

Vascular Mass on the Posterior Neck in a Newborn

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A newborn male was delivered via cesarean section at 38 weeks 5 days' gestation with a large vascular mass on the posterior neck. The mass previously had been identified on a 23-week prenatal ultrasound. Physical examination by dermatology at birth revealed a well-defined violaceous mass measuring 6×5 cm with prominent radiating veins, coarse telangiectases, and a pale rim. Magnetic resonance imaging demonstrated a well-circumscribed mass with avid arterial phase enhancement. The patient experienced transient thrombocytopenia that resolved following administration of methylprednisolone. No evidence of rapid involution was noted after 3 months of observation.

WHAT'S YOUR DIAGNOSIS?

- arteriovenous malformation
- congenital hemangioma
- infantile hemangioma
- kaposiform hemangioendothelioma
- tufted angioma

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THE DIAGNOSIS: Congenital Hemangioma

Surgical resection of the mass was performed at 4 months of age without complication (Figure 1). Histopathology revealed a lobular endothelial cell proliferation within a densely fibrotic stroma, multiple thin-walled vessels, and negative immunoreactivity to glucose transporter type 1 (GLUT-1) (Figures 2 and 3). Combined with the patient's clinical history and findings on imaging (Figure 4), the most accurate diagnosis was a congenital hemangioma (CH). The mass was determined to be a noninvoluting congenital hemangioma (NICH).

A variety of vascular anomalies manifest in newborns and can be differentiated by the patient's clinical history—particularly whether the lesion is present at birth or develops after birth. Imaging and histopathology of the lesion(s) may be utilized when clinical examination alone

is not sufficient to make a diagnosis. Histopathology and immunohistochemistry further aid in differentiating the type of vascular lesion.

Overall, vascular anomalies are classified broadly into 2 categories based on their pathogenesis: tumors and malformations. Vascular tumors are composed of proliferating endothelial cells that have the potential to resolve spontaneously over time. Examples include CH, infantile hemangioma (IH), kaposiform hemangioendothelioma (KHE), and tufted angioma (TA). In contrast, vascular malformations (ie, arteriovenous malformations) are composed of dysplastic vessels with normal endothelial cell turnover and do not resolve without intervention.¹

Congenital hemangiomas are rare vascular tumors that are fully developed at birth. These tumors proliferate in utero, enabling prenatal detection via ultrasonography as early as 12 weeks' gestation for large



FIGURE 1. A congenital hemangioma in a newborn was surgically resected without complication.

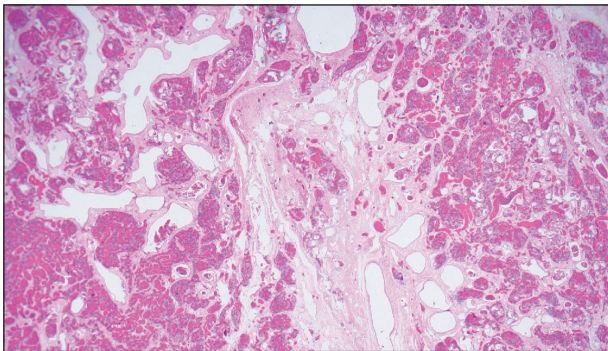


FIGURE 2. Histopathology of a congenital hemangioma revealed a lobular endothelial cell proliferation within a densely fibrotic stroma as well as multiple thin-walled vessels (H&E, original magnification $\times 200$).

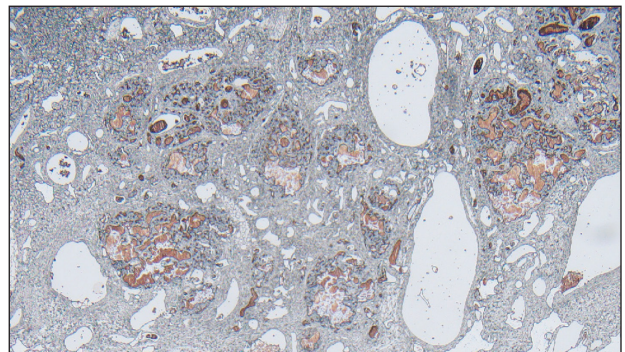


FIGURE 3. Immunohistochemistry of a congenital hemangioma demonstrated negative immunoreactivity to glucose transporter type 1 (GLUT-1).

