Rare Presentation of Rapidly Involuting Congenital Hemangioma of the Skull: A Case Report

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Patient: Female, newborn
Final Diagnosis: Rapidly involuting congenital hemangioma
Symptoms: No obvious symptoms
Clinical Procedure: —
Objective: Unusual clinical course

Background: Rapidly involuting congenital hemangioma (RICH) of the fetal skull is an extremely rare vascular disease which undergoes proliferation only in utero and progresses with maximal size at birth. RICH can be detected by prenatal imaging but is easily misdiagnosed.

Case Report: A 28-year-old nulliparous woman was referred at 38 weeks of gestation for routine screening with obstetric ultrasonography. The ultrasonography revealed a female fetus with a previously undetected head tumor (32×22 mm). Certain unusual sonographic features were observed: the lesion was fusiform, with a wide base adjacent to the frontal bone. Tumor growth appeared to be toward the brain parenchyma rather than outwards (ie, toward the skull), which suggested that the mass may have been derived from the skull. The mass may have remained undiagnosed due to its small size or due to the superimposition of the skull in poor quality ultrasound images. On the basis of ultrasound findings, the lesion was diagnosed as an intracranial tumor, but fetal MRI findings led to the suspicion of RICH of the fetal skull. Finally, the patient was followed up until 1 year after birth, by which time the lesion had completely disappeared.

Conclusions: Careful evaluation of prenatal ultrasound is necessary to ensure detection of any mass adjacent to the skull, and the ultrasonography technician should carefully examine the features of any suspected mass to diagnose it correctly to avoid affecting the treatment strategy.

Keywords: Hemangioma, Cavernous, Central Nervous System • Prenatal Diagnosis • Skull • Ultrasoundography, Doppler

Abbreviations: RICH – rapidly involuting congenital hemangioma; CHs – congenital hemangiomas; MRI – magnetic resonance imaging

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Introduction

Congenital hemangiomas (CHs) are very rare lesions, which makes it difficult to accurately determine their incidence [1]. A prospective study reported CH in 0.3% (2/594) of newborns [2]. Hemangiomas are vascular tumors characterized by endothelial cellular proliferation, which is stimulated after birth (tenth day) and followed by subsequent, slow involution. Consequently, infantile hemangiomas are rarely diagnosed during the prenatal period. Immunohistochemical staining for the marker GLUT1 is positive in all cases of infantile hemangiomas but not in any other type of infantile vascular tumors [3]. CHs are classified into rapidly involuting, partially involuting, and non-involuting types [4]. Unlike the case with infantile hemangiomas, CHs progress in utero and appear to be fully developed at birth. Rapidly involuting CH (RICH) usually exhibits spontaneous involution during the first 2 years, but non-involuting CHs do not show any postnatal regression and therefore often require surgery. RICH of the skull is an extremely rare benign lesion arising from the skull, mostly affecting infants. Prenatal diagnosis of RICH is important for childbirth and postnatal care and also helps avoid unnecessary surgery [1]. RICHs can be detected by prenatal imaging, but they are often misdiagnosed [5].

Herein, we present the case of a female neonate diagnosed with CH that was detected during the prenatal ultrasound and magnetic resonance examination and showed spontaneous complete involution within 1 year of birth. On the basis of our experience in this case and a review of similar cases in the literature, we have identified a few characteristic ultrasonographic findings of RICH of the skull. We believe that this report will be a useful resource on the diagnosis of CH in fetuses using ultrasonography.

Case Report

A 28-year-old nulliparous woman was referred at 38 weeks of gestation for obstetrical ultrasonography as part of routine screening, and the examination revealed a female fetus with a head tumor that had remained undetected until that time. Ultrasound examination revealed a 32×22 mm right-sided temporal heterogeneous mass in the parenchyma adjacent to the frontal bone, with an area of focal hypechogenicity. The mass had an indistinct boundary. A wide base was observed, with no other abnormalities. The findings of physical examination were otherwise unremarkable. The lesion was large enough to cause compression of the frontal lobe. Based on these findings, the mass was suspected to be a congenital hemangioma of the skull.

Following ultrasonography, fetal MRI was performed, which showed a well-circumscribed tumor with a maximum diameter of 39 mm. The mass showed heterogeneous hyperintensity with small hypointensity foci on T2-weighted images (Figure 1C) and heterogeneous isointensity with a few internal areas of hyperintensity foci on T1-weighted images. The mass arose from the dipole of the frontal bone, and the inner table was discontinuous and located outside the dura mater. The lesion was large enough to cause compression of the frontal lobe. Based on these findings, the mass was suspected to be a congenital hemangioma of the skull.

The newborn was delivered at 39 weeks of gestation without any complications, with Apgar scores of 7 and 8. Over the right temporal area, a slightly raised 1.2×1 cm lesion was noted, with no other abnormalities. The findings of physical examination were otherwise unremarkable. The findings of laboratory tests, including platelet count, serum α-feto protein, carcinoembryonic antigen (CEA), and β-HCG, were normal, as were the echocardiography findings. Ultrasonography and contrast-enhanced MRI were performed 1 day later after birth (Figure 1D, 1E).

Ultrasound examination revealed a right-sided temporal heterogeneous mass of size 43×32 mm in the parenchyma adjacent to the frontal bone, with hypechogenic foci. No distinct boundary of the mass was visible. Doppler imaging suggested the presence of punctiform flow into the lesion. The findings of postnatal conventional MRI were similar to those of the prenatal MRI. However, contrast-enhanced MRI revealed progressive enhancement of the tumor after contrast injection.

