Peninsula Shaped Craniectomy for Delayed Holocalvarial Synostosis

Geç Başvuran Holokalvaryal Sinostozlarda Yarımada Şeklinde Kraniyektomi

PAMİR ERDİNÇLER, MEHMET YAŞAR KAYNAR, BÜLENT CANBAZ, NEJAT ÇIPLAK, ALİ ÇETİN SARIOĞLU, CENGİZ KUDAY

İstanbul University Cerrahpaşa Medical Faculty Department of Neurosurgery (PE, AÇS, CK), and Institute of Neurological Sciences (MYK, BC, NÇ), İstanbul, Turkey

Abstract: Delayed holocalvarial synostosis is an uncommon form of craniosynostosis. We report in this article two children with delayed holocalvarial synostosis. Neurological examination was normal but skull X-rays showed diffuse digital impressio and synostosis of all calvarial sutures in both patients. The patients were operated with a modified pi craniectomy technique which relaxes entire calvarium in coronal, axial and sagittal plans. Control X-rays after surgery showed relief of digital impressio in both patients.

Key Words: Craniosynostosis, digital impressio, surgery

Özet: Tüm kalvaryal sütürleri kapsayan geç ortaya çıkan sinostozlar kranyosinostozun nadir görülen bir durumudur. Bu makalede gecikmiş holokalvaryal sinostoz bulguları ile başvuran iki çocuk hasta sunulmuştur. Nörolojik muayenelerinde bir özellik saptanmayan olguların kranyografilerinde yaygın dövülmüş bakır manzarası ve tüm kalvaryal sütürlerde sinostoz saptandı. Hastalar, tüm kalvaryumu koronal, aksiyal ve sajital planlarda serbestleştirecek şekilde, modifiye pi kranyektomi tekniği kullanılarak ameliyat edildiler. Kontrol kranyografileri iki olguda da dövülmüş bakır manzarasında azalma gösterdi.

Anahtar Sözcükler: Cerrahi, dövülmüş bakır manzarası, kranyosinostoz

INTRODUCTION

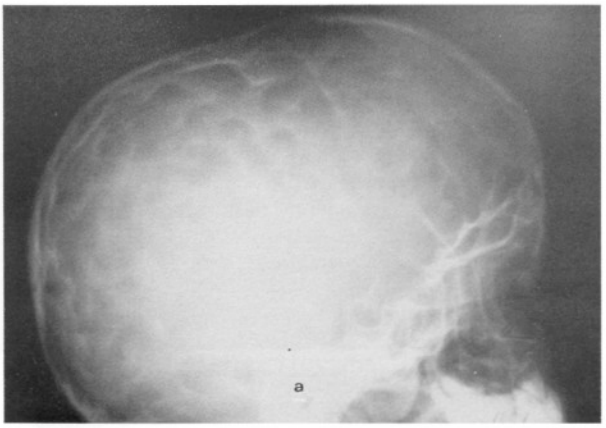
Most of the different types of craniosynostosis have known to be apparent at birth. Oxycephaly can have a clinical manifestation later, by the age of two, with signs of increased intracranial pressure (ICP). Reddy et al. (12) focused attention on patients with delayed and progressive multiple suture craniosynostosis. They concluded that the patient exhibiting these rare situations is distinctly uncommon than the other types of craniosynostosis.

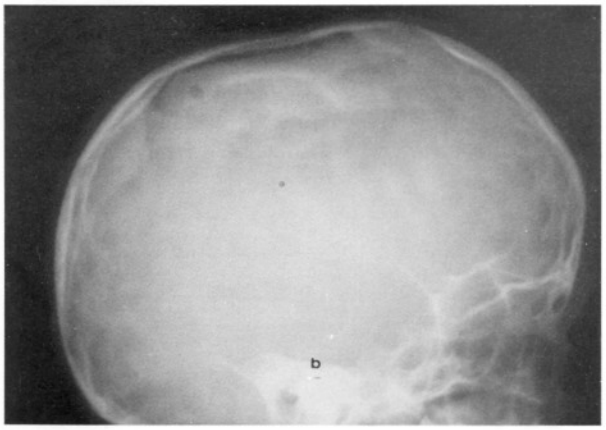
Recently, we have operated two patients with delayed holocalvarial synostosis. We discuss in this article the clinical and radiological pictures of delayed holocalvarial synostosis and our surgical technique for management.

