Isolated primary craniosynostosis in an adult: Imaging findings of a case

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Abstract

Craniosynostosis means premature closure of calvarial sutures. It may be primary or secondary. The patient presents with unexplained neuropsychological impairment and radiological imaging clinches the diagnosis. We present a case of 31-year-old female having primary isolated craniosynostosis who survived into adulthood without any surgical intervention. The imaging findings of such a case are rarely described in the literature.

Key Words

Craniosynostosis, convolutional, imaging, sutures, syndrome

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Ann Indian Acad Neurol 2014;17:92-4

Introduction

Craniosynostosis refers to closure of calvarial sutures prematurely resulting in restricted skull growth. It is classified as primary and secondary. The patient presents with unexplained neuropsychological impairment. Radiological imaging is necessary for establishing the diagnosis.

Case Report

A 31-year-old female was brought with a history of severe mental retardation since birth. She had a history of global developmental delay also. Both the parents were normal with no consanguinity. On physical examination, she had a peculiar face with hypertelorism and deformed calvarium. There was no mandibular hypoplasia or midfacial deformity. No microcephaly or abnormal nasal configuration was seen [Figure 1]. Her hands and feet were normal with no syndactyly. X-ray of the skull [Figure 2a and b] showed prominent convolutional markings on the calvarium with complete closure of all calvarial sutures. No harlequin eyes were seen. Plain computed tomography (CT) confirmed the X-ray findings. All the sutures were closed and showed bone bridging and heaping. Inner table of skull showed extensive convolutional markings [Figure 3a and b]. The brain parenchyma was normal with no atrophy. Corpus callosum, both leaves of the septum pellucidum, and posterior fossa structures were well seen. However, the

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DOI: 10.4103/0972-2327.128563

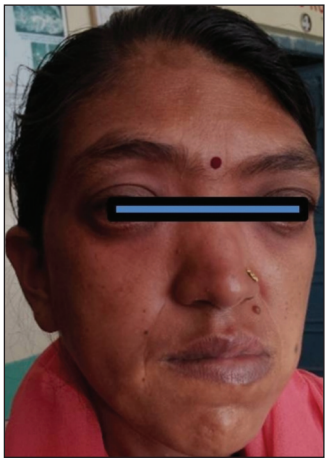

Figure 1: Photograph of the patient shows hypertelorism and deformed head (photograph taken with prior permission of the patient's attendant for online and in-print publication)

subarachnoid space and cisterns showed effacement. The ventricular system was normal [Figure 4]. The paranasal sinuses were hypoplastic. All these imaging findings along with the supporting history were consistent with primary isolated craniosynostosis.

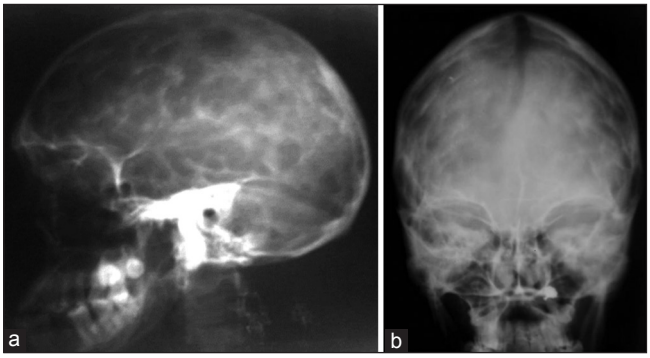

Figure 2: X-ray of the skull lateral (a) and frontal (b) view shows prominent convolutional markings and closure of cranial sutures. The frontal paranasal sinuses are hypoplastic

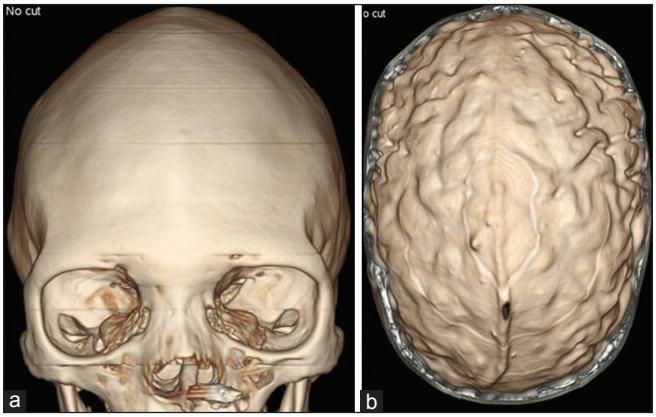

Figure 3: Volume-rendered computed tomography (CT) image (a) shows deformed skull with sutural closure and bone bridging and heaping. Axial cut (b) shows prominent convolutional markings on the inner table

