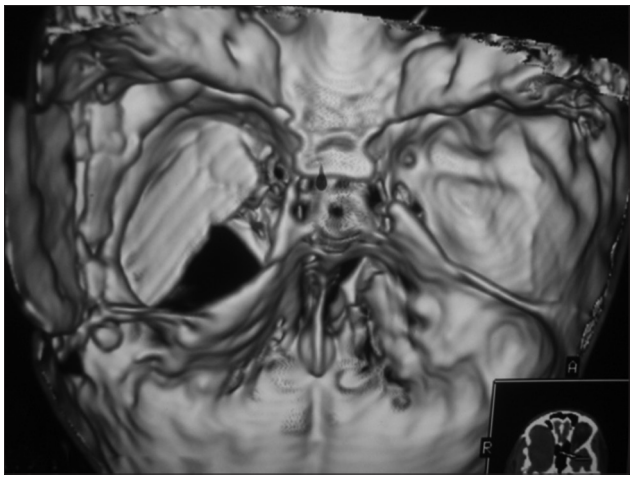

Figure 5: CT reconstruction showing graft *in situ*

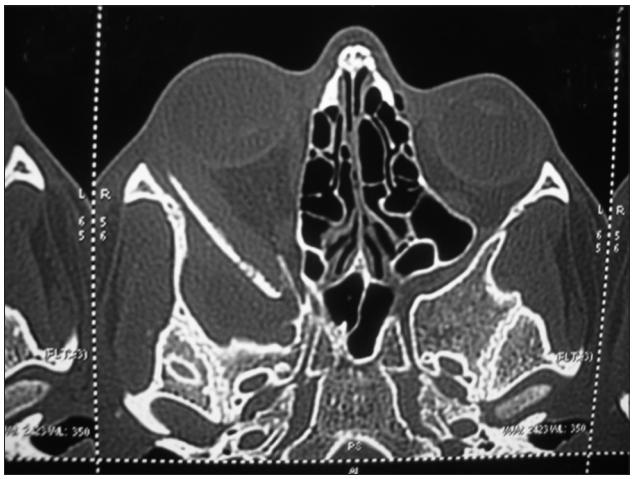

Figure 6: CT axial bonycuts showing graft in place

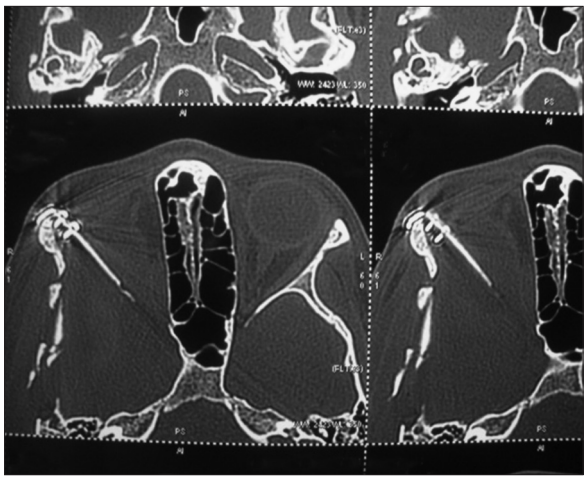

Figure 7: CT axial cut showing graft, plate, and screws in place

stage repair. The authors reconstructed the bone defect in superior and lateral wall of orbit and temporal bone with inner table of frontal bone; 4 months later they placed a hydroxyapatite ceramic implant subperiosteally to correct malposition of graft. Neurofibromatosis associated with skull deformation, sphenoid bone dysplasia, and defect. Authors of several reports have demonstrated a relationship between sphenorbital encephalocele and this disease.[5,6,8] In present case, we found, multiple subcutaneous nodules and axillary freckling with hereditary component. This case satisfies the criteria for neurofibromatosis.

Size and shape of bone defects in congenital encephalocele may vary, and small defects identified as orbital encephalocele can often be managed with small fragments of muscle and fascia.[8-10]

Hydroxyapatite ceramic is an excellent compensatory material for cranioplasty, having high affinity for biomaterials and the capability of being molded in to desired contour. It is suitable for repair of defect of cranial anomalies of whatever size and shape. Moreover, as hydroxyapatite contains multiple micropores into which osteogenesis can extend by osteoconduction.[11] However, the possibility of an imbalance developing between an implant and the autologous bone should always be taken into account. Reconstruction by using autologus split calvarial bone is an attractive option that can be fixed by micro plate and screw[7] as weadopted in our case to fix the implant and gave equally rigid fixation.

Authors of most of the previous reports described the resection of herniated brain for repair of encephalocele.[4,7,9,12] However, in our case, we also resected the herniated anterior temporal lobe. This helped us for comfortable repair of defect.

Conclusion

Sphenorbital encephalocele is a rare entity. Standard methods of treatment are not well established. Many authors have used the artificial constructed implants for repair. But we successfully repaired the sphenorbital encephalocele by harvesting partial thickness bone graft from same patient and fixed by micro plate and screws with satisfying results.

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