REPORT OF CASES

The first patient was a 5-year-old boy. He was referred to our unit due to headache and dolicocephalic skull shape. Neurological examination

was normal. Neuropsychiatric evaluation showed a slight psychomotor retardation, which was attributed to his being without a mother. Bilateral papilledema was detected at fundus examination. Skull X-rays showed diffuse digital impressio over the calvarium and synostosis of all calvarial sutures (Figure 1, a). Skull shape was slightly dolicocephalic but there was not a prominent frontal or occipital boss. In computerized tomography (CT) scans, skull base was normal, cerebral sulci were effaced and lateral ventricles were narrowed suggesting a high intracranial pressure.





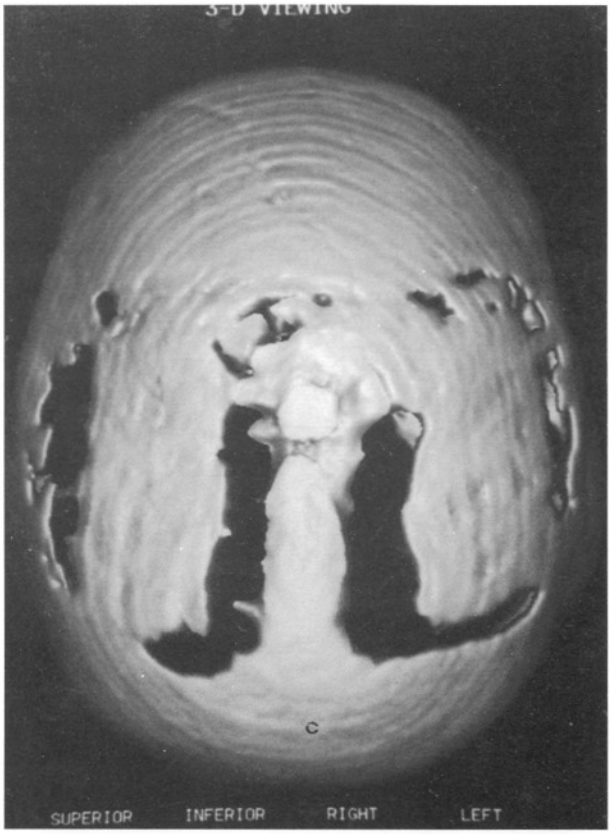


Figure 1. Pre and post operative radiologic evaluation of the first patient: a. Note diffuse digital impressio and synostosis of all calvarial sutures in preoperative lateral x-ray radiograph, b. Digital impressio were decreased 3 years after the surgery at lateral radiograph, c. 3D reconstructed CT scan show partial refusion of bones at the craniectomy borders.

The presenting complaint in the second, 2-year-old male child, was bossing over the bregmatic fontanel, detected by the parents, after a minor head trauma at 1 year of age. Neurological examination, neuropsychiatric evaluation and fundus examination were normal, but a "volcanic shape" bregmatic bossing was palpable over the calvarium. Skull X-rays showed diffuse digital impressio over the parietal and occipital bones, holocalvarial synostosis and bregmatic explosion of frontal and parietal bones, whereas skull X-rays at 1 year of age showed no abnormality and patency of all calvarial sutures (Figure 2 a, b and c). 3D reconstructed CT scans confirmed synostosis of all calvarial sutures and showed three dimensional view of this bregmatic boss (Figure 2 d).

Surgical technique and results

Both patients were operated with the same surgical procedure. Entire exposure of the calvarium was provided with gell-filled collar technique for operative positioning (2) and with a bicoronal skin incision. After the periost dissected in a similar fashion to the skin incision, a pi craniectomy, first described by Jane et al. (4), was done and the parasagittal craniectomies were extended towards both lambdoideal sutures. Then, the ends of coronal and lambdoideal craniectomy were curved