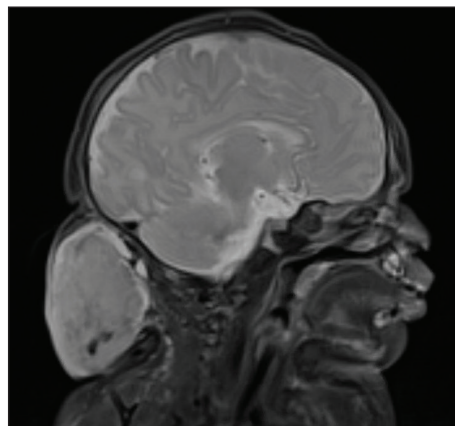


FIGURE 4. Magnetic resonance imaging of a congenital hemangioma demonstrated a well-circumscribed mass with avid arterial phase enhancement.

heterogeneous vascular masses.²⁻⁴ Congenital hemangiomas are described as solitary, well-circumscribed, raised, violaceous lesions most commonly located in the head and neck region.⁴⁻⁶ Histopathologically, they are characterized by lobules of proliferating capillaries surrounded by fibrous stroma and dysplastic vascular channels.^{6,7}

Congenital hemangiomas are categorized based on their postnatal involution patterns.² Fetally involuting CH both develops and begins regression in utero and often is completely regressed at birth.⁸ Rapidly involuting CH begins regression in the first few weeks of life and usually is completely involuted by 14 months of age.^{6,9-11} Conversely, NICH does not regress, often requiring surgical excision due to functional and cosmetic issues.^{12,13} Partially involuting CH is intermediary, beginning as rapidly involuting but not involuting completely and persisting as lesions that resemble NICH.¹⁴⁻¹⁶ Although generally benign and asymptomatic, these tumors can cause transient thrombocytopenia and coagulopathy at birth, as seen in our patient.^{17,18}

Infantile hemangioma is the most common vascular tumor of infancy.¹⁹⁻²¹ Although a precursor lesion may be present at birth, generally this tumor becomes apparent after the first few weeks of life as a solitary vascular plaque or nodule with a predilection for the head and neck.²²⁻²⁵ Once it arises, IH quickly enters a period of rapid growth, followed by a period of slower continued growth, with most reaching maximum size by 3 months.²² Thereafter, IH enters a slow period of involution (range, 3–9 years)²⁶; more recent data suggest near resolution by 5 years of age.²⁷ Infantile hemangioma is categorized based on its depth in the skin and subcutaneous tissues and can be classified as superficial, mixed, or deep.^{22,24,28,29} Superficial IH appears as a red plaque and may exhibit lobulation, while deep IH can be identified as flesh-colored or blue subcutaneous masses. Mixed IH may manifest with both superficial and deep features depending on the extent of its involvement in the dermal and subcutaneous layers. The pattern of involvement may be focal, segmental, or indeterminate.²⁴ In contrast, CH typically is a solitary vascular mass with prominent telangiectases, nodules, and radiating veins.⁶ Histologically, IH is composed of proliferative plump endothelial cells that form capillaries, and the lesion stains positively for GLUT-1, whereas CH does not.³⁰

