Craniosynostosis Associated with Lacunar Skull: Three-Dimensional Computed Tomography Features

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ABSTRACT

Craniosynostosis is the premature closure of the cranial sutures. We present the radiological findings of a six-month old case who presented with small head and frontal triangular shape and who was found to have generalized pitting and softness on palpation of the head. The three-dimensional images computed tomography (CT) showed a sagittal, coronal and metopic suture synostosis with exaggerated convolutional markings in the form of lacunar skull. This case report showed that patients with craniosynostosis should be evaluated by three-dimensional images CT.

***Key words**: Craniosynostosis, Three-dimensional computed tomography, Lacunar skull*

INTRODUCTION

Craniosynostosis is the premature fusion of one or more of the cranial sutures and can occur as part of a syndrome or as an isolated defect (1). The prevalence of craniosynostosis was estimated to be one per 1,800 to 2,200 births (2). The process of growing inhibition due to craniosynostosis is not always restricted to the neurocranium, it may have an influence on the development of the viscerocranium too. In this article, the importance of three-dimensional CT in the evaluation of patients with craniosynostosis was discussed by presenting a 6-month-old male patient.

CASE

A 6-month-old male patient with narrow, triangular forehead and small cranium was referred to our pediatric clinic for microcephaly. The breast-feeding patient was delivered by cesarean section after a normal prenatal period. There was collateral consanguinity between the parents and one of the siblings older than him had craniosynostosis. On physical examination, both anterior and posterior fontanels were

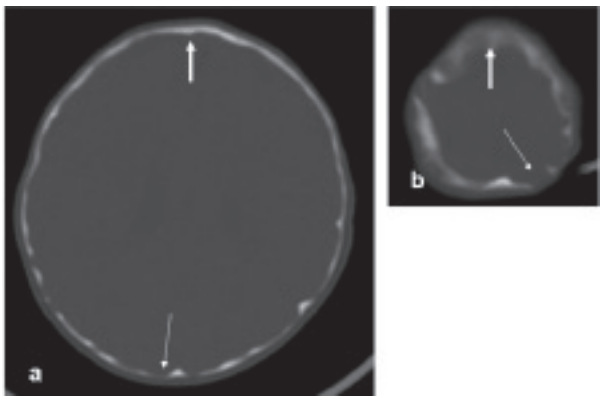

***Figure 1**. Routine cranial examination; transverse scans; Multiple thinning and defective areas with convexities toward external direction in calvarium, dominantly located in the parietooccipital region (a, b thin arrows) and also moderate sharpness in the frontal region (a, b thick arrows); Coronal and also the lambdoid sutures are not seen*

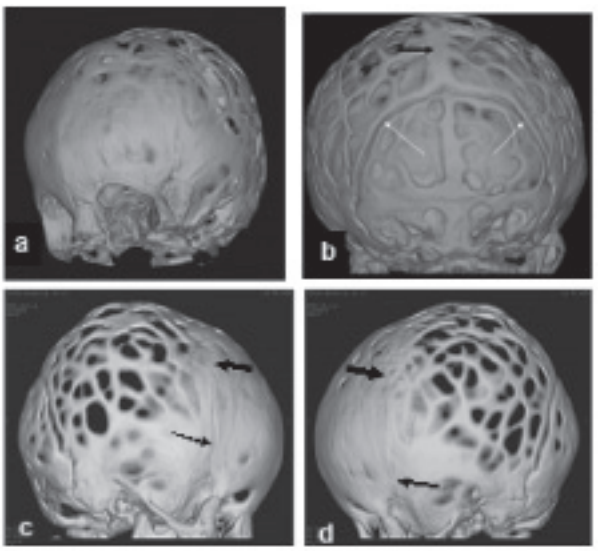

Figure 2. (a), Three-dimension CT reformatted image, left anterior oblique projection: Fusion and slight frontal triangular formation (arrow) on metopic suture except its short caudal segment (star) (b), Lambdoid sutures are open (white arrows); sagittal sutur is closed (black arrows) (c), (d), Fusion at coronal sutures bilaterally, partial in the caudal (thin arrow) and complete in the superior (thick arrow) region. Excessive exaggerated increased convolutional signs in the cranial bones more prominent in the preoccipital region (Hemi cranium is cut and removed in order to prevent delusion because of superposition).

closed, but there was generalized softness on palpation of the head. The patient was below 3 percentile for weight, length and head circumference, the measurements being 5200 g, 56 cm and 36,5 cm respectively. Both the other physical examination and the laboratory findings such as biochemistry, complete blood count and also TORCH panel were unremarkable. Cranial CT showed that both the posterior fossa and the supratentorial region was normal. Whereas the cranial vault had multiple thinned areas, convexities of which were facing outwards and focal defects, especially in the parietooccipital region. There was also slight triangulation of the frontal region (Figure 1 a,b). Coronal sutures were not observed and the lambdoid suture patency could not be differentiated and evaluated in between the defective regions previously mentioned. The patient underwent thin-slice high resolution cranial CT examination using multislice CT scanning and reconstruction images were obtained for three-dimensional analysis. Metopic suture was closed with a resultant slight triangular configuration of the frontal region, better depicted in anterior oblique projection (Figure 2a). Lambdoid sutures were patent, while the sagittal suture was closed almost totally (Figure 2b). On lateral projections, three-dimensional images revealed that both of the coronal sutures were prematurely fused (Figure 2c, d). Cranial vault demonstrated multiple exaggerated convolutional markings in the form of lacunar skull, especially in the parietooccipital region (Figure 2b-d). The diagnosis of compound craniosynostosis was established by the basis of radiological findings.

DISCUSSION

Craniosynostosis is premature closure of one or multiple cranial sutures. The sagittal suture is affected in 40 to 60 percent of cases, the coronal suture in 20 to 30 percent of cases, and the metopic suture in less than 10 percent of cases; true lambdoid synostosis is rare (3). Suture and fontanelle closure takes place in different times (4). For example, range of normal closure of the anterior fontanel is 4 to 26 months and posterior fontanel is birth to 2 month. Cranial development and mature sutural closure occurs by the age of 12, while the completion of sutural fusion by ossification is an ongoing process until the third or fourth decade of life (4,5).

Of affected individuals, 2-8% have primary craniosynostosis. Secondary causes include rickets, hyperthyroidism, hypercalcaemia, bone marrow hyperplasia, or inadequate brain growth (microcephaly and shunted hydrocephalus). Usually, craniosynostosis is present at birth, but it is not always diagnosed when mild. Generally it is diagnosed as a cranial deformity in the first few months of life.

The diagnosis of craniosynostosis depends on physical examination, plain radiography, and cranial CT. The diagnostic value of the CT scan outweighs that of plain radiography because the sutures can be identified more accurately on a CT scan. Modalities of suture imaging such as three-dimensional and spiral CT have improved the accuracy in diagnosis (6). Three-dimensional surface reconstruction using CT scanning can help the surgeon to accurately describe the craniofacial deformity and plan surgical reconstruction (5). In addition suture patency and range of craniosynostosis is shown well three-dimensional CT. To better understand the suture anatomy, CT evaluation provides valuable intelligence for correction of the deformity (7). In our case, there were increased convolutional images, lacunary skull with bilaterally synostotic sutures in CT examinations of the case. These were probably secondary to pulsatile pressures toward the internal tabular of cranium. The aggregated convolution images may have prevented an evident deformity development as a compensatory mechanism for the brain. Our case is unique for presence sagittal, coronal and metopic suture synostosis in one case.

In conclusion, we accentuated the importance of three-dimensional CT scan in patients with craniosynostosis.

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