Prenatally detected giant congenital hemangioma of the fetal neck

Shuichiro Uehara a,b,*, Toshimichi Hasegawa a, Hiroomi Okuyama a, Hisayoshi Kawahara a, Akio Kubota a, Keigo Osuga c, Eiichi Morii d

a Department of Pediatric Surgery, Osaka Medical Center and Research Institute for Maternal and Child Health, Osaka 594-1101, Japan
b Department of Pediatric Surgery, Department of Surgery, Osaka University Graduate School of Medicine, Osaka 565-0871, Japan
c Department of Diagnostic and Interventional Radiology, Osaka University Graduate School of Medicine, Osaka 565-0871, Japan
d Department of Pathology, Osaka University Graduate School of Medicine, Osaka 565-0871, Japan

ARTICLE INFO

Article history:
Received 16 February 2013
Received in revised form
1 March 2013
Accepted 1 March 2013

Key words:
Congenital hemangioma
Antenatally detected
Magnetic resonance imaging

ABSTRACT

Hemangiomas and vascular malformations constitute a variety of non-cancerous birthmarks and lesions. We experienced a rare case of a fetus with a giant congenital hemangioma detected on the fetal ultrasonography and magnetic resonance imaging (MRI). The patient was delivered via a planned cesarean section and underwent extirpation of the tumor on the 16th postnatal day due to concerns of bleeding from the tumor. A pathological examination demonstrated glucose transporter (GLUT)-1-negative lobular capillary proliferation compatible with a diagnosis of congenital hemangioma. The infant was discharged without complications on day 14 after tumor resection. Currently, at 5 years of age, no tumor recurrence has so far been observed since the extirpation. The antenatal images of this case are presented and the perinatal management is discussed.

Congenital hemangiomas (CHs) are fully formed at birth, having undergone proliferation in utero, do not exhibit the typical postnatal evolutive pattern of infantile hemangioma (IH) and can be diagnosed prenatally using ultrasonography [1]. CH is a subtype of benign vascular tumor, that is further characterized as either rapidly involuting (RICH) or non-involuting (NICH). In most infants with RICH, involution is complete, leaving anetodermic skin within 6–14 months of life, whereas NICH lesions grow proportionally with the child’s growth. NICHs never disappear and therefore require eventual excision. However, when CH is prenatally detected and there are concerns of bleeding or heart failure due to the high blood flow of the tumor, surgical resection may be performed immediately after birth.

We herein report a rare case of a fetus with a giant congenital hemangioma detected on the fetal ultrasonography and magnetic resonance imaging (MRI) that was treated with surgical resection.

1. Case report

A 29-year-old female was referred at 29 weeks of gestation due to an ultrasonographically detected tumor located on the fetal posterior neck. Ultrasonography revealed a $65 \times 67 \times 43$-mm heterogeneous tumor that made contact with the infant’s spine and skull (Fig. 1A). Color Doppler imaging revealed rich vascular formation and blood flow in the tumor. T2-weighted images of magnetic resonance imaging (MRI) obtained at 34 weeks of gestation revealed iso-high signal intensity (Fig. 1B). These findings were compatible with a diagnosis of hemangioma. During the pregnancy, fetal echography showed no increases tumor size; however, a mild right heart load was detected.

At 38 weeks of gestation, a planned cesarean section was performed with appropriate informed consent to prevent dystocia and bleeding from the tumor during vaginal delivery. A 2622-g-male infant was delivered with Apgar scores of 8 and 9 at one and 5 min, respectively. The infant did not require resuscitation or respiratory support. The tumor originated from the posterior of the neck, as observed on prenatal images. The color of the tumor became more red with each passing day, and the surface of the tumor gradually demonstrated enlargement, creating an ulcer on the surface of the tumor (Fig. 2). No signs of involution of the tumor were observed until the 15th postnatal day. Since the tumor appeared ready to bleed from the surface, tumor resection was performed on the 16th postnatal day. First, a spindle-shaped skin incision was made around the tumor. Tumor mobilization was achieved with ligations of the vessels in the subcutaneous layer, preserving the posterior neck muscles. The tumor was successfully resected completely. Skin grafting was not required to close the wound. The amount of blood loss was 132 ml, and the operative time was 3 h and 13 min. A pathological examination revealed lobular capillary

* Corresponding author. Division of Pediatric Surgery, Department of Surgery, Osaka University Graduate School of Medicine, 2-2 Yamadaoka, Suita, Osaka 565-0871, Japan. Tel.: +81 6 6879 3753; fax: +81 6 6879 3759.
E-mail address: uehara@pedsurg.med.osaka-u.ac.jp (S. Uehara).
proliferation (Fig. 3A) and a GLUT-1-negative status in the vascular endothelial cells, in contrast to the positive findings in red blood cells observed in the vessels as an internal control of GLUT-1 (Fig. 3B), compatible with a diagnosis of congenital hemangioma. The infant was discharged without complications on day 14 after tumor resection. Currently, at age 5 years of age, no tumor recurrence has not been observed since the tumor resection.