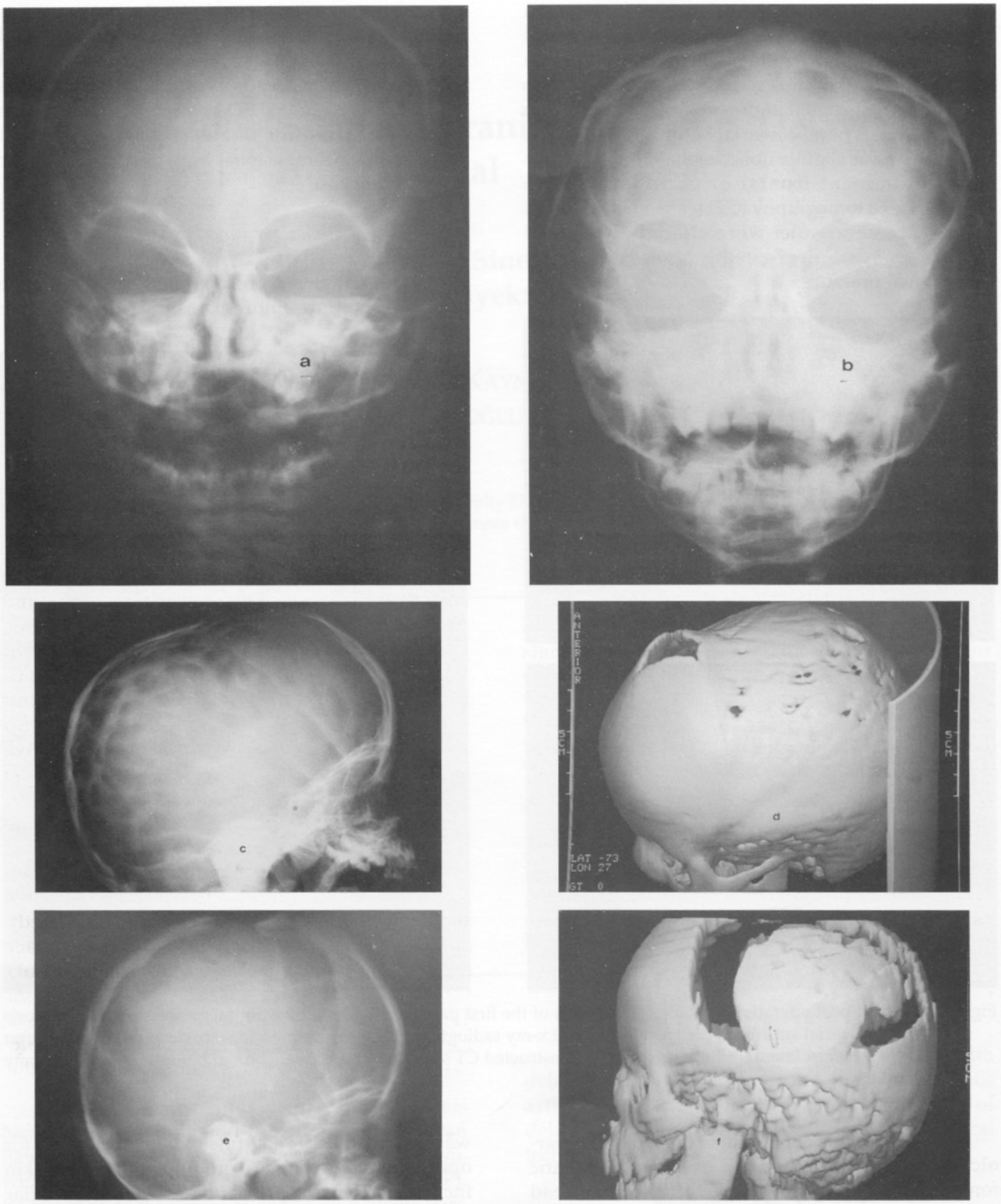


Figure 2. Pre and post operative radiologic evaluation of the second patient: a. Antero-posterior radiograph of second patient at 1 year of age show no any abnormality; b, c. Antero-posterior and lateral radiograph of the same patient shows digital impressio over the parietal and occipital bones, holocalvarial synostosis and bregmatic bossing, 1 year after, d. Three dimensional view of the volcano shaped bregmatic boss and synostosis of all calvarial sutures are well seen in 3D reconstructed CT scan, e. Digital impressio were completely disappeared 4 months after the surgery in skull x-rays, f. 3D reconstructed CT scan shows the craniectomy size and the created peninsula shaped parietal bones.

respectively posteriorly and anteriorly and thus, large "peninsula shaped" parietal bones were created. The bregmatic boss in the second patient was left in place like an island in the bregmatic part of the craniectomy flap in order to avoid any possible damage to the superior sagittal sinus.

