Singapore Med J 1997; Vol 38(5):217-220

Cloverleaf skull - When Should One Operate?

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ABSTRACT

Cloverleaf skull or *kleeblattschadel* usually occurs as a result of premature closure of the cranial sutures, with accompanying hydrocephalus, proptosis, cranial base and mid-face hypoplasia. The incidence of this abnormality is rare, with less than 130 reported cases in the literature, and is associated with various congenital syndromes. Conventional surgical management advocates shunting prior to definitive craniotomy and bone remodeling. We report an unusual case of Pfeiffer syndrome with associated cloverleaf skull deformity, in which early surgery was vital for decompression of raised intracranial pressure and shunt independence for the first six months. A secondary craniofacial procedure was performed at nine months of age to improve cosmesis. In such cases, good outcome can be achieved if aggressive surgery is combined with good paediatric anaesthesia and intensive care.

Key words: cloverleaf, craniotomy, early surgery, hydrocephalus

INTRODUCTION

The cloverleaf skull syndrome or *kleeblattschadel* presents at birth with a dramatic cosmetic deformity involving an elevated tower-shaped skull, marked convexity of the squamous portion of the temporal bones, ocular proptosis and mid-face hypoplasia. It may occur as a result of coronal or lambdoid suture synostosis or a combination, with hydrocephalus and cranial base dysplasia[1]. Owing to the rarity of this condition, with less than 130 cases in the literature, surgical management is still controversial in terms of timing of surgery and the need for cerebrospinal fluid (CSF) shunting. We report a case of cloverleaf deformity of the skull in which early surgery was beneficial in preserving vision, reducing raised intracranial pressure and improving cosmesis.

CASE REPORT

A full-term male baby weighing 3.5 kg was born by normal vaginal delivery in our hospital. The mother was aged 35 years, with two normal children, and the father was aged 40 years. Both parents were in good health with normal physical appearances. The baby however was noted to have severe proptosis bilaterally with a cloverleaf-type skull deformity (Fig 1). In addition, some features of Pfeiffer syndrome were present, namely broad distal phalanges of the thumb and hallux, low-set ears, small nose with low nasal bridge, choanal stenosis and maxillary hypoplasia[1,2].

On plain radiographs, the characteristic tower-shaped skull with hyperostotic bone was noted. Lateral skull X-ray demonstrated the three main radiological features of cranio synostosis, ie skull deformity, exaggeration of convolutional impressions and absence of part or whole of one or more vault sutures (Fig 2). Axial CT scans of the brain confirmed the plain film findings. Both lambdoid sutures were closed in part (Fig 3). The coronal sutures were unfused whereas the sagittal suture was not clearly seen. The temporal and frontal horns, and body of the lateral ventricles were dilated, with a normal third ventricle (Fig 4).

Treatment

Early surgery was performed at four weeks of age because of the alarmingly rapid progression of height of the skull vault, increasing anterior fontanelle tension, and corneal abrasions consequent to severe proptosis. The body weight at the time of surgery was 4 kg.