Kaposiform hemangioendothelioma is classified as a locally aggressive vascular tumor that manifests either prenatally or in early infancy.³¹ It is described as a solitary, ill-defined, firm, purple plaque most commonly located on the extremities and retroperitoneum.³²⁻³⁴ Histopathologically, these lesions are characterized by dilated lymphatic channels and irregular sheets or lobules of spindle-shaped endothelial cells infiltrating the dermis and subcutaneous fat.^{33,35} In contrast to CH, KHE lesions show immunoreactivity to the markers podoplanin, lymphatic vessel endothelial receptor 1, and prospero homeobox 1 protein.^{36,37} Notably, 70% of these tumors are complicated by the presence of Kasabach-Merritt

phenomenon, a potentially life-threatening emergency that occurs when platelets are trapped within a vascular tumor, leading to the consumption of clotting factors, intralesional bleeding, and rapid enlargement of the tumor.³² The Kasabach-Merritt phenomenon manifests clinically as microangiopathic hemolytic anemia, severe thrombocytopenia, and disseminated intravascular coagulation.³⁸ Although CH lesions also can be associated with thrombocytopenia and coagulopathy, they generally are mild and self-limited.¹⁸

Tufted angioma is a vascular tumor that arises within the first 5 years of life as firm violaceous papules or plaques, often with associated hyperhidrosis or hypertrichosis.^{39,40} Although TA grows slowly for a period of time, it eventually stabilizes and persists, rarely regressing completely.⁴¹ These tumors share many similarities with KHE, and it has been suggested that they may be part of the same spectrum.⁴² As with KHE, TA lesions show immunoreactivity to the markers podoplanin, lymphatic vessel endothelial receptor 1, and prospero homeobox 1 protein, which are negative in CH.^{36,37} Although TA also can be complicated by Kasabach-Merritt phenomenon, the incidence is much lower (up to 38%).^{43,44} As such, TAs tend to be recognized as more superficial benign lesions. However, they still can cause notable cosmetic and functional impairment and should be monitored closely, especially in the presence of associated symptoms or complications.

Arteriovenous malformation is a vascular lesion that results from errors during the embryonic development of vascular channels.⁴⁵ Although present at birth, it may not become clinically apparent until later in life. Arteriovenous malformations enlarge postnatally, and their growth is proportional to the developmental growth of the affected individual rather than the result of endothelial proliferation.⁴⁶ In infants, AVM may manifest as a faint vascular stain that can evolve over time into a pink patch associated with a palpable thrill during adolescence.⁴ On Doppler flow imaging, AVMs are identified as fast-flow anomalies arising from an abnormal communication between high-pressure arterial systems and low-pressure venous systems without the presence of a capillary bed.⁴⁷ One of the differentiating factors between AVM and CH is that AVMs do not regress spontaneously and tend to have high recurrence rates, even with intervention.⁴⁸ In contrast, CH can be categorized based on its postnatal involution pattern. Another distinguishing factor is that AVMs tend to be larger and more invasive than CHs.⁴⁶ Therefore, early diagnosis and intervention are crucial to prevent complications such as bleeding, seizures, or neurologic deficits associated with AVMs.¹

REFERENCES

- Enjolras O, Wassef M, Chapot R. Introduction: ISSVA Classification. In: Enjolras O, Wassef M, Chapot R, eds. *Color Atlas of Vascular Tumors and Vascular Malformations*. Cambridge University Press; 2007:3-11.
- Fadell MF, Jones BV, Adams DM. Prenatal diagnosis and postnatal follow-up of rapidly involuting congenital hemangioma (RICH). *Pediatr Radiol*. 2011;41:1057-1060.

