Unusually Large Anterior Fontanellar Bone and Diffuse Capillary Malformation with Overgrowth in a Three-Month-Old Child

A Computed Tomography Case Report

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Key words: os bregmaticum, capillary malformation, overgrowth

SUMMARY – *The case presented describes the simultaneous occurrence of an unusually large anterior fontanellar bone with a syndrome of vascular malformation and overgrowth in a three-month-old child, which to our knowledge has not yet been reported. This combination may strengthen the arguments for a possible genetic contribution to the occurrence of supernumerary ossicles in the skull. Although of minor clinical importance, the shape and variations of these Wormian bones should be well-known to prevent misleading interpretations of imaging Results.*

Introduction

Wormian bones are considered to result from abnormal ossification centers in the cranium. Whereas the role of genetic or “epigenetic traits” versus environmental conditions in their formation is a matter of debate, their clinical impact is usually considered to be an abnormal variant. However, in cases of multiplicity, a number of pathologic conditions can be associated such as craniosynostosis, osteogenesis imperfecta, rickets, cleidocranial dysplasia, hypothyroidism, Down syndrome and others¹.

These supernumerary ossicles present as sutural or fonticular bones and among them, a localization in the region of the anterior fontanel is regarded as unusual². Here, we present the case of a three-month-old child with an extraordinarily large anterior fontanellar bone in combination with a diffuse capillary angioma and a hyperplasia of the left extremities which match the criteria of a “diffuse capillary malformation with overgrowth”³.

Case Report

This male baby was born with a diffuse port-wine stain nevus affecting the right half of his face and both sides of his chest and both arms, and a moderate overgrowth of his left arm and leg (Figure 1). Clinical examination and Doppler sonography did not show any signs of dilated veins or arteriovenous shunts thus ruling out Klippel-Trenaunay and Parkes-Weber syndromes⁴.

Because of spells of absence showing typical 3 Hz spike and wave discharges and because magnetic resonance imaging was not available for logistic reasons, computed tomography (CT) was performed at the age of three months. CT showed a normal brain with just slightly enlarged frontal sulci, but an unusually large accessory rhomboid-shaped bone covering the complete area of the anterior fontanel (Figure 2). This ossicle was divided horizontally by the frontal fissure into a smaller triangular-shaped posterior and a larger oval-shaped anterior portion. The surrounding sutures were

Figure 1 Photograph of the 3-month-old baby showing the port-wine stain nevus covering the right half of his face, the upper part of his chest and both arms, as well as the slight enlargement of his left arm.

Figure 2 Surface reconstruction from skull CT data as viewed from an anterior-superior position. The anterior fontanel is covered by an ossicle extending downwards between the two parts of the frontal bone, which are divided by the metopic suture. The ossicle is divided into a smaller superior and a larger inferior part by the frontal suture.

Figure 3 Same CT, horizontal slices below the ossicle (left), through its inferior part (middle) and through its superior part (right). All sutures are open, however the suture bordering the right side of the ossicle is slightly smaller than the left one.

open as well as all other sutures including the metopic suture, and the posterior and lateral fontanels. However, the suture separating the ossicle from the right half of the frontal bone was slightly smaller than the contralateral one

(Figure 3). Since then, the child has developed normal without further seizures for over three years now showing a normal-shaped skull, and further growth of the affected extremities was proportional to the rest of the body.

Discussion

As stated above, replacement of the anterior fontanel with a fontanellar bone is an uncommon finding. To our knowledge, the case presented is unique not only because of the combination with a vascular malformation and overgrowth syndrome, but also because of the large size of the specimen and its separation into two parts.

An extensive anatomical description of ossicles covering the frontal fontanel was given by Le Double⁵ as "os bregmatique" which as in our case, can be divided by the frontal suture into a superior and an inferior part ("Os hmi-bregmatique suprieur et infrieur"). This description exactly matches our findings. The irregular and varying shape of the ossicle, which does not always correspond to the fontanellar shape, was also mentioned in this early report. Since then, several anatomic and radiologic reports have been published showing single and multiple small splinter-like and larger fragments mainly in the inferior parts of the anterior fontanel, but also a special pentagonal Wormian bone in a predominant parietal localization possibly deriving from "a proper additional ossification center"¹.

The occurrence in two siblings⁶ and the combination with a syndrome of vascular malformation and overgrowth as in our case might point to a genetic origin of ossicle formation. However, further external and internal factors such as artificial skull deformations and

altered tensile forces caused by prematurely closed sutures also have to be considered as possible causes⁷. These mechanical factors are believed to increase the number of Wormian bones in an individual case, but not their incidence¹. The slight asymmetry between both sutures forming the lateral borders of the inferior part of the fontanellar ossicle in our case may be interpreted in this way although a definitive premature synostosis can be excluded by imaging and further development of the child.

The time of onset of ectopic ossification has been reported to range from late fetal stages⁸ as it may be in our case, to early infancy including the two and a half years after birth⁹. From a clinical point of view, a large ossicle of the frontal fontanel without additional skull abnormalities may be considered a variant which does not interfere with cranial growth or normal development, and special treatment is not indicated².

Conclusion

Our case of an unusually large anterior fontanellar bone in combination with a syndrome of vascular malformation and overgrowth may strengthen the arguments for a possible genetic contribution to the occurrence of supernumerary ossicles in the skull. The shape and variations of these Wormian bones should be well-known to prevent misleading interpretations of imaging Results.

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