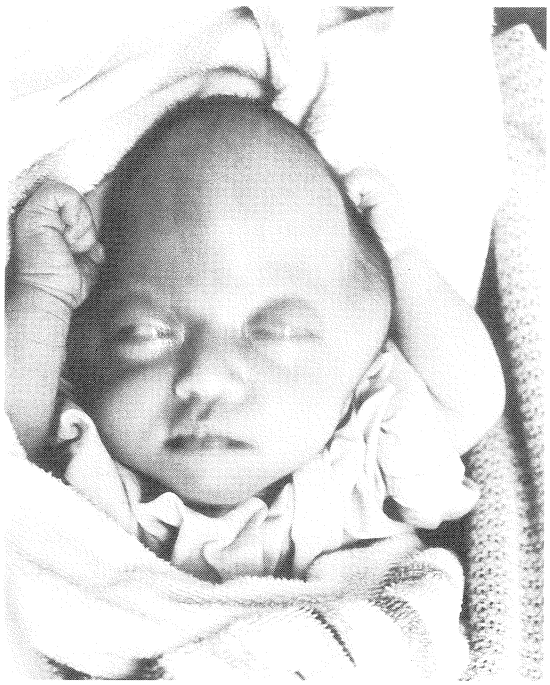

Fig 1 - Clinical photograph of appearance of cloverleaf deformity

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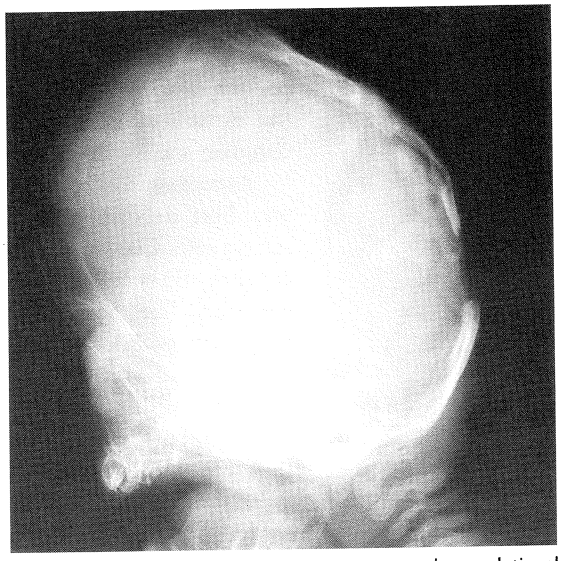

Fig 2 - Lateral skull X-ray showing exaggerated convolutional markings, vault deformity and absence of sutural markings.

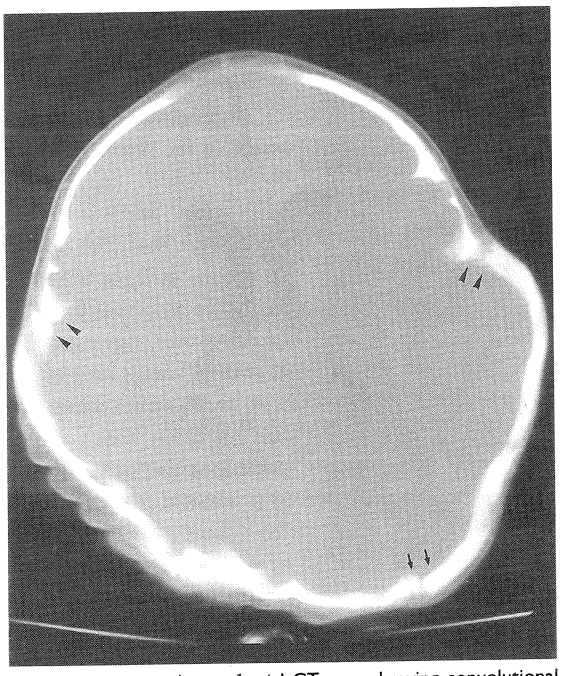

Fig 3 - Bone windows of axial CT scan showing convolutional markings and fused right lambdoid suture. The unfused left lambdoid suture (arrows) and coronal sutures (arrowheads) are seen.

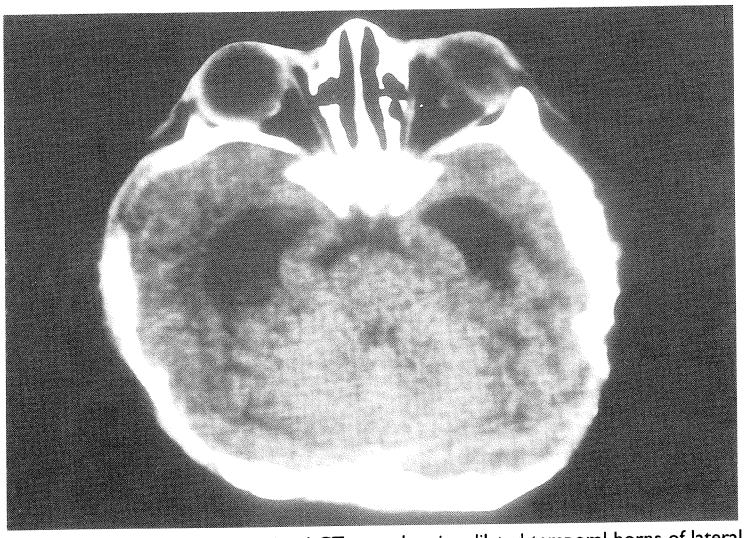

Fig 4 - Axial CT scan showing dilated temporal horns of lateral ventricles.

The modified prone position was chosen for optimum access to both frontal and occipital regions (Fig 5). A linear biparietal incision was made and scalp dissection was performed in the supraperiosteal plane to minimise blood loss. The metopic suture was noted to be bulging, with evidence of hyperostotic bone at both lambdoid sutures, particularly the left, and posterior half of the sagittal suture. The tight prematurely fused sutures posteriorly caused compensatory bulging through the anterior fontanelle, patent metopic suture and squamous temporal bones bilaterally, thus resulting in a cloverleaf-like skull deformity. In addition, noticeable bone softening from raised intracranial pressure was seen at various points.

