Lateral frontal encephalocele associated with dysplasia of orbit, eyeball, and eyelid

A. Kulali and Ö. Rahmanli

Department of Neurosurgery, Dicle-University School of Medicine, Diyarbakir, Turkey

Received January 17, 1989 / Revised May 25, 1989

Abstract. A case is reported of a right lateral frontal encephalocele without communication with the midline structures and in concurrence with dysplasia of the orbit, eyeball, and eyelid.

Key words: Lateral encephalocele – Coloboma – Scalp defect – Split skull cranioplasty

Cephaloceles are congenital malformations consisting of a defect in the cranium and the dura mater with extracranial herniation of intracranial structures and they occur about once in 4,000 to 5,000 live births [1]. Occipital and frontal regions are the sites of predilection. Frontal encephaloceles occur once in every 35,000 live births. With few exceptions, they are found in the midline of the cranium. In rare instances, the sac may be found on the lateral aspects of the cranium usually near a suture line. Concurrent optic malformations including unilateral coloboma or hypoplasia of the eye and orbit with encephaloceles have been reported in few cases [9].

We have treated a case of encephalocele on the lateral aspect of the frontal region with coloboma and hypoplasia of the eye and orbit. Our search did not lead to any report of a similar occurrence in the literature.

Offprint requests to: A. Kulali

Fig. 1. Soft swelling just above the right orbit protruding over the eyebrow and eyelid with hypertelorism, exophthalmus, and exotropia

Fig. 2. Frontal view of skull radiograph showed an oval large bone defect in the right frontal region just above the eyebrow. Right orbit is smaller than the left one
Case report

A 4-year-old boy presented with a soft swelling in the right lateral aspect of the forehead just above the right eyebrow which had been there since birth and was increasing in size. There were blindness and split of the eyelid on the right side. On examination, a soft swelling just above the right orbit protruding over the eyebrow and eyelid was found. There was a scar which represented a previous attempt to biopsy the swelling (Fig. 1). There were hypertelorism, exophthalmos, and exotropia on presentation. No other anomalies were neurologically found.

The radiographs of the skull showed an oval large bone defect in the right frontal region just above the eyebrow. The right orbit was smaller than the left one (Fig. 2). Computed tomography scans revealed a large osseous defect extending from the roof of the right orbit to the frontosphenoidal suture. Herniation of the large part of the right cerebral hemisphere through the osseous defect resulted in the rotation and hypoplasia of the right eyeball and orbit (Fig. 3).

Surgery through a bicoronal scalp incision revealed an oval bony defect in the lateral frontal region, 4 to 6 cm in diameter, just above the right orbit without any extension into midline, but however into suture frontozygomatic and sphenofrontalis surrounded by smooth thickened bone edges. The frontal pole of the brain covered with scarred and thickened dura herniated with a wide neck through the same defect under the well-developed scalp. The dura was loosed from the surrounding smooth edges of the bony defect and opened in cross-shaped incision. The solid mass was resected from its origin at the pole of the right frontal lobe. The dural sac also was reduced in size and closed watertight. To prevent future recurrence and restore the plastic reconstruction of the cranium defect in the forehead we carried out a split-skull cranioplasty by using the outer table of the autogenous bone graft removed from the left frontoparietal region (Fig. 4).

The postoperative course has been uneventful (Fig. 5) and no additional clinical and neurological deficits have occurred postoperatively (Fig. 6). The histological examination confirmed the preoperative diagnosis of an encephalocele.