Postoperative course was uneventful in both patients, except a decrease in hematocrit levels which didnot necessitate blood transfusion. Skull X-rays and 3D reconstructed CT scans of the first patient showed the craniectomy size and partially resynostosed frontal and parietal bones, 3 years after the surgery (Figure 1 b and c). The digital impressio were decreased and the patient had no any other complaints and he had a normal performance at the school. The second patient showed a dramatically decrease of digital impressio in skull X-rays at the end of four months after the operation (Figure 2 e). The 3D reconstructed CT scans showed the extent of the craniectomy to the both lambdoideal sutures (Figure 2 f). The bregmatic boss completely disappeared at the end of 18 months after surgery.

DISCUSSION

Various attempts have been made in the past to better understand the pathogenesis of craniosynostosis. The role of the skull base for programming the future aspect of craniofacial growth is now widely accepted (10,11,15). The results of premature fusion of sutures are in part manifested by an abnormal cosmetic appearance, and on the other hand, by various neurological complaints such as increased ICP and related psychomotor retardation, epilepsy or neuro-ophtalmological signs (1, 6, 13, 14). The two patients in this article had no cosmetic deformity except a bregmatic bossing in one.

As seen in CT and 3D reconstructed CT scans, there was no involvement of skull base or facial bones in our patients as in 4 cases reported by Reddy et al. (12). Zygomatic angle which seems to be non-affected in 3D CT scans was the indirect sign of the absence of basal synostosis (9). This situation must be reserved for patients in whom craniosynostosis was manifested at birth. In delayed holocalvarial synostosis, most of the calvarial suture fusion takes place after the facial and basal skull development is completed (12). Head shapes were normal in one patient, but slightly dolicocephalic in the other, suggesting a first attempt of sagittal suture. Delayed holocalvarial synostosis must be differentiated from oxycephaly. Marchac et al. (8) call oxycephaly the result of a rather late closure of the coronal suture and announce that they have never seen oxycephaly in children younger than 2 or 3 years of age. Although a round skull is sometimes seen, a pointed appearance of the vault, a short orbital roof and resultant exophtalmus, absence of fronto-nasal angle are considered as the keywords of this anomaly (6, 8). These features were absent in our patients. In the second case, an "explosion" of the bregmatic fontanel after first year of age was the alerting sign of an anomaly. It's probably a herniation of the brain from a single opening of all sutures synostosed calvarium. This event reflect the high capacity of the brain to grow in the first years of life and does not reflect oxycephaly.

The surgical correction of craniosynostosis has evolved from simple removal of the suture to the various applications of cranio-facial principles for better esthetic and cosmetic results (3,5,14,17). Linear craniectomy techniques are generally proposed for scaphocephaly in infancy (14,16). The treatment procedure proposed by Reddy et al. (12) for holocalvarial synostosis was calvarial reshaping and creation of new sutures by morcellation craniectomy. The important factors for better relaxation of the brain can be created in a multitude of ways e.g. vault reconstruction utilizing split cranial bone grafts and judicious use of plates or transposition techniques such as advocated by Marchac and Renier (7). We have programmed our procedure not only for the creation of new sutures by linear craniectomy but also for the relaxation of the brain by extending the craniectomy as far as the temporal fossa as described above. The extending "peninsula shaped" parietal bones to both of the coronal and lambdoideal sutures and to the temporal fossa, as we have done correctly in the second case, is an important factor for better relaxation of the brain. The neck of the "peninsula shaped" parietal bones must be left in the squamosal part of the temporal bone in order to give a malleability even if in the face of thickened bones at later ages. We have observed preoperatively, the relaxation of the brain manifested by amplitude augmented brain pulses, immediately after the entire craniectomy was achieved in both cases.

This minimalist approach is safe, does not need any fixation or reshaping and allows a complete relaxation of the brain in coronal, sagittal and axial plans.

Correspondence: Pamir Erdinçler
PK 19, 34312
Kocamustafapaşa, İstanbul, Turkey
Tel: 212-587 65 85
Fax: 212-273 24 77

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