3. Feysin T, Khalek N, Moldenhauer JS. Fetal brain, head, and neck tumors: prenatal imaging and management. *Prenat Diagn.* 2020;40:1203-1219.
4. Foley LS, Kulungowski AM. Vascular anomalies in pediatrics. *Adv Pediatr.* 2015;62:227-255.
5. Bruder E, Alaggio R, Kozakewich HPW, et al. Vascular and perivascular lesions of skin and soft tissues in children and adolescents. *Pediatr Dev Pathol.* 2012;15:26-61.
6. Berenguer B, Mulliken JB, Enjolras O, et al. Rapidly involuting congenital hemangioma: clinical and histopathologic features. *Pediatr Dev Pathol.* 2003;6:495-510.
7. North PE, Waner M, James CA, et al. Congenital nonprogressive hemangioma: a distinct clinicopathologic entity unlike infantile hemangioma. *Arch Dermatol.* 2001;137:1607-1620.
8. Maguiness S, Uihlein LC, Liang MG, et al. Rapidly involuting congenital hemangioma with fetal involution. *Pediatr Dermatol.* 2015;32:321-326.
9. Keating LJ, Soares GM, Muratore CS. Rapidly involuting congenital hemangioma. *Med Health R I.* 2012;95:149-152.
10. Schafer F, Tapia M, Pinto C. Rapidly involuting congenital haemangioma. *Arch Dis Child Fetal Neonatal Ed.* 2014;99:F422.
11. Boon LM, Enjolras O, Mulliken JB. Congenital hemangioma: evidence of accelerated involution. *J Pediatr.* 1996;128:329-335.
12. Liang MG, Frieden IJ. Infantile and congenital hemangiomas. *Semin Pediatr Surg.* 2014;23:162-167.
13. Enjolras O, Mulliken JB, Boon LM, et al. Noninvoluting congenital hemangioma: a rare cutaneous vascular anomaly. *Plast Reconstr Surg.* 2001;107:1647-1654.
14. Nasser E, Piram M, McCuaig CC, et al. Partially involuting congenital hemangiomas: a report of 8 cases and review of the literature. *J Am Acad Dermatol.* 2014;70:75-79.
15. Wassef M, Blei F, Adams D, et al. Vascular anomalies classification: recommendations from the International Society for the Study of Vascular Anomalies. *Pediatrics.* 2015;136:E203-E214.
16. Boull C, Maguiness SM. Congenital hemangiomas. *Semin Cutan Med Surg.* 2016;35:124-127.
17. Drolet BA, Frommelt PC, Chamlin SL, et al. Initiation and use of propranolol for infantile hemangioma: report of a consensus conference. *Pediatrics.* 2013;131:128-140.
18. Baselga E, Cordisco MR, Garzon M, et al. Rapidly involuting congenital haemangioma associated with transient thrombocytopenia and coagulopathy: a case series. *Br J Dermatol.* 2008;158:1363-1370.
19. Kanada KN, Merin MR, Munden A, et al. A prospective study of cutaneous findings in newborns in the United States: correlation with race, ethnicity, and gestational status using updated classification and nomenclature. *J Pediatr.* 2012;161:240-245.
20. Munden A, Butschek R, Tom WL, et al. Prospective study of infantile haemangiomas: incidence, clinical characteristics and association with placental anomalies. *Br J Dermatol.* 2014;170:907-913.
21. Léauté-Labrèze C, Harper JI, Hoeger PH. Infantile haemangioma. *Lancet.* 2017;390:85-94.
22. Chang LC, Haggstrom AN, Drolet BA, et al. Growth characteristics of infantile hemangiomas: implications for management. *Pediatrics.* 2008;122:360-367.
23. Hidano A, Nakajima S. Earliest features of the strawberry mark in the newborn. *Br J Dermatol.* 1972;87:138-144.
24. Martinez-Perez D, Fein NA, Boon LM, et al. Not all hemangiomas look like strawberries: uncommon presentations of the most common tumor of infancy. *Pediatr Dermatol.* 1995;12:1-6.
25. Payne MM, Moyer F, Marcks KM, et al. The precursor to the hemangioma. *Plast Reconstr Surg.* 1966;38:64-67.
26. Bowers RE, Graham EA, Tomlinson KM. The natural history of the strawberry nevus. *Arch Dermatol.* 1960;82:667-680.
