CRANIOSYNOSTOSIS : MANAGEMENT IN INFANCY

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Introduction

Craniosynostosis is defined as premature fusion of one or more cranial sutures, which may occur in isolation or in association with a syndromic constellation, resulting in cranial deformity. Functionally, craniosynostosis may be defined as the premature conversion of dynamic region of growth and resorption between two adjacent bones of the cranium into a static region of bony union. In the treatment of such cases, each case must be judged individually and a multidisciplinary approach is recommended. We present a case of microcephaly with bilateral coronal craniosynostosis treated with early wide linear craniectomy followed by relative normalization of skull and facial appearance.

Case Report

This six month old first-born male baby was brought to paediatric surgery department with complaints of delayed developmental milestones and small size of head since birth. His anthropometric parameters revealed a length of 59 cm (5th centile), a weight of 4.5 Kg (5th centile) and OFC 30 cm (5th centile). Clinically the baby had microcephaly with closed fontanelle, fused coronal and metopic sutures, flattened forehead on sides with central prominence, and bilateral proptosis (Fig 1). Prominent ears and a high arched palate were also noticed. The child was mentally deficient but there was no focal neurological deficit. Fundus examination was normal. X-ray skull confirmed obliteration of coronal and metopic sutures (Fig. 2). CT scan brain showed microcephaly with fusion of metopic and coronal sutures suggestive of craniosynostosis (Fig 3). There was no evidence of generalized hydrocephalus. The case was taken up for surgical correction after joint consultation with neurosurgeon on 24 Jan 2000.

After induction, the child was placed in a supine position. Sterile preparation of the head was done and a bicoronal skin incision was made from one antitragus to the other. The scalp flap was elevated by blunt and sharp dissection of the galea from the periosteum anteriorly upto the supraorbital rim. Burr holes were made along the lateral aspect of anterior fontanelle and a 1.5 cm wide strip of coronal suture removed with the help of bone

rongeur and continued laterally to the sphenozygomatic sutures. Similarly, a wide synostectomy was carried out along the metopic suture down to the nasion. Finally a linear craniectomy, running about 1 cm above and parallel to the orbital rim was carried out to join the coronal and metopic craniectomies to obtain bilateral free frontal bone flaps. These flaps were removed and orbital roof decompression was carried out. Now a mosaic of small postage stamp bone fragments obtained from frontal bone flaps were laid freely above the duramater (Fig 4). Complete haemostasis was achieved by bipolar coagulation and local absorbable gelatin sponge (Gelfoam) application. The scalp was resutured back in the bicoronal plane after inserting a closed drainage system (Redivac). This drainage tube was removed on 5th post-operative day. Postoperative recovery of the patient was uneventful and there was

Fig. 1 : Clinical photograph of the baby showing flattened forehead with central prominence and bilateral proptosis

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Fig. 2 : X-ray skull showing obliteration of coronal and metopic sutures

Figs. 3a & b : Two different sections of CT brain showing fusion of metopic and coronal sutures.

dramatic improvement in the cranial shape and facial appearance over a period of 6-8 weeks (Fig 5).

Fig. 4 : Operative photograph showing mosaic of bone fragments obtained from bone flaps laid on durameter.

Fig. 5 : Post-operative clinical photograph after 8 weeks of surgery showing marked improvement in shape and facial appearance

Discussion

Craniosynostosis is the premature fusion of one or more cranial sutures with secondary changes in shape or volume of the skull or both. The term fusion implies partial or total ossification of the strips of connective tissue interposed between adjacent vault bones. Abnormal fusion of cranial sutures interferes with their main function of allowing its enlargement to accommodate for the physiologic growth of the neural mass [1]. The biology underlying this fusion remains poorly understood. A number of craniosynostotic disorders have recently been ascribed to mutations in one of the three fibroblast growth factor receptor genes (FGFR1, FGFR2, and FGFR3). Common feature of these FGFR-associated conditions is unilateral or bilateral premature ossification of the coronal suture. Thus, from both diagnostic and prognostic points of

view, early FGFR mutational screening in craniosynostotic conditions will be helpful [2]. Sagittal suture is the commonest affected in craniosynostosis; with a striking preponderance of males while coronal synostosis has predilection towards females. Rarely, all sutures are involved. The rate of prevalence of craniosynostosis is 0.04% to 0.1% [3]. The main clinical feature is skull deformity but sometimes mental retardation, ocular problems and associated congenital defects may co-exist. Skull deformity depends on the suture or sutures involved. The commonest condition, sagittal craniosynostosis is associated with a long, narrow skull recognizable at or soon after birth. Bilateral coronal craniosynostosis gives rise to a head which is broad in its lateral dimensions and narrow antero-posteriorly with shallow orbits and usually some proptosis, as seen in our case. Coronal suture is found involved in 13% of cases [4]. The rare combination of coronal and metopic synostosis produces an appearance called turribrachycephaly.

Evaluation of these patients should include a history, physical examination and developmental analysis. A good quality skull x-ray is essential in deciding which sutures are affected. Hyperostotic bony lesion and obliteration of the suture characterize the lesion. High resolution three dimensional CT reconstruction scanning is the radiological procedure of choice for determining the extent of premature bony fusion [5].

Various approaches to surgical correction of deformities are described [6,7,8,9]. There is still a controversy regarding the optimum time to perform surgery for craniosynostosis. Some recommend surgery soon after birth and others delay it until the age of 12 months. Sgouros S et al recommend early surgical correction to prevent neurologic sequelae and to provide the best cosmetic result [10]. He also suggests that measurement of intracranial volume could be used to "fine tune" the optimum time for surgery [11].

The principle of treatment is to allow initial expansion of the dura and its enclosed neural tissue and create an artificial suture by a linear craniectomy in the region of the prematurely fused suture, removing a strip of bone and also the pericranium for 2 or 3 cm beyond the edge of the gap in the bone, as was done in our case. The goals of surgery are to ensure normal brain growth, to prevent increased intracranial tension, to prevent compromise of visual and auditory function

and to improve skull and facial appearance [12]. Several surgical procedures are propounded to correct coronal synostosis but most surgeons prefer to leave a mosaic of bone to be possibly moulded by an external device [13] or to use various bone transposition techniques [14].

To conclude simple strip craniectomies are sufficient to obtain satisfactory functional and cosmetic results in patients of less severe forms of bicoronal craniosynostosis, who are operated in early infancy.

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