At 1 and 3 months of age, physical examinations revealed a small mass on the right temporal region, whereas MRI revealed a gradual but significant reduction in the size of the mass. Finally, the tumor disappeared completely by the time the infant was 1 year old (Figure 1F).

Discussion

This case report presents a cause of RICH that was detected during the third trimester, as reported in most case reports published previously. A review of existing literature revealed that the tumor was detected in utero in only 5 cases of CH of the skull, including our case [3,5,6]. In all of these cases, the tumor was detected after 26 weeks of gestation.

Ultrasonography revealed a whole solid component with a homogeneous or slightly heterogeneous pattern and signs of a small amount of blood flow within the mass. Therefore, we ruled out hematomas. As observed in our case, small echogenic foci...
Gray-scale (A) and color Doppler (B) ultrasound images of the case at 38 weeks revealed a 32×22 mm right frontal heterogeneous mass with various focal hypoechogenic (red arrow). A few small hyperechogenic foci can be observed in the lesion and a wide base of case can be detected close to frontal bone. (C) Axial T2-weighted image of case at 38 weeks on MRI. The mass appears hyperintense of the mass in the frontal bone (red arrow). (D) Axial T2-weighted image of case on MRI revealed 1 day later after birth (red arrow). (E) Ultrasound revealed right-sided temporal heterogeneous and fusiform mass in the frontal bone 1 day after birth. (F) The tumor was complete involution in the T1-weighted image 1 year later.
suggestive of calcifications were observed in the imaging studies of 4 other cases in the literature. Gorincour et al. [7] also reported that 37% of RICHs had features of calcification within the lesions. Considering these findings, we may infer that calcification may be one of the features of RICHs located in the skull.

Although the imaging features of RICH of the skull have been reported previously [3,5,6], most of the reports describe extracranial lesions lying adjacent to the skull, which enables easy diagnosis. However, we noted a few ultrasound imaging features of CHs involving the skull that warrant attention since they are different from those reported previously. We believe that knowledge of these features can improve recognition of the lesions. First, the mass in this case showed growth toward the brain parenchyma rather than the exterior of the skull. This may lead to the suspicion that the mass was derived from the brain parenchyma. However, we noted that the lesion had a wide base attached to the skull, which suggested that the mass was not an intracranial lesion. In addition, the mass was fusiform in shape—a feature suggesting that the mass was an extracranial lesion. Moreover, the cranial cortex attached to the mass was discontinuous. Together, these ultrasonographic findings may provide useful diagnostic information in cases of RICH. In a few cases reported in the literature [8,9], an obvious hyperechogenic line, called the periosteal reaction, was noted extending from the adjacent bone, overlying the lesion. However, this finding was not observed in our case, whereby the condition could have easily been misdiagnosed.

Although several signs can lead to a diagnosis of RICH, a differential diagnosis including a number of similar conditions is still necessary. Infantile hemangioma is the lesion most similar to RICH and is easily misdiagnosed. Antenatal ultrasonography has been shown to detect up to 75% of CHs as early as 12 weeks of gestation [10]; therefore, imaging plays a vital role in the characterization of CHs. Although histopathologic examination is the criterion standard for diagnosis, it is rarely required for RICHs, as in our case. Olsen et al. have shown that the presence of venous lakes, venous ectasia, and calcification on ultrasonography can help distinguish CHs from infantile hemangiomas [1]. Additionally, GLUT-1 has been reported to be a highly sensitive and specific immunohistochemical marker for infantile hemangiomas; therefore, when the clinical picture is unclear, the absence of GLUT-1 expression can help differentiate CHs from IHs [11]. Another differential diagnosis included in our case is intracranial solid tumor because the tumor in this case extended intracranially. Therefore, it is necessary to evaluate the imaging features of the lesion and accurately determine its origin. Yet another condition to be differentiated from CHs is cephalohematoma, but this condition was relatively easy to rule out because the lesion in our case showed minimal blood flow, unlike cephalohematomas.

In all the reported cases, fetal MRI had been performed, but it showed no specific signs attributable to the lesion. The clinical presentation in our case was different from that of common hemangiomas because hemangiomas generally present with a hyperintense pattern on T2-weighted postnatal MRI [12]. We therefore infer that hypointense or isointensity on T2-weighted imaging of fetal MRI does not exclude the diagnosis of RICH of the skull [1]. We believe that the best method to identify RICH is ultrasonography or contrast-enhanced MRI, if possible. Follow-up to evaluate changes in the mass is also necessary.

Regarding treatment, spontaneous resolution of the lesions occurred within 1 year of birth in 4 cases, including ours, with no specific treatment required during the follow-up period. Surgical excision was performed only in 1 of the reported cases, on the third postnatal day, for esthetic reasons [6].

Conclusions

We present the characteristic prenatal identification and serial postnatal imaging findings of a RICH, providing an insight into the evolution and involution of the lesion during the first year of life. These tumors pose a diagnostic challenge both in the fetus and neonate, and recognition of the characteristic imaging and clinical features would help avoid unnecessary medical intervention.

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Ethics Statement

This study was approved by the Ethics Committees of the Institutional Review Board of Women and Children’s Hospital of Chongqing Medical University.

Declaration of Figures’ Authenticity

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References: