A 7-day-old Boy with a Congenital Vascular Nodule

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A 7-day-old boy infant presented for evaluation of a lesion on the trunk that was present at birth. The mother noted that the mass was warm but not painful when touched. Bleeding and ulceration were not noted. The patient was the 3.4 kg (50th percentile) product of a full-term, 38-week gestation birth and was born via spontaneous vaginal delivery. The pregnancy was uncomplicated, and there was no maternal history of preeclampsia or placental abnormalities. Vascular abnormalities were not seen on prenatal ultrasounds. Review of systems was unremarkable. There is no family history of vascular anomalies.

On physical exam, he was a well-developed, well-nourished infant with a 4.6 cm x 4.2 cm discrete, warm vascular nodule with a rim of pallor and central prominent telangiectasias on his trunk (see Figure 1). No bleeding or ulceration was noted. There was no pain on palpation. The remainder of his physical exam was unremarkable, including a normal cardiac and abdominal exam.

Follow-up evaluation of this child in 2 months revealed softening of the lesion, decreased erythema, and warmth. Further follow-up evaluation, which was 6 months after initial presentation, showed a flat, telangiectatic plaque with a rim of pallor (see Figure 2).

For diagnosis, see page 64.

Editor’s note: Each month, this department features a discussion of an unusual diagnosis in genetics, radiology, or dermatology. A description and images are presented, with the diagnosis and an explanation of how the diagnosis was determined following. As always, your comments are welcome.
They also noted other histologic differences between CH and IH.

RICH are characterized by a vascular tumor present at birth, usually located on the head, neck, or extremities. They are usually a violaceous color with overlying telangiectasias. After rapid involution in the first months to year of life, redundant skin and surface telangiectasias may be noted.

In contrast, a NICH does not involute and persists throughout childhood as a high-flow vascular lesion. The increased flow has been described by ultrasound and magnetic resonance imaging (MRI). Clinically, a NICH is an erythematous to violaceous, warm, vascular tumor with prominent overlying telangiectasias and is usually flatter in appearance than a RICH. A rim of pallor around the lesion can be seen in RICH and NICH, although it is more common in the latter. Often, it is difficult to differentiate a RICH from a NICH; some congenital hemangiomas seem to partially involute and are considered to be on the RICH to NICH spectrum.

Histopathologic evaluation of a RICH shows capillary lobules of variable size with moderately plump endothelial cells.

The differential diagnoses of a RICH include an IH, a kaposiform hemangioendothelioma (KHE), and a tufted angioma. As previously discussed, IH are very rarely fully formed at birth. A KHE usually presents as a purple to violaceous plaque or nodule that is firm and often rapidly expansile. Tufted angiomas usually do not manifest at birth but rather appear during infancy and childhood. Clinically, they present as slow growing papules or plaques. These latter two entities are associated with Kasabach-Merritt phenomenon, an entity with thrombocytopenia, microangiopathic hemolytic anemia, and a coagulopathy. Kasabach-Merritt phenomenon is not associated with congenital or infantile hemangiomas.

CONCLUSION

Most often, management of a RICH or a NICH is expectant with observation for rare complications of bleeding or ulceration. Although removal of excess skin after involution of a RICH may be indicated, a NICH may require complete excision later in life.

REFERENCES


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