An ophthalmological examination revealed punctate keratitis. Her visual acuity was 20/40 in the 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 the arm, forearm, and trunk with axillary freckling. Her family history revealed that her father has multiple subcutaneous nodules in the face, neck, and arm and was diagnosed to have neurofibromatosis type 1. Genetic screening was not performed.

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

Magnetic resonance imaging revealed the tip of the right anterior temporal lobe protruding toward the right orbit with hypointense sac anterior to the 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 turned to the 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 the upper border of the zygomatic arch. Subperiosteal dissection along the orbital rim freed the periorbita from the orbital wall. Standard frantotemporal craniotomy performed. The tip of the temporal lobe protruded forward through bone defect to the orbit. As there were adhesions between periorbita and duramater of the temporal lobe, anterior temporal lobectomy, and adhesiotomy done. The defect in
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\(^1\) 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, sphenethmoidal, 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
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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 autologous split calvarial bone is an attractive option that can be fixed by micro plate and screw[7] as we adopted 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.

References


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