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GIANT BREGMATIC WORMIAN BONES REPLACING THE ANTERIOR FONTANELLE IN TWO CHILDREN

İki Çocukta Anterior Fontanelin Yerini Alan Dev Bregmatik Wormian Kemikler

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ABSTRACT

Wormian bones are accessory bones within sutures or fontanelles. Replacement of the anterior fontanele with a bregmatic wormian bone, so-called as fontanellar bone, is an uncommon findings. They may be solely presented with absence of the anterior fontanelle or associated with craniosynostosis and also form part of various syndromes, skeletal dysplasias. We herein report two uncommon cases of giant bregmatic wormian bones replacing the anterior fontanelle in two children. Computerized tomografi (CT) is the best choice to detect both the wormian bones and possible associated craniosynostosis or central nervous system (CNS) abnormalities. Adding 3D reconstruction to routine CT imaging facilitates detection of wormian bones.

Key words: bones and bone; anterior fontanelle, tomography

ÖZET

Wormian kemikler sütürler içerisinde veya fontanellerde yerleşmiş aksesuar kemiklerdir. Fontaneller kemik olarak adlandırılan, bregmatik kemiğin anterior fontanelin yerini alması sık rastlanmayan bir bulgudur. Fontaneller kemik sadece anterior fontanelin yokluğu ile ortaya çıkabilir veya kraniosinostoz ve ayrıca çeşitli sendromların, iskelet displazilerinin bir parçası olarak görülebilir. Biz burada iki çocukta anterior fontanelin yerini alan, sık görülmeyen iki dev bregmatik wormian kemik vakasını sunuyoruz. Bilgisayarlı tomografi (BT) hem wormian kemiklerin hem de olası kraniosinostoz veya santral sinir sistemi anormalliklerinin saptanmasında en iyi seçenektir. Rutin BT imajlarına 3D rekonstriksiyon imajlarının eklenmesi wormian kemiklerin saptanmasını kolaylaştırır.

Anahtar kelimeler: kemik ve kemikler; anterior fontanel, tomografi

INTRODUCTION

Vormian bones (supernumerary bones or ossicles) are accessory, irregularly shaped bones developed from independent ossification centers found along cranial suture lines and fontanelles (1). Replacement of the anterior fontanele with a bregmatic wormian bone, so-called as fontanellar bone, is an uncommon findings (2). To our knowledge, there are a few reports concerned about bony replacement the anterior fontanelle with an additional bone with or without associated craniosinostosis in the literature. They may be isolated findings as well as associated with other abnormalities such as CNS abnormalities, skeletal dysplasias (1-4). We herein report two uncommon cases of giant bregmatic wormian bones replacing the anterior fontanelle in two children with computed tomography findings.

CASE 1

A one and half year-old boy, who had been operated for repair of cleft palate at the age of one month was admitted to pediatry department. On physical examination, hypertelorism, inguinal hernia and neuromotor retardation were found. He was referred to our department because of severe neuromotor retardation. Multidetector computed tomography (MDCT) was carried out with a 64-row MDCT unit (Aquilion 64; Toshiba, Tokyo, Japan) using a 0.5-mm section width, Reconstructed images were processed, and then 3D volume-rendering (VR) images was processed on a separate workstation (Vitrea 2; Vital Images, Plymouth, Minn, USA). On CT imaging, lateral ventricles were parallel to each other, suggesting corpus callosum agenesis or dysgenesis. Three dimensional VR images showed a trigonal shaped wormian bones at bregmatic region, measuring 6,5x6cm. Metopic suture was closed (Fig.1.A). Following this, magnetic resonance imaging (MRI) was performed by 1.5 MR scanner (Vision Plus, Siemens, Germany) to confirm the agenesis or dysgenesis of the corpus callosum., The parallel configuration of lateral ventricle on axial images and also “Viking helmet” sign on coronal images were observed (Fig.1.B,C). MRI verified agenesis of corpus callosum.

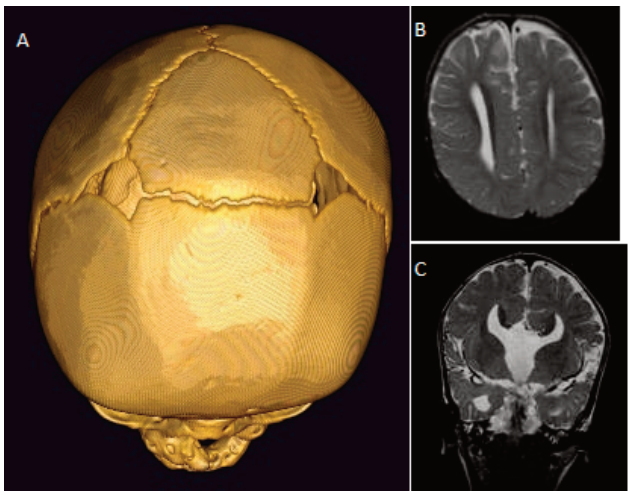

Figure 1. On computed tomographic image with 3D reconstruction, the triangular shaped wormian bone located within anterior fontanelle is seen. Metopic suture is closed (A). The paralel configuration of lateral ventricle on T2-weighted axial image (B) and "Viking helmet" sign on T2-weighted coronal image (c), highly suggestive findings of corpus callosum agenesis are observed.

