

Prenatal imaging findings in rapidly involuting congenital hemangioma of the skull

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KEYWORDS: angioma; imaging; prenatal; RICH; skull; ultrasound

ABSTRACT

We report two cases of rapidly involuting congenital hemangioma (RICH) of the skull diagnosed in the third trimester of gestation, and also present a brief review of the literature. In both of our cases ultrasound examination showed a soft tissue vascular mass of the skull with a specific sonographic finding: a thin hyperechogenic line over the lesion and continuous with the calvaria, suggesting a subperiosteal origin and possibly accounting for a mass effect on the underlying skull. This was slight in one case and marked in the other (and associated with involvement of the calvaria). On prenatal T2-weighted magnetic resonance imaging, the signal of each of the lesions was less marked than the hypersignal encountered in the postnatal period. Postnatal clinical and radiological follow-up over the first few months after delivery confirmed the diagnosis of RICH in each case by demonstrating a significant decrease in the size of the tumor and regression of the vascular component, with complete involution of the lesion within a year. Copyright © 2008 ISUOG. Published by John Wiley & Sons, Ltd.

CASE REPORTS

Case 1

A 39-year-old woman, gravida 3 para 2, was referred at 34 weeks' gestation following ultrasound detection of a head tumor in a male fetus 1 week earlier. Ultrasound examination at 33 weeks' gestation revealed a 30 × 38-mm right-sided occipital heterogeneous mass with multiple hypoechogenic foci. A thin hyperechogenic line continuous with the calvaria was observed at

the boundary of the lesion and was interpreted as corresponding to the periosteum (Figure 1). The occipital bone was slightly deformed. A few small hyperechogenic foci were also visible within the mass, suggesting microcalcifications. Doppler imaging suggested a high-flow lesion. No other abnormalities were detected.

Magnetic resonance imaging (MRI), performed at 34 weeks' gestation, showed the tumor to have a maximum diameter of 42 mm. On T2-weighted imaging, the tumor appeared homogeneously hypointense with no intra- or perilesional signal voids (Figure 2). No T1-weighted images were obtained. Because of the large size of the lesion, its high vascularity and its location, a diagnosis of congenital hemangioma of the skull was suspected.

The infant was examined at 46 days of age. On physical examination, a warm and bluish mass with a small portwine stain was present on the right side of the occiput (Figure 3). Ultrasound examination of the mass revealed a relatively homogeneous tumor with high-flow vessels and a few echogenic foci suggestive of microcalcifications. At 3 months of age, the mass was significantly smaller, less warm and less tense. Ultrasound examination showed echogenic fibrous tissue replacing the mass with a significant decrease of its vascular component. At 14 months of age, the mass had completely disappeared.

Case 2

A 27-year-old woman was referred at 33 weeks' gestation following detection of a left frontal tumor in a female fetus on ultrasound examination. The tumor was homogeneous and echogenic. An echogenic border continuous with the calvaria was observed. The tumor had a wide insertion on the periosteum and markedly deformed the frontal bone

