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Case Report

Trigonocephaly pre- and post-operative evaluation by multidetector computed tomography

Rahul Talwade[1], Sabyasachi Basu[2], E Adarsh[3], Anil Kumar Shukla[1], K R Shankar[1], Dibanath Chakrabarty[2]

From Departments of [1]Radiodiagnosis, [2]Neurosurgery, [3]Paediatrics, Raja Rajeswari Medical College and Hospital, Bengaluru, Karnataka, India

Correspondence to: Dr. Anil Kumar Shukla, Department of Radiodiagnosis, Raja Rajeswari Medical College and Hospital, Bengaluru - 560 074, Karnataka, India. Phone: +91-9342508923. E-mail: shookla2007@yahoo.co.in

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ABSTRACT

Trigonocephaly is the third most common single suture synostosis which is seen as bulging of the forehead due to early fusion of the metopic suture. It is linked with an increased level of neurodevelopmental delays. On imaging, multidetector computed tomography shows anterior fontanelle ossification, hypoteliorism, narrowing of the anterior cranial fossa, and compensatory increase of the middle cranial fossa with atrophic features of brain. Its reported incidence is 1 in 700-15,000 live births. Operative multiple osteotomies were done and imaged in follow-up.

Key words: *Craniosynostosis, Hypoteliorism, Metopic synostosis, Multi detector computed tomography, Trigonocephaly*

Premature closure of the metopic suture results in a growth restriction of the frontal bones, which leads to a skull malformation known as trigonocephaly (metopic synostosis). Over the course of recent decades, its incidence has been rising, currently making it the third most common type of craniosynostosis. Treatment consists of a cranioplasty, usually performed before the age of 1 year. It is linked with an increased level of neurodevelopmental delays. The imaging findings range from a reduced volume of the anterior cranial fossa to intrinsic malformations of the brain [1]. This case report aims to provide an overview and roadmap of this entity by pre- and post-operative multidetector computed tomography (MDCT) which also plays a vital role in follow-ups.

CASE REPORT

A 3-month-old female baby was brought to hospital in view of bony midline frontal ridge noted by her mother. There was no history of seizures or delayed milestones. Clinical inspection showed triangular shape of the forehead and midline frontal ridge. The ridging metopic suture could be palpated and temporal narrowing was noted (Fig. 1a and b). Swelling was non-compressible and no signs of inflammation were noted. Central nervous system examination revealed normal reflexes, and there was no neurological deficit. The patient was referred to the department of radiology for MDCT. Written informed consent was obtained from the father of the child, and multidetected computed tomography was performed.

We used a 128 slice Siemens perspective CT machine and images obtained at sub mm sections in the axial, coronal, and sagittal planes in both bone and soft tissue windows. Post-processing techniques such as shaded surface displays and volume rendering technique (VRT) were used. In bone window, there was complete closure of the metopic suture (Fig. 1c). Axial sections showed triangular anterior fossa and triangular frontal lobes of brain with minimal atrophy and prominent adjacent sulcal spaces (Fig. 1d). Ratio of interparietal and intercoronal distance (ICD) is increased (Fig. 2a). Frontal angle was decreased to 97° (Fig. 2b). Orbits show decreased interorbital distance suggestive of hypoteliorism (Fig. 2c and d). VRT images showing prominent gyral impressions on calvaria in bilateral parieto-occipital regions (Fig. 3a-d).

Child underwent decompressive craniotomies with sutural release involving frontal bone and was stable post procedure. Follow-up CT scan was done 1 month after surgical correction and it appearing as "floating forehead" (Fig. 4a and b). Few overlapping bony fragments noted with adequate distance between other fragments (Fig. 4). However, orbits did not show significant changes compared to pre-operative images. The cosmetic appearance was improved, and no neurological deficit was noted on follow-up.