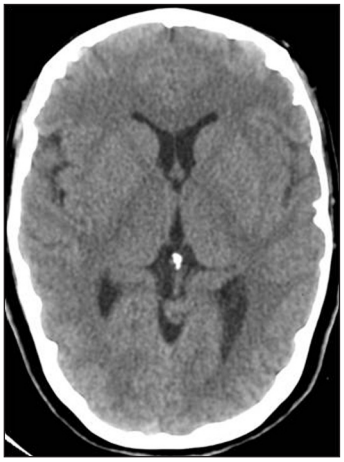

Figure 4: Plain computed tomography (CT) axial image at the level of frontal horns shows prominent convolutional markings on the inner table of skull. There is effacement of subarachnoid space. The brain parenchyma is normal

The uniqueness of our case lies in the presentation of imaging findings of the patient with primary craniosynostosis who survived into adulthood without undergoing any corrective surgical procedure, whereas the literature describes such findings in infants and children only.

Discussion

Craniosynostosis means premature closure of calvarial sutures. The term was first coined by Virchow in 1851.[1] The prevalence rate is 3-5/10,000 live-born infants. The affected child has neuropsychological impairment and a lower intelligence quotient. The possible explanation is raised intracranial pressure and decreased intracranial volume owing to the restricted skull growth in these patients.[2]

Craniosynostosis can be classified[3] as primary, which can be in isolated form or may be a part of a syndrome and secondary, which is associated with many metabolic and hematological disorders, bone dysplasias and dysostoses and ventriculovenous shunts.[4]

Isolated primary craniosynostosis is a rare congenital disorder.[5] In general, sagittal suture closure is most common and is seen in 50% of the cases.[1] Primary craniosynostosis is subclassified in accordance to the type of suture involved as tabulated [Table 1].

However, in syndromic craniosynostosis, synostosis of bilateral coronal sutures is the most common;[6] 40% of the primary craniosynostosis are syndromic.[2] The common syndromes associated with craniosynostosis are Crouzon syndrome, Apert syndrome, Carpenter syndrome, Treacher Collins syndrome, and craniotelencephalic dysplasia.[4]

The primary radiological features of craniosynostosis are narrowing and indistinctness of suture with bony bridging along it. There may be heaping of bone at the suture. The secondary signs include defective calvarial shape, changes in shape and timing of fontanelle closure, and facial anomalies.[1] There may be effacement of the subarachnoid space underneath the prematurely closed sutures. The subarachnoid space beneath the part of skull showing compensatory growth may be prominent requiring dural plication. There may be fibrous union of the suture with no primary radiological signs. Even then, the presence of marked secondary signs only warrants surgical correction.

Table 1: Classification of primary craniosynostosis[1]

Type	Sutures involved
Simple synostosis	
Brachycephaly	Coronal and/or lambdoidal
Dolichocephaly/scaphocephaly	Sagittal
Trigonocephaly	Metopic
Pachycephaly	Lambdoidal
Plagiocephaly	Unilateral coronal or lambdoidal
Compound synostosis	
Acrocephaly/oxycephaly/ turricephaly	All sutures or coronals plus one other
Kleeblattschadel (cloverleaf skull deformity)	Multiple or all

Radiological evaluation is necessary to characterize the calvarial deformity. It yields information about the type and extent of suture involved, which guides the corrective surgical procedure. The ideal time for the surgery is the first year of life for better patient outcome.[2] CT better defines the skull changes and also evaluates associated intracranial and facial abnormalities. Other accompanying findings of orbit, paranasal sinuses, and nasal skew should also be assessed.

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How to cite this article: Thakur S, Jhobta A, Kumar S, Thakur CS. Isolated primary craniosynostosis in an adult: Imaging findings of a case. Ann Indian Acad Neurol 2014;17:92-4.
Received: 20-08-13, **Revised:** 25-09-13, **Accepted:** 25-09-13
Source of Support: Nil, **Conflict of Interest:** Nil

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