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Accepted: 8 February 2008

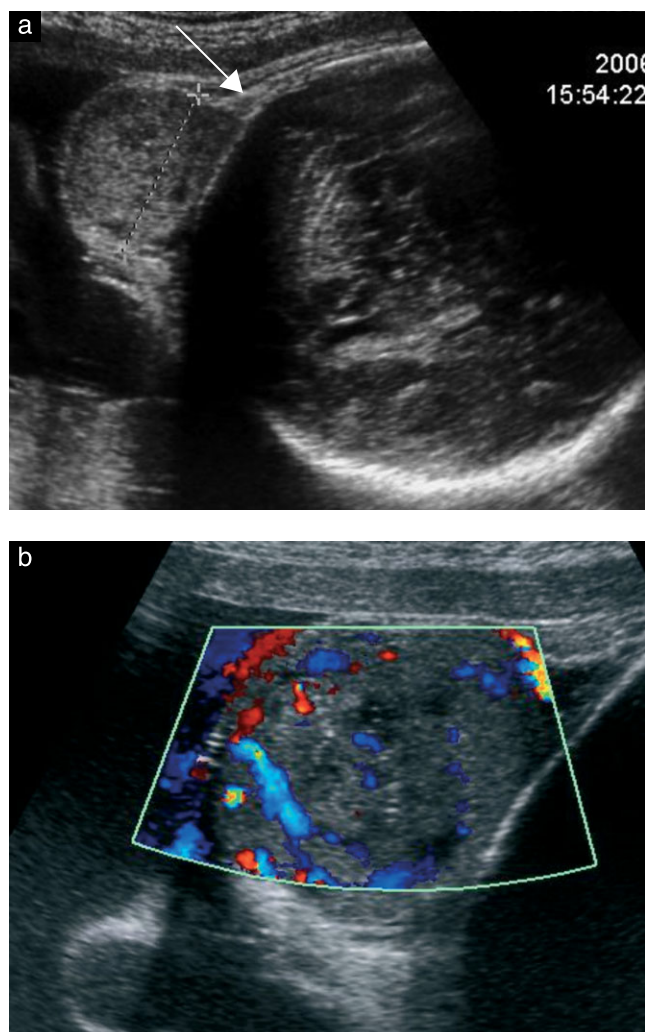


Figure 1 Gray-scale (a) and color Doppler (b) ultrasound images of Case 1 at 33 weeks showing a 30 × 38-mm occipital right-sided vascular heterogeneous mass with multiple hypoechoogenic foci. A thin hyperechogenic line continuous with the calvaria can be observed at the boundary of the lesion (arrow).

(Figure 4a). Doppler imaging showed arterial-type flow with low resistance (Figure 4b).

MRI at 34 weeks' gestation showed a left frontal tumor (dimensions 50 × 20 × 45 mm) with a slightly heterogeneous signal and hypointense foci, with no obvious signal void on T2-weighted imaging (Figure 5). On T1-weighted imaging the signal was also slightly heterogeneous with small hypointense foci. The frontal bone showed a marked depression adjacent to the mass, and was heterogeneous and very thin. Ultrasound examination at 35 weeks' gestation showed a homogeneous echogenic mass of unchanged size with arterial and venous components. For the same reasons as for Case 1, a diagnosis of congenital hemangioma of the skull was suspected.

After birth, the newborn was examined at 2 days of age. The firm and warm left frontal mass was covered by normal skin. On gray-scale and Doppler ultrasound examination, the tumor was relatively homogeneous, with arterial and venous blood flow and a scalloping of the outer table. Computed tomography demonstrated

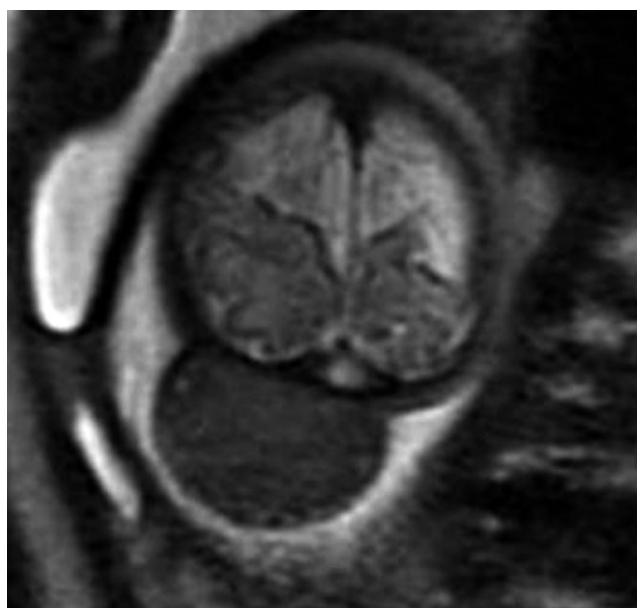


Figure 2 T2-weighted magnetic resonance image of Case 1 at 34 weeks showing a coronal slice. The tumor appears homogeneous and slightly hypointense, with a small depression of the occipital bone.



