Life-Threatening Hemorrhaging in Neonatal Ulcerated Congenital Hemangioma
Two Case Reports

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Infantile hemangiomas (IHs) are the most common benign vascular tumors found in children; they appear around 2 weeks after birth, proliferate rapidly for up to 1 year, and then slowly spontaneously regress by the age of 3 to 5 years. Conversely, congenital hemangiomas (CHs) are rare and represent less than 3% of all hemangiomas. Congenital hemangiomas fall into 2 clinical subtypes: rapidly involuting congenital hemangiomas (RICH), which tend to rapidly involute in the postnatal period so that the lesion is fully resolved by age 8 to 14 months, and noninvoluting CHs, which do not regress but instead grow in proportion to the child and often require surgical resection. Recently, a third clinical subtype based on partially involuting clinical behavior has been described. Congenital hemangiomas are hypervascularized lesions. Because of their favorable prognoses, IH and CH seldom require treatment and usually only result in a scar.

Ulceration is the most common complication of IH, which can result in pain and infection. Ulceration of IH can also induce bleeding episodes that are mainly mild and benign. In contrast, little is known about the prognosis of ulcerated CH. However, it has been observed that ulcerated CH may be complicated by life-threatening bleeding episodes.

To our knowledge, only 3 well-documented cases of severe bleeding have been reported in patients with CH. We report 2 cases of ulcerated RICH that were complicated by episodes of severe bleeding during the neonatal period. Institutional review board approval was not required for this study. Parents of the patients provided written consent.

Report of Cases

Case 1
A newborn girl was referred to our institution for a large cutaneous congenital vascular tumor. She was born at term by vaginal delivery after an unremarkable pregnancy. Results of prenatal ultrasonography performed at 32 weeks’ gestation were normal. At birth, she had a telangiectatic violaceous protuberant tumor 5 cm in diameter on her right knee (Figure 1A). A small central ulceration with a crust was observed. She also had a superficial veinous ectasia at the inner edge of the thigh with a vibratory sensation felt on the skin at palpation, suggesting a fast-flow tumor.

Findings from color Doppler ultrasonography revealed a moderately fast-flow vascular tumor with a large dilatation of...
superficial veins, in particular the right saphenous vein with another dilated branch. There was no visible shunt or abnormal vessels. Findings from magnetic resonance imaging showed a mass with T2 high-intensity signals associated with arteriovenous shunts, tortuous veins, and flow void. Results of echocardiography were normal, and the patient did not have thrombocytopenia or coagulopathy. Results of cutaneous biopsy were consistent with a benign vascular tumor with lobular proliferation and large ectatic veins (Figure 2). Immunohistochemical staining was negative for GLUT1 and positive for WT1, suggesting a diagnosis of CH.

Active treatment was not initially advised. Two weeks later, the patient suddenly developed a massive hemorrhage from the crust area of the CH that led to hemorrhagic shock and necessitated transfer to the intensive care unit. The bleeding was partially controlled using a compression dressing. Treatment with oral propranolol, 2 mg/kg/d, was started but was discontinued after 9 days as it proved ineffective in rapid volume decrease. At 3 weeks of life, given the persistence of the hemorrhage, we gave the patient several blood transfusions. A selective vascular embolization was then performed with coils and N-butyl cyanoacrylate with metacryloisulfolane. A series of arteriograms of the common femoral artery showed that voluminous pedicles were supplying the CH (Figure 3). The embolization allowed an 80% devascularization of the tumor. However, when the patient was almost 2 months old, the persistence of severe hemorrhagic episodes required several more transfusions. Therefore, we decided to perform selective hemostatic surgery, which finally stopped the bleeding. At this point, the CH rapidly
regressed, and by 4 months of age, only a telangiectatic scar with redundant skin remained (Figure 1B).

Case 2
A newborn boy was hospitalized in the pediatric ward for a hemorrhage resulting from an ulcerated CH. A CH diagnosis was suspected during the antenatal period after ultrasonography was performed at 26 weeks’ gestation. The boy was born at term by vaginal delivery after an unremarkable pregnancy. He had a 3.0 × 2.5-cm firm telangiectatic purplish tumor on his upper left thigh. The lesion was surrounded by a halo-like rim of pallor with a large central ulceration (Figure 4).