Bifrontal, biparietal and occipital craniotomies were performed for decompression and separate frontal, parietal and occipital bone grafts were elevated on each half of the skull, taking particular care to avoid blood loss from injury to the sagittal sinus. The areas of hyperostotic bone were smoothened and thinned down using the burr-attachment of the high-speed craniotome. Individual bone grafts were remodeled by a combination of radial osteotomies and controlled fractures using a small bone rongeur. Once satisfactory decompression from the various constricting points was achieved and the brain was free to expand uniformly, no dural plication for contouring nor bone work to reduce the height of the skull vault was required. The overall shape and appearance improved significantly. The separate bone grafts were then replaced using 3-0 absorbable sutures and fibrin glue.

At the end of surgery, the cloverleaf skull configuration had been restored to a more acceptable cosmetic appearance, and the post-operative course was uneventful. The corneal abrasions healed with no subsequent visual impairment. The baby continued to gain weight and at six months of age, was able to sit with support. His body weight was 7.8 kg (75th percentile). He was noticed to have increased anterior fontanelle tension at seven months of age, although there was no deterioration in clinical signs. A CT scan and MRI of the brain showed ventriculomegaly as a result of aqueduct stenosis. This appeared secondary to a small posterior fossa with limited space for hindbrain expansion (Fig 6).

A ventriculo-peritoneal shunt was inserted, followed by fronto-orbital advancement two months later. At one year of age, he was able to stand with minimal support, play and feed well, and was beginning to vocalise. The cosmesis was acceptable (Fig 7), with full eye closure achieved.

DISCUSSION

Craniosynostosis has been described in as many as 62 syndromes, associated with single gene disorders, chromosomal disorders, environmental teratogens and sporadic events[3]. The cloverleaf skull deformity is characterised by a tower-like bulging of the cranial vault and marked outpouching bilaterally at the temporal areas, with downward displacement of the

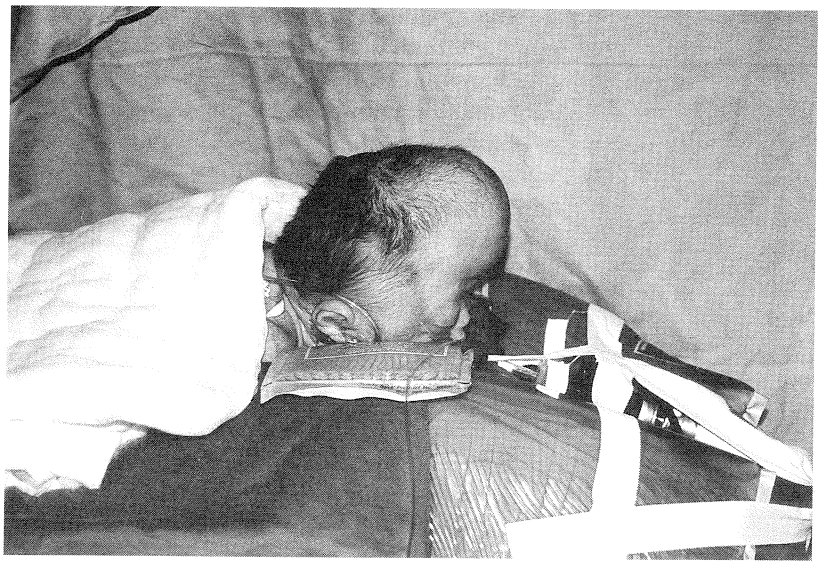

Fig 5 - Modified prone position for surgery.

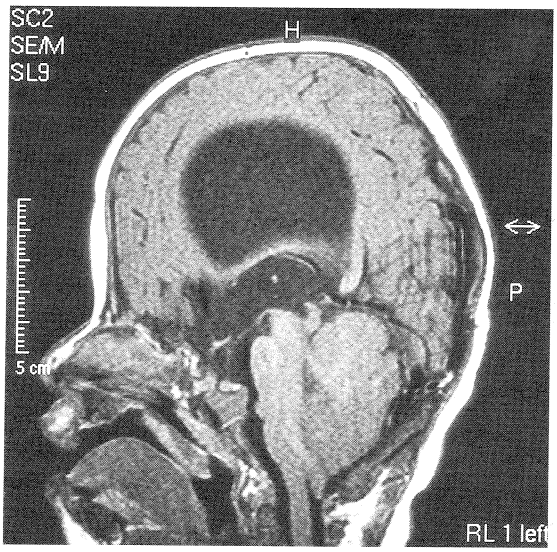

Fig 6 - Sagittal MRI showing a small, distorted posterior fossa with mild tonsillar herniation.