Figure 3 Appearance of the hemangioma in Case 1 46 days after birth.

a depressed frontal bone showing a lacunar appearance of the cranial vault, but without extension into the inner table of the calvaria. The tumor was hyperdense and enhanced after contrast injection. Postnatal MRI showed a heterogeneous intermediate to hyperintense lesion with signal voids on T2-weighted imaging. The signal was hypointense on T1-weighted images, with marked enhancement from the periphery to the center of the lesion after gadolinium injection.

By 3 months of age the tumor had significantly decreased in size, supporting the diagnosis of rapidly involuting congenital hemangioma (RICH). At 1 year of age, the mass had completely disappeared but a marked depression of the frontal bone remained.

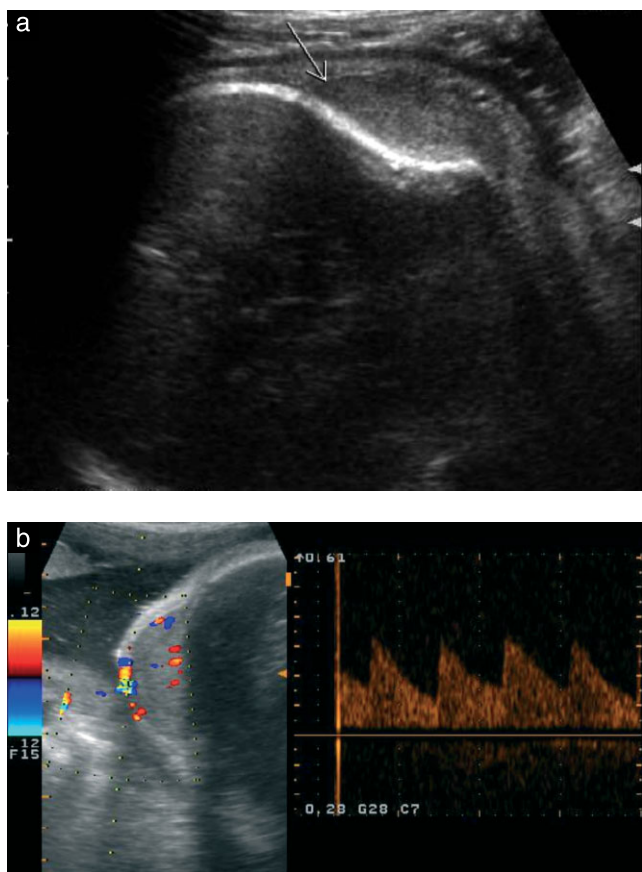


Figure 4 Gray-scale (a) and color Doppler (b) ultrasound images of Case 2 at 33 weeks. The frontal tumor is homogeneous and echogenic. An echogenic border continuous with the calvaria can be observed (arrow). The tumor has a wide insertion on the periosteum and markedly deforms the frontal bone (a). Doppler imaging shows an arterial low-resistance spectrum (b).

DISCUSSION

With advances in prenatal imaging, vascular anomalies of the soft tissues are being diagnosed prenatally with increasing frequency¹. Vascular anomalies are categorized into two major types of lesion according to endothelial characteristics: vascular tumors and vascular malformations^{2,3}. Among vascular tumors, infantile hemangioma (IH) is the most common. However, IH appears postnatally and is rarely present at birth. Conversely, congenital hemangioma progresses during fetal life to manifest as fully developed at birth⁴. Congenital hemangiomas are subdivided into RICH and non-involuting hemangioma. RICH is the most frequent type and commonly appears as a raised tumoral lesion; these lesions resolve more rapidly than common IHs to regress completely before the age of 14 months^{4,5}. Because RICHs are more frequent, especially on the skull and in the peri-auricular region, and have a bulging tumoral presentation, these vascular tumors of the skull can potentially be diagnosed by prenatal imaging.

