Radical Cranio-orbital Reconstructive Procedures for Cloverleaf Skull Deformity in Adult: Operative Technique for the Longest Survivor—Case Report—

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Abstract

Cosmetic cranio-orbital reconstructive surgery was carried out on a 22-year-old male, the longest surviving case of cloverleaf skull syndrome reported. He previously underwent classical linear suturectomy for synostotic sutures and temporal cranioplasty. Fortunately, hydrocephalus became arrested so did not require continuous cerebrospinal fluid drainage through shunt tube. His intelligence quotient was in the 40s. The present problem was mainly of cosmetic cranio-orbital corrections of shallow orbits with resultant exophthalmos, frontal dysgenesis, and marked temporal bossings. Bilateral orbital advance, lateral canthal/pterional reshaping, frontal remodeling, and temporal reduction cranioplasty were performed. The postoperative outcome was satisfactory. The cloverleaf skull deformity is etiologically and pathologically heterogeneous, so radical surgical reconstructive procedures should be planned and designed individually.

Key words: cloverleaf skull, craniofacial surgery, orbital advancement, frontal remodeling, reduction cranioplasty

Introduction

The cloverleaf skull syndrome was first described by Holtermüller and Wiedermann in 1960 using the evocative German term “Kleeblattschädel-Syndrome.” They found 12 previous cases in the German literature dating from 1824. The classical features of this syndrome are 1) a trilobar skull configuration with ears downwardly displaced, 2) facial deformities including hypertelorism, flattened nasal root, and jaw abnormalities, 3) micromyelia and skeletal abnormalities, 4) radiological features of trilobar or cloverleaf skull and hydrocephalus, and 5) poor prognosis. Generally, the trilobar appearance of the head is caused by a premature intrauterine closure of the coronal and lambdoid sutures associated with internal hydrocephalus and cerebral protrusion through the open sagittal and squamosal sutures. However, different sutures may be involved. For example, synostosis of the sagittal and squamosal sutures with cerebral protrusion through a widely patent anterior fontanel may occur. Trilobar skull has occurred with complete synostosis of all cranial sutures, or with widely patent sutures and no evidence of prenatal craniosynostosis. Recent analyses of anatomic and histological observations have found striking variations. These studies suggest that cloverleaf skull malformation is etiologically and pathologically heterogeneous. Radical corrective surgery must be based on individual variations.

We report a surgical approach for cloverleaf skull in a male who previously underwent linear craniectomy and temporal cranioplasty at 4 and 12 years old. This is the longest surviving case reported.

Case Report

A 22-year-old male with trilobar skull deformity underwent sagittal paramedian linear craniectomy and strip suturectomy of the bilateral coronal, lambdoid and part of the squamosal sutures when aged 4 followed by bitemporal cranioplasty at 12 years old. A ventriculoperitoneal (VP) shunt was emplaced at 5 years old but was removed after a week because of
severe low pressure syndrome. Head circumference reached 64 cm at 12 years old but has not increased since then so the hydrocephalic state apparently became arrested. He graduated from junior high school and works as a farmhand. He was admitted to our hospital for reconstructive cosmetic surgery.

On admission, he was alert and well oriented, but was mildly retarded (intelligence quotient = 40, Tanaka-Binet method). The trilobar head was prominent and 64 cm in circumference (Fig. 1 upper). Cranial nerve functions were intact. There were no motor or sensory deficits. Plain skull x-ray films demonstrated the previous craniectomies of the trilobar skull. The frontal fossa was shallow and contracted anteroposteriorly. The bilateral middle fossae were greatly expanded and deepened. The petrosal bone was located far inferior and the external auditory meatus was open at the foramen magnum level. The skull base was nearly flat along the laterally and inferiorly elongated petrous bone edges and temporal fossae (Fig. 1 lower). Computed tomographic (CT) scanning and magnetic resonance (MR) imaging confirmed severe ventriculomegaly especially of the bilateral temporal horns, body, and anterior horns of the lateral ventricles (Fig. 2 upper). The frontal paranasal cavities were well developed. The shallow orbits were caused by the hypoplastic frontal fossa and posterolateral compression from the expanded temporal bones. Temporoparietal synostosis had caused bilateral pillars (Fig. 2 lower).