27. Couto RA, Maclellan RA, Zurakowski D, et al. Infantile hemangioma: clinical assessment of the involuting phase and implications for management. *Plast Reconstr Surg.* 2012;130:619-624.
28. Drolet BA, Esterly NB, Frieden IJ. Hemangiomas in children. *N Engl J Med.* 1999;341:173-181.
29. Chiller KG, Passaro D, Frieden IJ. Hemangiomas of infancy: clinical characteristics, morphologic subtypes, and their relationship to race, ethnicity, and sex. *Arch Dermatol.* 2002;138:1567-1576.
30. North PE, Waner M, Mizeracki A, et al. GLUT1: a newly discovered immunohistochemical marker for juvenile hemangiomas. *Hum Pathol.* 2000;31:11-22.
31. Gruman A, Liang MG, Mulliken JB, et al. Kaposiform hemangioendothelioma without Kasabach-Merritt phenomenon. *J Am Acad Dermatol.* 2005;52:616-622.
32. Croteau SE, Liang MG, Kozakewich HP, et al. Kaposiform hemangioendothelioma: atypical features and risks of Kasabach-Merritt phenomenon in 107 referrals. *J Pediatr.* 2013;162:142-147.
33. Zuberberg LR, Nickoloff BJ, Weiss SW. Kaposiform hemangioendothelioma of infancy and childhood. an aggressive neoplasm associated with Kasabach-Merritt syndrome and lymphangiomatosis. *Am J Surg Pathol.* 1993;17:321-328.
34. Mac-Moune Lai F, To KF, Choi PC, et al. Kaposiform hemangioendothelioma: five patients with cutaneous lesion and long follow-up. *Mod Pathol.* 2001;14:1087-1092.
35. O'Rafferty C, O'Regan GM, Irvine AD, et al. Recent advances in the pathobiology and management of Kasabach-Merritt phenomenon. *Br J Haematol.* 2015;171:38-51.
36. Le Huu AR, Jokinen CH, Rubin BP, et al. Expression of prox1, lymphatic endothelial nuclear transcription factor, in kaposiform hemangioendothelioma and tufted angioma. *Am J Surg Pathol.* 2010;34:1563-1573.
37. Debelenko LV, Perez-Atayde AR, Mulliken JB, et al. D2-40 immuno-histochemical analysis of pediatric vascular tumors reveals positivity in kaposiform hemangioendothelioma. *Mod Pathol.* 2005;18:1454-1460.
38. Haisley-Royster C, Enjolras O, Frieden IJ, et al. Kasabach-Merritt phenomenon: a retrospective study of treatment with vincristine. *J Pediatr Hematol Oncol.* 2002;24:459-462.
39. Wilmer A, Kaatz M, Bocker T, et al. Tufted angioma. *Eur J Dermatol.* 1999;9:51-53.
40. Herron MD, Coffin CM, Vanderhoof SL. Tufted angiomas: variability of the clinical morphology. *Pediatr Dermatol.* 2002;19:394-401.
41. North PE. Pediatric vascular tumors and malformations. *Surg Pathol Clin.* 2010;3:455-494.
42. Chu CY, Hsiao CH, Chiu HC. Transformation between kaposiform hemangioendothelioma and tufted angioma. *Dermatology.* 2003;206:334-337.
43. Osio A, Fraïtag S, Hadj-Rabia S, et al. Clinical spectrum of tufted angiomas in childhood: a report of 13 cases and a review of the literature. *Arch Dermatol.* 2010;146:758-763.
44. Johnson EF, Davis DM, Tollefson MM, et al. Vascular tumors in infants: case report and review of clinical, histopathologic, and immunohistochemical characteristics of infantile hemangioma, pyogenic granuloma, noninvoluting congenital hemangioma, tufted angioma, and kaposiform hemangioendothelioma. *Am J Dermatopathol.* 2018;40:231-239.
45. Christison-Lagay ER, Fishman SJ. Vascular anomalies. *Surg Clin North Am.* 2006;86:393-425.
46. Liu AS, Mulliken JB, Zurakowski D, et al. Extracranial arteriovenous malformations: natural progression and recurrence after treatment. *Plast Reconstr Surg.* 2010;125:1185-1194.
47. Young AE, Mulliken JB. Arteriovenous malformations. In: Mulliken JB, Young AE, eds. *Vascular Birthmarks: Haemangiomas and Malformations.* WB Saunders; 1988:228-245.
48. Duggan EM, Fishman SJ. Vascular anomalies. In: Holcomb GW III, Murphy JP, St Peter SD, eds. *Holcomb and Ashcraft's Pediatric Surgery.* 7th edition. Elsevier; 2019:1147-1170.