2. Discussion

The concept of a classification system for pediatric vascular anomalies was first established in 1982 based on the pathologic findings reported in a study by Mulliken and Glowacki and later modified by the International Society for the Study Group of Vascular Anomalies [2]. Congenital hemangioma (CH) is a subtype of benign vascular tumor that is further characterized as rapidly involuting (RICH) or non-involuting (NICH). Infantile hemangioma (IH) and CH are distinguished based on their clinical features and the presence of a GLUT-1-positive expression in the endothelial cells of infantile hemangioma (IH) [3]. The involution observed in cases of RICH is very similar to that observed in cases of IH, which regresses slowly after occurring during the first year of life [4]. In contrast, NICH does not exhibit the regression. Consequently, the possibility of NICH being a later stage of RICH has been suggested. Therefore, to determine the fate of RICH and NICH, monitoring for an observation period of a few years and performing serial postnatal imaging can provide insight into imaging evolution and involution of RICH during the first year of life.

Both CH and IH appear as fast-flow lesions on ultrasound and exhibit flow voids on MRI. RICH and NICH are more likely to be heterogeneous on US and much more likely to contain identifiable calcification. On MRI, these lesions exhibit high intensity on T2-weighted images and iso-intensity on T1-weighted images; prior to involution, they enhance avidly and homozygously [5]. Fast-flow is also detected in arterial vascular malformations; however, it can be easily differentiated on MRI. Prenatal screening with ultrasonography allows for the early detection of CH as early as 12 weeks of gestation [4]. A distinction between RICH and NICH lesions cannot be made on prenatal ultrasound [6]. If CH is diagnosed antenatally, the lesion can be followed on ultrasonography and MRI to further define the tumor characteristics and monitor growth [7].

In our case, immunohistochemical staining revealed negative GLUT-1 results, suggesting a diagnosis of CH. However, it is difficult to distinguish RICH from NICH before and at birth without the knowledge obtained from histological findings and observation for at least one year. Since the tumor in our case was likely to be injured by mechanical friction, we believed that it should be excised without observation to distinguish involution.

As Nolan et al. [8] previously reported, life-threatening hemorrhage associated with passage through the birth canal can result in birth trauma if the tumor is large and located on the surface of the neonate. In such cases, cesarean section should be used for delivery. Bleeding may also occur, even after birth, due to mechanical friction. Powell et al. [9] reported that two cases of severe bleeding episodes occurred during the first week of life. Close antenatal evaluation, including the frequent use of US to monitor lesion size and blood flow, should be conducted. Large hemangiomas can cause congestive heart failure due to hyperdynamic circulation leading to increasing cardiac output; therefore, fetal circulatory parameters should also be monitored. When these findings are observed, surgical excision should be considered.

Fig. 1. Prenatal images of color Doppler ultrasonography at 29 weeks of gestation showing the 65 × 67 × 43-mm heterogeneous tumor that made contact with the infant’s spine and skull. Rich vascular formation and blood flow in the tumor was observed (A). T2-weighted images of magnetic resonance imaging (MRI) obtained at 34 weeks of gestation revealed iso-high signal intensity (B).

Fig. 2. Postnatal photograph showing the neonate with a tumor on the anterior neck.
immediately after birth. Otherwise, the first line of treatment should be close observation.

3. Conclusion

We herein reported a rare case of a fetus with a giant congenital hemangioma detected on the fetal ultrasonography and MRI that was treated with the extirpation. Close follow-up before and after birth led to concerns of bleeding after birth, and a surgical approach was ultimately selected.

Conflicts of interest
None.

Sources of funding
None.

Acknowledgement
The authors thank Dr. Brian Quinn, Japan Medical Communication for editing this manuscript.

References


Fig. 3. Microscopic examination showing capillaries and feeding vessels with flattened endothelial cells on hematoxylin eosin staining (A, ×200). GLUT-1 immunostaining was negative (B, ×200), consistent with a diagnosis of congenital hemangioma.