DISCUSSION

Craniosynostosis or craniostenosis is the premature fusion of cranial sutures. It can be isolated or associated with craniofacial syndrome with alteration of shape of the cranial vault. Broad categories include "simple craniosynostosis," involving only one suture, or "compound craniosynostosis," where two or more sutures are involved [2]. By etiology, it is primary - due to intrinsic defect in suture or secondary - due to premature closure of normal sutures and deficient brain growth [3]. Trigonocephaly is derived

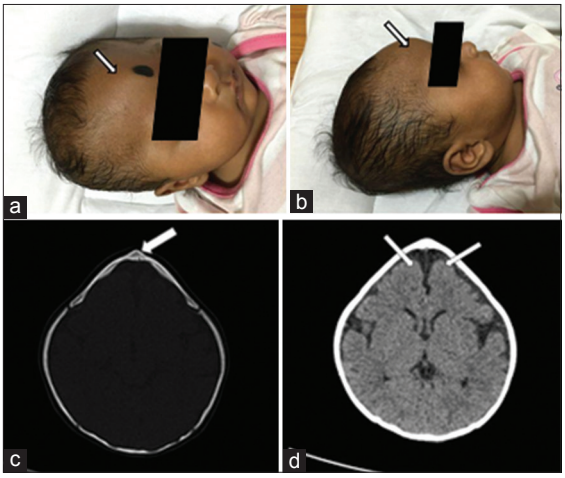

Figure 1: (a and b) Photographs of the baby showing midline frontal ridge (arrows). Multidetector computed tomography showing (c) complete closure of metopic suture (arrow) and (d) frontal lobe minimal atrophy

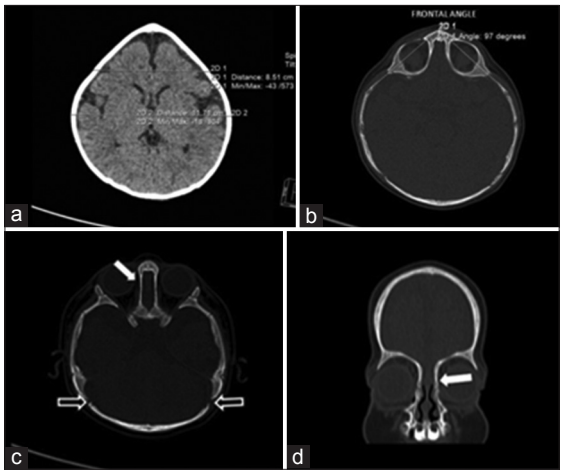

Figure 2: Multidetector computed tomography showing (a) increased ratio of interparietal and intercoronal distance (b) showing decreased frontal angle (c and d) decreased interorbital distance

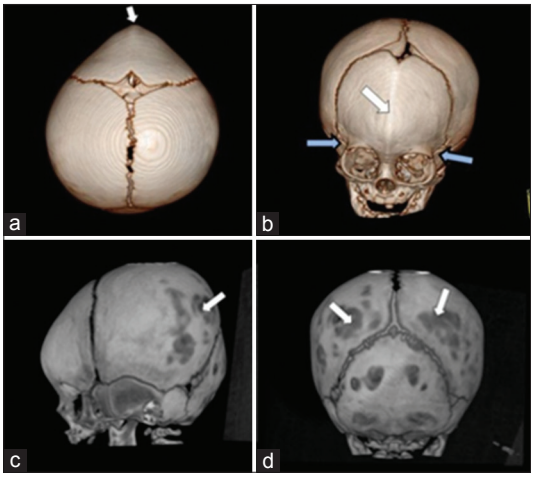

Figure 3: Multidetector computed tomography volume rendering technique images showing (a and b) midline frontal eminence (white arrow) and bitemporal indentations (blue arrows) (c and d) gyral impressions on calvarial and bilateral parieto-occipital regions (arrows)

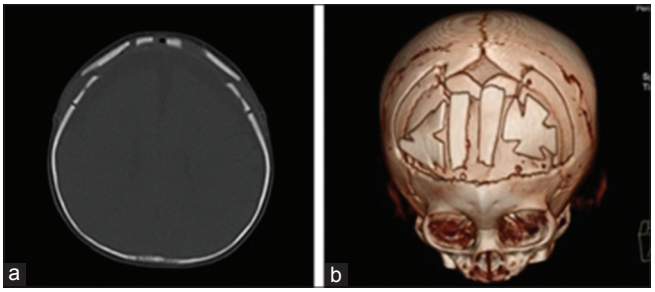

Figure 4: (a and b) Post-operative multidetector computed tomography volume rendering technique and bone window images showing multiple osteotomies appearing as "floating forehead"

from the Greek word "trigon on," which means triangle, and "kephale," which means head. Forehead is triangular, or wedge-shaped due to premature fusion and subsequent ossification of the metopic suture (Greek "metopon" = forehead). The term trigonocephaly was first proposed by Walker in 1862 [1].