CASE 2

A 3-month-old boy referred to our department with suspicion of premature clossure of anterior fontanelle. CT imaging was performed and then 3D VR images were constituted by same device. There were no abnormalities found in the cerebral and cerebellar hemispheres. Axial bone window CT images demonstrated additional bone at the bregmatic region (Fig.2.A). 3D VR images nicely revealed a wormian bone filling the anterior fontanelle. Axial CT image showed the rhomboid shaped wormian bone measured 4x4 cm. The metopic, sagittal, coronal, and lamboid sutures were normal (Fig.2.B).

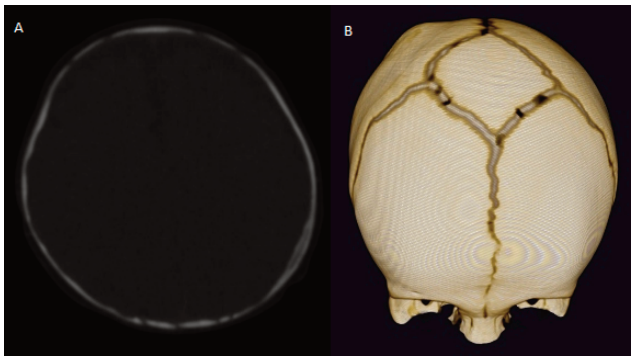

Figure 2. Additional bone at the bregmatic region is seen on axial bone window CT image (A) 3D reconstruction easily shows a rhomboid shaped vormian bone filling the anterior fontanelle. The metopic, sagittal, and coronal sutures are normal (B).

DISCUSSION

Wormian bones are accessory bones within sutures or fontanelles. Their developmental pathogenesis is disputable. Two hypothesis that explain development of wormian bones are widely accepted. The first hypothesis indicates that presence and frequency of wormian bones are primarily under genetic factors, and external influence such as cultural cranial deformation do not play an important role in wormian bone formation. In second hypothesis, enviromental factors (e.g. cultural cranial deformation, experimentally created craniosinostosis) was blamed for development of wormian bones (5,6).

The anterior fontanelle, is located at the junction of the metopic, sagittal, and coronal sutures. Normally present at birth, the anterior fontanelle is diamond shaped, and measures about 4 cm in its antero-posterior and 2,5 cm in its transverse diameter. The anterior fontanelle closes by progressive ossification of the surrounding bones, to close completely by 12 to 18 months of life (4). Evaluation of anterior fontanelle is clinically important because of sunken fontanelle indicating dehydration and bulging of the fontanelle recognized sign of raised intracranial pressure (4). The wormian bone at anterior fontanelle is certainly a rare occurence (2). The incidence of bregmatic bones changes from 0% to 2,5 % in several series (2). There are a few reports concerned about bony replacement the anterior fontanelle with an additional bone with or without associated craniosynostosis in the literature (1-4). To date, the two largest vormian bones reported to date measure about 8x7 cm and 5,7x5,5cm (3,4). The wormian bones in our report were also large size. We measured the wormian bones 6.5x6 cm in our first case and 4x4 cm in our second case. The wormian bones may cause localized distortion of skull growth. They may be solely referred with absence of the anterior fontanelle or associated with craniosynostosis and also form part of various syndromes, skeletal dysplasias (4). Sometimes these bones may be incidental findings (2). The patients with bregmatic bones associated with can be treated surgical procedures. Operative intervention is not required in the patients in whom the bregmatic bones are incidental findings or no association with craniosynostosis (4).

The bregmatic wormian bones can be detected by imaging methods or operatively or at autopsy (1-4). CT is the noninvasive and best modality to both confirm the diagnosis, detect associated craniosynostosis, CNS abnormalities. VR images provide easy detection of wormian bones. We did not recognize these bregmatic bones on axial CT images before assessing with 3D reconstruction.

In conclusion, because wormian bones may be unnoticed on axial imaging, we recommend to add 3D reconstruction for evaluation of these bones that may be part of craniosynostosis or syndromes.

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