Bilateral temporofrontal reshaping, orbitotomy and orbital reconstruction were designed based on the skull deformity characteristics. The diagnostic imaging showed a marked depression at the pterion because of the temporoparietal synostosis pillars, which also caused a vertical shift of the lateral sphenoid bone parts and configuration of the temporal bone (Figs. 1 and 2). There was localized hyperostotic thickening in the right frontal bone.

Fig. 1 upper: A 22-year-old male with cloverleaf skull. Note the oxycephalic head with bitemporal bossing and the low-set ear. The orbits are shallow and relative proptosis is observed. The operative scars are from previous procedures. lower: Plain skull x-ray films demonstrating the previous bilateral craniectomy sites. The frontal fossa is shallow and the bilateral temporal middle fossae are extremely expanded and deepened. The petrosal bone is located far inferior and the skull base is nearly flat.

Fig. 2 upper: Coronal and sagittal MR images (TR 3100 msec, TI 500 msec) revealing severe ventriculomegaly especially of the massively expanded temporal horns. lower: The orbits are small and shallow, being compressed posterolaterally by expanded temporal bones.

Neurol Med Chir (Tokyo) 31, February, 1991
The thin bilateral orbital rims and depressed pterional regions resulted in relative proptosis (Fig. 3 upper). Therefore, orbital reconstruction with pterional/frontal reshaping and temporal reduction cranioplasty were planned. The right frontal bone, superolateral orbital rims, orbital roofs, and temporal bones were removed. The massively expanded temporal bones prevented identification of the inferior orbital fissures and zygomatic arches. The temporoparietal pillars were a good landmarks for the temporal reshaping line. The superolateral orbital rim advance and pterional reshaping were achieved with multiple bone fragment bridges (Fig. 3 lower). The dural tension was within normal limits or slightly hypotensive after frontal base expansion. The temporal reduction cranioplasty was then achieved without significant compressive effects. The frontal and temporal remodeling used an individually designed methyl methacrylate plate (Fig. 4 left). Similar procedures mainly for orbital advance and temporal reduction cranioplasty were performed on the left.

Postoperative course was uneventful. His neurological status remained unchanged. Wound healing was uneventful except for some subgaleal fluid ac-

The skull deformity viewing superolaterally and after skin eversion. Note depression of the pterional region. lower: Osteotomy for orbital hypoplasia, frontal dysplasia, and temporal bossing and orbital advance/lateral canthal/pterional reconstruction.

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Discussion

Cloverleaf skull typically demonstrates premature fu-
sion of the coronal and lambdoid sutures. However, cranial base development displays varying premature cranial base synchondrosis ossification. In severe cases, surgery aims to open the nasopharyngeal airways by advancing the naso-maxillary-zygomatic complex, decompress the brain, remodel the skull, and expand the bony orbits. The prognosis depends on these critical morphological skull base changes, progression of associated hydrocephalus, brain dysgenesis and/or systemic anomalies. Fortunately, in the present case hydrocephalus became arrested without shunt dependence and there were no associated severe intra- or extracranial morphological malformations other than the craniofacial deformity. The classical decompressive procedure may have preserved the brain function and this is the longest surviving case. Previously reported cases survived for 6.8,14 and 14 years. Shunt emplacement is generally essential for good prognosis. Cloverleaf syndrome without associated hydrocephalus occurs in a few cases diagnosed in utero, but the present case is exceptional.

Watters et al. described the survival rate in 24 reported cases was 16% (four patients). In 1975, Muller and Hoffman reported early radical surgery for cloverleaf skull syndrome which achieved a reasonable cosmetic result. They found only two previous reports of surgical treatment and believed theirs was the first successful repair of a cranial deformity. A few cases have since undergone radical reconstructive surgery in early infancy. Kroczek et al. performed subtotal craniectomy, supraorbital ridge advance and midfacial advance on a 3-month-old male infant. Complications of cerebrospinal fluid leakage and unsatisfactory midface fixation because of thin cranial bones and limited support for the small metal plates occurred postoperatively. The patient died of acute meningitis. Muller and Hoffman chose two-stage complete circumferential craniectomy and medial displacement of the frontal bones after lumboperitoneal shunt placement. Frank et al. described the appropriate chronological development of a 4-year-old child after VP shunt and cranial suture release. Early aggressive multidisciplinary intervention for respiratory support and control of hydrocephalus can achieve relatively normal growth and development in children with this anomaly. The procedures used in the present case, early intracranial decompression including shunt operation and late cosmetic craniofacial surgery may therefore be the best choice for cloverleaf skull syndrome.

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