Deep congenital hemangioma: prenatal diagnosis and follow-up

Abstract: Congenital hemangiomas (CHs) are rare benign vascular tumors that are present at birth after full development in utero. With the increasing importance of prenatal screening and improved imaging techniques, vascular tumors will be detected more frequently during examination. Ultrasonographers and obstetricians should be aware of these vascular tumors, their differential diagnosis and prognosis. Not only caregivers need optimal counseling, but also the professionals involved should have knowledge about these anomalies because of an increased risk of complications during and after the delivery of the child. Here, we present a child with a CH of the leg and discuss prenatal diagnosis, intra-partum management, and postnatal follow-up.

Keywords: Hemangioma; multidisciplinary management; prenatal ultrasound; vascular tumor.

Case presentation

A 30-year-old woman, G2P1 with a spontaneous bichorionic twin pregnancy, was referred to our hospital at 29 weeks of gestation, after routine sonography had revealed a superficial growing mass on the leg of one of the fetuses, suggestive of a hemangioma. The mother’s medical history and the antenatal course of her current pregnancy were unremarkable.

At 23+1 weeks of gestation, ultrasound (US) examination showed a homogeneous soft tissue mass (~2.7 × 0.8 cm) of the right lower leg in one of the fetuses, with multiple feeding arteries and drained by the popliteal vein, suggestive of a hemangioma (Figure 1A and B). US examination at 29+6 weeks of gestation showed moderate insufficiency of the tricuspid valve, and Doppler examination revealed increased flow in the umbilical vein with a normal pulsatility index of the ductus venosus. Biometry of both fetuses was appropriate for gestational age. During follow-up, the mass increased to a size of 1.5 × 5.0 × 4.0 cm at 32+6 weeks of gestation (Figure 1C and D). Despite previous signs of hyperdynamic circulation, the child did not develop cardiac decompensation and circulation parameters turned normal. At that time, on basis of these findings, the lesion was suspected to be a congenital hemangioma (CH). The parents were informed about the possible diagnoses and the benign course of these lesions. Close antenatal follow-up was advised using US to score the size and the development of the lesion. The professionals involved were informed about the risks during and after delivery and a cesarean section (CS) was advised.

At 35+1 weeks of gestation, the mother presented with spontaneous labor and a planned CS was performed. The first child was delivered without complications, seemed healthy at first physical exam and was admitted because of prematurity. Then, the affected boy, weighing 2740 g, was delivered. Because of persistent respiratory distress, the infant had to be transferred to the neonatal intensive care unit, where continuous positive airway pressure (CPAP) was started. Respiratory distress was thought to be the result of wet lung disease and resolved within 2 days.
following CPAP. No signs of cardiac decompensation were observed. Further neonatal physical examination was completely normal besides the known tumor of the right leg and the respiratory symptoms.

An elastic purple blue tumor with a pale edge and a dark purple spot was seen on the medial side of the right distal leg, ~5 cm in length (Figure 2A). Postnatal US showed a well-defined inhomogeneous lesion with extended vascularization measuring $4.4 \times 5.5 \times 0.6$ cm. The lesion appeared to be subcutaneous without involvement of underlying structures. Magnetic resonance imaging (MRI) corresponded with the suggested diagnosis of CH; the hypo-intense signal on T1-weighted and hyper-intensity on T2-weighted MRI scan, in combination with the presence of normal superficial veins in the subcutaneous lesion, supported the presumed benign character of the lesion. Outpatient follow-up showed impressive blanching and involution of the tumor within 6 months (Figure 2B), which confirmed the diagnosis of rapidly involuting CH.

**Figure 1** US images.
(A) First US at 23+1 weeks of gestational age showing a mass on the mediadorsal side of the right lower leg, ~2.7 cm in length (cyan line). Doppler imaging shows extended vascularity of the lesion with afferent and efferent vessels. (B) On US at 28+6 weeks of gestation, the well-defined homogeneous soft tissue mass has extended to a length of 3.9 cm (white line). (C) US at 32+6 weeks of gestation showing an increasing size of ~1.5×5 cm (yellow lines). (D) On the same US, incomplete circumferential growth of the tumor is visible (circumference 11.3 cm, yellow dotted line).

**Discussion**

CHs are benign vascular tumors that reach maturity *in utero* characteristically with no further growth after birth. There are two types of CH: non-involuting CH (NICH) and rapidly involuting CH (RICH). NICHs do not regress, whereas RICHs show complete spontaneous regression within 12–24 months in most cases [2]. Therefore, RICH does not require treatment initially and observation is justified [2, 9]. However, occurrence of complications, such as those affecting vital organs or ulceration, may necessitate intervention [7]. Although positive results have been reported, it is unclear whether new therapies for infantile hemangioma (IH), such as a β-blocker, are effective in treating CH [3, 4, 7, 9].

When detecting a well-vascularized, deep tumor *in utero*, other vascular tumors, such as kaposiform hemangioendothelioma and tufted angioma, also have to be considered because these latter lesions could be associated...
with Kasabach-Merritt syndrome (KMS). This rare consumption thrombocytopenia carries a significant risk of perinatal complications, including death. However, this phenomenon is not seen in CH or IH [9]. Hemangiomas typically exhibit a homogeneous, hypo-echoic signal, intense diffuse vascularity, and high flow on US-Doppler, which is not typical for other vascular tumors [3–5, 8]. High flow is also typical for (combined) arterial malformations (AMs) and IHs, which can be differentiated by MRI. Cystic lesions are more suggestive of lymphatic malformations, whereas calcifications are suggestive of venous malformations [7]. Both findings can also be found in teratomas [6]. When distinguishing possible diagnoses before birth has serious consequences for planning perinatal care or when uncertainty about the diagnosis (e.g., CH or AM) or prognosis of the lesion remains after enhanced Doppler US, fetal MRI is indicated [1, 6].

Close antenatal evaluation includes frequent US to monitor the lesion size and blood flow. Large hemangiomas are associated with congestive heart failure due to a hyperdynamic circulation leading to increased cardiac output; therefore, fetal circulatory parameters should also be monitored [7, 9, 10]. Termination of pregnancy has been considered in published cases in which the lesion was interfering with vital functions [10]. Regarding delivery, an elective CS is often recommended in the literature because of the possibility of fetal thrombocytopenia due to KMS with an increased chance of bleeding during normal or instrumental vaginal delivery [9]. Because of the higher a priori chance of an emergency CS or intrapartum fetal manipulation with the risk of trauma to the hemangioma in this twin pregnancy, we also performed a CS.

After birth, follow-up with a multidisciplinary approach, including frequent observation of the tumor, will confirm the prenatal and postnatal diagnosis. Especially, involution postnattally excludes malignant tumors and differentiates RICH from NICH. A biopsy can further distinguish or confirm diagnoses based upon typical histopathological characteristics. A GLUT-1 staining, for example, can differentiate an IF from a RICH and other vascular tumors [3, 5]. However, diagnosis can often be made on clinical ground and US alone [1, 5].

In conclusion, we reported a case of a prenatally diagnosed deep hemangioma of the right lower leg in a twin pregnancy. During close follow-up after birth, it appeared to be a RICH, justifying the conservative approach. When confronted with a vascular lesion, ultrasonographers and obstetricians should be aware of the differential diagnosis. Caregivers of the child and the involved professionals should be adequately informed by a multidisciplinary team of experts, which will closely monitor the vascular tumor pre-, peri-, and postnattally.

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References


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