The metopic suture separates the two frontal bones at birth and is the first skull suture to close physiologically, starting at 3 months and completely fused at the age of 8 months [4]. Premature fusion shows a ridge over the midline of the forehead due to ossification of the suture and lateral growth restriction of the frontal bones. According to the theory of Virchow, this wedge shape is even further enhanced by the increased compensatory growth of the remaining skull sutures while the skull keeps expanding [1]. This can occur from *in utero* fusion, and one-third of the cases are syndrome and may be associated with other midline anomalies involving the brain or palate [5].

Due to MDCT, many imaging features are observed in our patient such as a triangular, pointed forehead with flattened frontal bones, and bossing of the parieto-occipital regions. Our patient had hypoteliorism, narrow anterior cranial fossa, hypoplasia of the ethmoid sinuses and "quizzical" orbits. Inferior angle of the anterior fontanel is elongated with frontal notching and tight anterior extraaxial subarachnoid spaces. Temporal shortening and parietal widening with normal cephalic index were also observed [6].

Synostosis of this kind is the third most common type followed by unicornal and sagittal and has an incidence of 1:700 to 1:15,000 of newborns, having male to female ratio of 3:1 [1]. On clinical evaluation, there is reduction in skull size, lengthening, and ridging of the skull along the fused suture. Due to stenosed suture, there is growth restriction with compensatory more of growth at the other open sutures sites. By fetal ultrasonography, it can also be identified after 9 week of gestation [7]. Due to mainly bony changes submillimeter slices by MDCT is very accurate, and it also shows features of brain growth, deformities and ventricular volumes. MDCT is very useful in planning of surgical correction and subsequent post-operative follow-ups [8].

By MDCT supraorbital retrusion, classical feature of trigonocephaly can be classified as - Frontal angle: It is the angle between the two lines drawn through pterion (bilaterally) and nation. Trigonocephaly is classified as: (1) Severe with an angle of <89°, (2) moderate between 90°and 95°, (3) mild between 96° and 103°, our case was in this category as angle is 97°, and

(4) normal when 104° or more (Fig. 2b) [1]. Frontal stenosis, It is defined as the increased ratio of the interparietal distance (IPD) to the ICD. The IPD/ICD to be 1.21 in normal children in our case it is increased 1.37 with small anterior fossa (Fig. 2a) [1].

Any craniosynostosis leads to high incidence of poor scholastic and mental development. Diagnosed patient should be operated as soon as possible as it has very good prognosis and it also improves the cosmetic appearance and developmental milestones. For surgical treatment, timing and extent are directed by the severity of the frontal changes with normalization of the forehead with reconstitution of the normal supraorbital rim if necessary. Individuals presenting solely with a prominent midline keel may be best served by simple contouring of the frontal bone or by removal of the frontal bone flap followed by reconfiguration. Patients with significant trigonocephaly and hypertelorism need orbital reconstruction and may also require lateral expansion of the orbits at the same time. The type and timing of the correction must be tailored to the individual patient, and long-term results are excellent with minimal complications [9]. In our case, decompressive craniotomies with sutural release were done and are seen by post-operative follow-up MDCT as "floating forehead" (Fig. 4a and b).

CONCLUSION

MDCT is the imaging modality of choice in trigonocephaly in the initial pre-operative stage to take various measurements and in post-operative status to assess osteotomies and to assess bony and cerebral components in the further follow-ups. Regular pediatric clinical, neurological and whenever required MDCT follow-ups are recommended in these cases. Our patient had no underlying neurological deformity; however, in later follow-ups, we will assess improvement in the cognition.

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