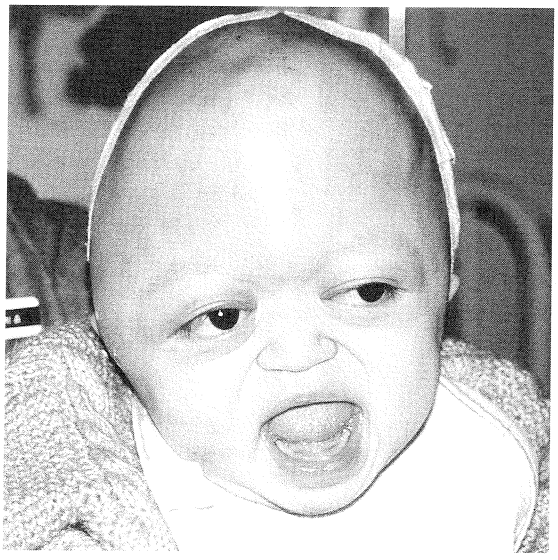

Fig 7 - At 10 months of age, after fronto-orbital advancement.

ears to an almost horizontal position, low nasal root, and proptosis[4]. It may occur as an isolated phenomenon or as part of several clinical syndromes, the most well-known of which are the Apert, Crouzon and Pfeiffer syndromes[4].

According to Cohen, Pfeiffer syndrome, which was first described in 1964, consists of three subtypes[2]. Classic Pfeiffer syndrome or type 1 consists of brachycephaly, syndactyly, broad thumbs and toes. The mode of inheritance is autosomal dominant with complete penetrance in familial pedigrees, of which 7 have been reported. Type 2 is associated with cloverleaf skull, Pfeiffer hands and feet, and ankylosis of the elbows, whereas type 3 is similar to type 2 but without the cloverleaf skull. Type 2 and 3 occur sporadically, with poorer prognosis, more severe neurological deficits and shorter survival[2]. This particular case is of interest because of features consistent with type 2 Pfeiffer syndrome (sporadic, no family history, cloverleaf skull and broad thumbs and toes), yet without the usual poor prognostic factors, such as seizures and mental retardation.

Frequently in cases of cloverleaf skull, there are coexistent complex abnormalities such as hydrocephalus, intracranial hypertension, hindbrain herniation, and skull base dysplasia[5]. Shiroyama suggested that the hydrocephalus may be either communicating or obstructive, and that the aetiology might be associated with infratentorial and basilar compression, aqueduct stenosis or impairment of venous drainage and cerebrospinal fluid flow at the level of the constricting sutural band[6]. Arnold-Chiari malformation, acquired on the basis of cephalocranial disproportion in such cases, has also been reported as a possible cause of hydrocephalus[7].

Opinion however is divided as to the management of the hydrocephalus. Golabi described 10 cases of craniosynostosis with hydrocephalus, of which 5 had cloverleaf skulls[8]. All 10 cases underwent a shunting procedure prior to definitive craniotomy. Jane also advocated a similar practice[9]. There are however others who believe that hydrocephalus is not an invariable complication present at the time of birth, but rather secondary to the obliteration of the cortical subarachnoid spaces and the presence of a cranial ring[10]. As such, total craniectomy may reverse the hydrocephalus satisfactorily.

In this particular case, fear of possible visual impairment from corneal ulceration and an alarming progression of skull deformity and anterior fontanelle tension prompted early surgery, primarily as a life-saving procedure to reduce raised intracranial pressure. Satisfactory decompression was achieved in addition to a more acceptable skull appearance, and shunting was subsequently avoided for six months. However, these cases are often associated with small posterior fossae and hindbrain abnormalities[5], resulting in hydrocephalus, for which a shunt had to be performed eventually. The craniostenotic state which has been

known to recur despite cranial vault surgery and is probably accelerated by shunting[5], has not recurred in our patient at one year of age. Early surgery may have played a not insignificant part in this respect. Having performed a near total craniectomy for him, we suggest that mild hydrocephalus may have been beneficial in allowing skull shaping during the period of rapid brain growth (the first six months in this case) and avoiding early restenosis.

CONCLUSION

In complex craniostenosis, raised intracranial pressure in the neonatal period is of primary concern. We recommend early surgery in preference to a temporising shunt procedure. This can be safely performed if there are satisfactory paediatric anaesthetic and intensive care facilities. Further elective cosmetic procedures can be performed when the child is older. The cloverleaf skull continues to present a particular challenge, not only in its rarity but also in its surgical management.

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