CASE REPORT

CRANIOSYNOSTOSIS: A CASE REPORT

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ABSTRACT

Background: Craniosynostosis has a prevalence of 1 in 2000 to 2500 live births. We report on a case of craniosynostosis managed surgically.

Method: A 2-year old male, case of craniosynostosis which presented to Ziauddin Hospital North Nazimabad OPD. This child had a history of multiple falls since the past 4 months for which sutures were required twice. His head circumference was above 95 percentile for his age. He was diagnosed as a case of trigonocephaly based on the findings of the 3D CT scan. He underwent surgical remodelling

Result: Patient was discharged after an unremarkable post-operative period.

Key words: Craniosynostosis, case report, remodeling

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INTRODUCTION

Craniosynostosis is said to be when there is early fusion of the cranial plates or sutures[11,12]. The human brain grows fast early in the first year of life meanwhile the sutures of the major cranial bones are forming new bone in relationship to the rapid growing brain. If any of the plates unite or fuse early, this is known ascraniosynostosis. [11]Craniosynostisishas a prevalence of 1 in 2000 to 2500 live births. [2,12] In 85% of the cases only one suture is involved and is nonsyndromic while multiple sutures are involved in syndromic cases along with other clinical abnormalities involving the hand, feet, cardiac and skeletal[8]. The syndromes associated with this condition include Apert and Pfeiffer syndrome.[A]

This abnormal or early fusion is genetically transmitted in 10-14% of the cases [12]. The etiology regarding craniosynostosis is very complicated and many genes are involved but the mutations in signaling pathways are significant, involving the craniofacial morphogenesis like the Hedgehog pathway and Notch signaling pathway both of which have an important role in early craniofacial development. [2,11]

Craniosynostosis alters the cranial vault frame significantly leading to restricted cranial growth and increase in intracranial pressure which can lead to optic nerve compression, optic atrophy, blindness and cognitive disability. [10,2] In the cases of visual compromise, urgent intervention is required to ensure that there is no permanent loss of vision. [A]

CASE REPORT

History

A 2-year old boy,weighing 14 kg came to the OPD with complaints of laceration of about 2×3 cm on the forehead approximately 1 cm above the glabella through which bone was visible. This child had a history of multiple falls since the past 4 months for which sutures were required twice. His head circumference was above 95 percentile for his age. There were no apparent facial deformities and age appropriate developmental milestones were achieved. His birth history included an uneventful spontaneous vaginal deliveryat 38 weeks, his natal weight was 3 kg and there was no NICU admission. The patients parents had a consanguineous marriage but both of his parents deny any history of

congenital defects like spina bifida or craniosynostosis in their family. This patient also has a 2 months old younger brother who is healthy according to parents. The child blood work, behavioral and nutritional history were unremarkable.

CT findings

The patient underwent thin slice high resolution cranial CT examination using multi-slice CT scanning and reconstruction images were obtained for 3 dimensional analysis. The scan revealed premature fusion of metopic suture with triangular and pointed shape of forehead due to premature fusion of frontal bones. There was no evidence of hemorrhage, infarction or space occupying lesion in the brain. He was diagnosed as a case of trigonocephaly based on the findings of the 3D CT scan.

Expansive craniotomy associated with skull reconstruction was planned.Bicoronal flap was raised, but due to extent of maturation of the frontal sinuses which greatly increased the risk of hemorrhage, we instead opted to remodel the patient's skull using a drill. The ossification centres associated with frontal hyperostosis were removed. The hypertrophied bone matrix was shaved until the bone was thin enough to visualize the underlying dural color. Scalp wounds were closed, the laceration was repaired with inside sutures and radivac drain was placed. Postoperative course was uneventful and the patient was subsequently discharged.

DISCUSSION

In our case the risk factors for craniosynostosis identified were consanguineous marriage and nulliparity.[1,2,3] There was no cognitive impairment in our patient; which is more commonly seen in craniosynostosis associated with syndromes.[4] Our patient also had frontal hyperostosis which is more commonly associated with nonsyndromic craniosynostosis.[5] As there is no positive family history of craniosynostosis in this patient it is reasonable to conclude that our case is not associated with any syndromes. However, it is important to note that after Caucasians, craniosynostosis is most commonly seen in Pakistanis. This may be attributed to multiple of reasons including consanguinity.[6]

Metopic craniosynostosis are associated with cognitive impairments, however a detailed history of our patient did not reveal any delays or regression of developmental milestones. [10]

Severe craniosynostosis when involving multiple sutures becomes a medical emergency which requires airway protection, nutritional support and management of ICP. A multidisciplinary approach may be required involving paediatricians, ENT specialists and ophthalmologists as well as a neurosurgeon. Emergency management may include tracheostomy, calvarialexpansion,foramen magnum decompression andVP shunts in cases of obstructive hydrocephalus. [A]

60% of craniosynostosis cases are operated on between 6 months to 2 years of age and our patient falls in this range.Preoperative workup includes CT scan of the head with 3-D reconstruction, as well as a venogram when a hazardous surgery is forecasted due to abnormal drainage. In thiscase, however, only the former was required.

In cases of trigonocephalywhich is the case in our patient, simple suturectomy which can be done in other varieties of craniosynostosis is considered insufficient. Extensive reconstruction is required which can be done via fracturing the frontal bone and using the subsequent bone as a rotational graft. In our patient due to the extent of the spread of the frontal sinuses we instead opted to do remodelling with a bone file and a drill. We were able to achieve a satisfactory cosmetic outcome with this method.[A,B]

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