Simple occlusive dressings were initially used for treatment. At 10 days of life, massive pulsatile bleeding occurred during the replacement of the dressing. Findings from the computed tomographic angiography revealed a 3.6 × 3.0-cm mass fed by numerous small branches from the deep femoral artery. Drainage occurred through the large veins into the left external iliac vein by the saphenous vein. Treatment with oral propranolol, 2 mg/kg/d, was started and combined by pressure bandage. However, because of the persistent episodes of massive bleeding that required several transfusions, a selective vascular embolization was performed when the patient was 3 weeks old, first using alcohol and then using microbeads. After this treatment, the bleeding quickly became less abundant, and the evolution of the tumor was favorable. The CH regressed, and the bleeding episodes stopped. The ulceration healed completely within 2 months. Propranolol treatment was stopped at 2 ½ months of age. By 1 year of age, although the CH continued to regress, a purplish swelling still persisted, with central lipoatrophy and a vascular mass seen on magnetic resonance imaging. The parents refused surgical excision of the remaining lesion.

Discussion
We report 2 neonatal cases of ulcerated CH that were complicated by life-threatening hemorrhagic episodes. While IHs are frequent vascular benign tumors in children, CHs are rare. RICH are violaceous tumors, firm to hard on palpation with telangiectasias on the surface, surrounded by a halo-like rim of pallor (Figure 4). The most frequently involved sites are the head and the limbs near a joint. Infantile fibrosarcomas may display similar clinical and radiologic features; therefore, a biopsy is recommended to confirm the diagnosis if there is any doubt.7,10,12 The histologic presentation of CH is characterized by relatively small lobules of capillaries surrounded by fibrous tissue (Figure 2). Endothelial cells from CH are negative for GLUT1 immunostaining.3 Seventy-five percent of CHs can be diagnosed using antenatal ultrasonography and can be detected from as early as 12 weeks’ gestation. However, diagnosis is more frequently made in the second trimester of pregnancy,12 as in our second case. In our first case, even though the tumor at birth was very large, it was not diagnosed during antenatal ultrasonography. This could be because these tumors grow extremely rapidly during the later stages of pregnancy. Although very large CHs may be complicated by cardiac failure, thrombocytopenia, or coagulopathy,13 these lesions are usually benign, and no treatment is recommended.

Ulceration is a common, predominantly benign complication in IH and bleeding seems to occur only rarely and most often is moderate.8,9 In contrast, little is known about the prognosis and the number of ulcerated CHs. Some case reports have suggested that ulcerated CH can lead to massive hemorrhages1,10,14,15 but the risk factors for this severe complication are unknown. Although some cases of life-threatening hemorrhages have also been described in ulcerated IH, especially in segmental IH, the data reported in the literature are sometimes too unclear to differentiate between true IH cases and misdiagnosed CH cases.14,15 Therefore, severe hemorrhages seem to be a rare complication more specifically observed in CH than in IH. Massive bleeding in ulcerated CH may be because of the erosion of the large superficial vessel that is associated with this tumor. As with IH, CHs are vascular tumors with a high blood flow. However, CH differs from IH based on the presence of the larger vessels that are seen in imaging test results.6 Furthermore, the vessels in CH are likely to be more superficial, making small cutaneous ulceration more likely to cause a severe hemorrhage. In our first case, as in previous reports,10,14,15 a large superficial vessel was eroded and resulted in high-flow hemorrhaging. It is unknown whether ulcerated CH localization or subtype could be a risk factor for severe hemorrhaging, but it is remarkable that our 2 cases and 2 of 3 other well-documented cases in the literature6,10 were of the RICH subtype, located on the thigh, and vascularized by the femoral fast-flow vessels. Therefore, Doppler ultrasonography, magnetic resonance imaging, or computed tomographic angiography should be discussed early in cases of ulcerated RICH to evaluate the
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size and depth of the vessels involved, as well as their distance to the ulcerated area.

Treatment of severe hemorrhages in ulcerated CH is similar to management of arteriovenous malformations and consists of selective vascular embolization and/or surgery.10 In our first case, embolization was initially performed because surgery was considered too risky.

Powell et al12 also propose to treat severe bleeding episodes caused by CH with tranexamic acid, an antifibrinolytic agent that can be used topically and helps to stabilize the clot.

In contrast to cases of IH,9 propranolol treatment is now known to be ineffective in reducing CH volume, as our 2 cases clearly illustrated. Furthermore, because propranolol can be harmful for hemodynamically unstable children, this treatment should be avoided in children with severe bleeding that can occur in ulcerated CH.

Conclusions

Although it is a rare complication, neonates with ulcerated CH are at risk for severe hemorrhaging. Close clinical monitoring is recommended for cases of CH with ulceration, even when the ulceration is small. Early Doppler ultrasonography, magnetic resonance imaging, or computed tomographic angiography should be used to visualize the subcutaneous vessels and assess the possible damage. Embolization should be the treatment of choice in case of severe bleeding as the natural history of RICH is to spontaneously regress.

REFERENCES