Based on our experience, and on a review of the recent literature of nine cases^{1,6–10}, we have aimed to identify the imaging findings that are highly suggestive of RICH of the skull. We included in our review a ‘cavernous

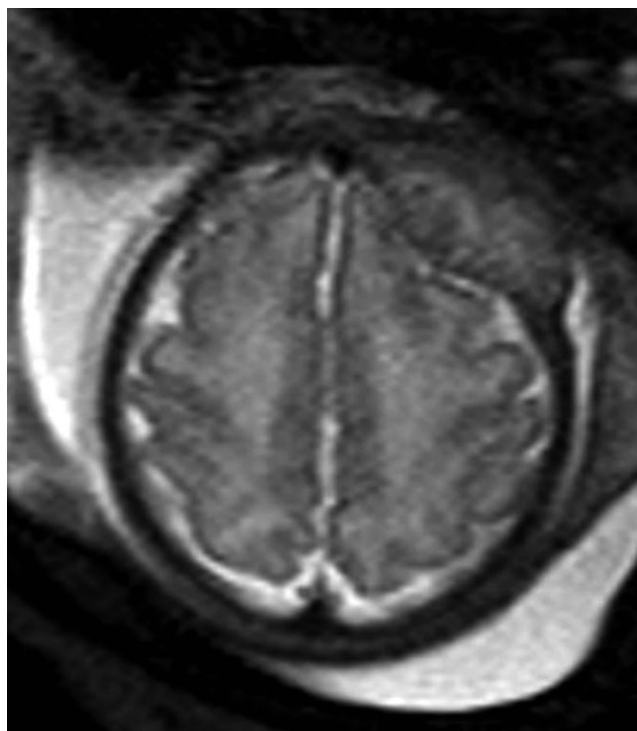


Figure 5 T2-weighted magnetic resonance image of Case 2 at 34 weeks showing an axial slice. The tumor has a slight heterogeneous intensity with hypointense foci.

hemangioma’, which can refer to either a hemangioma or a venous malformation¹. However, on the basis of the authors’ description, we considered this lesion to be a RICH⁷.

In both of our cases the lesions were diagnosed during the third trimester, as most commonly reported in previous cases. In seven of nine cases included in our review the diagnosis was made at or after 24 weeks (late second or third trimester). However, the lesion was detected as early as 16 and 18 weeks in the two other cases¹. In all cases, ultrasound imaging showed a solid appearance with a homogeneous or slightly heterogeneous pattern, similar to placental echogenicity without any cystic component. Vessels accounted for the heterogeneous pattern, as shown by hypervascularization which was found in all but one case⁷. In one of our two cases small echogenic foci suggestive of calcifications were found using a high-frequency probe; these were also found on postnatal examination, and in 37% of cases in a series of postnatal ultrasound analyses of RICH published by Gorincour *et al.*¹¹.

In our cases, there are two sonographic findings regarding the relationship of the tumor to the underlying skull that should be focused on. A deformity of the underlying bone was observed in both of our cases. However, this has also been reported in other scalp hemangiomas^{7,9,11}. In addition, our cases clearly showed a hyperechogenic line extending from the adjacent bone over the lesion, which can be observed retrospectively in prenatal images of other cases^{6,9,11}. This line was interpreted as the periosteum, suggesting a subperiosteal location of the hemangioma rather than a hypodermic

origin. These two sonographic findings may provide useful diagnostic information.

In the six cases in which a fetal MRI was performed (including our cases), the signal of the lesion was either markedly or slightly hyperintense (four cases) or hypointense (two cases) on T2-weighted imaging (compared with brain parenchyma)^{6,7,9}. This should be emphasized because hemangiomas are characteristically hyperintense on T2-weighted postnatal MRI⁷. Therefore, a decreased signal on fetal MRI, even with a hyposignal (as observed in two patients with proven RICH), should not exclude the diagnosis of RICH of the skull.

It should be stressed that no hemodynamic complication such as congestive heart failure or hydrops was associated with RICH of the skull, despite these hemangiomas being high-flow lesions. The skull location appears to present a lower risk of hemodynamic complication than does prenatal isolated hepatic hemangioma, which is considered to be a RICH¹².

In the postnatal period, our cases were managed conservatively with a typical involution in the first year after delivery, as in four of the nine cases reviewed. However, five lesions were surgically excised in infancy for esthetic reasons because of bleeding or ulceration, as a scar related to excision is less pronounced than a scar following ulceration^{1,6,7,9}.

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