SKIN

SKINmages

Multiple Congenital Hemangiomas

Christopher Chino-Marin*, BA[1], Kristina M. Derrick*, MD, ScM[1,2], John McGovern, MD[1,2], Sharon A. Glick, MD, MS[1,2]

[1] The State University of New York Downstate School of Medicine, Department of Dermatology, Brooklyn, NY, USA
[2] Maimonides Medical Center, Department of Dermatology, Brooklyn, NY, USA
* Mr. Chino-Marin and Dr. Derrick are considered joint first authors

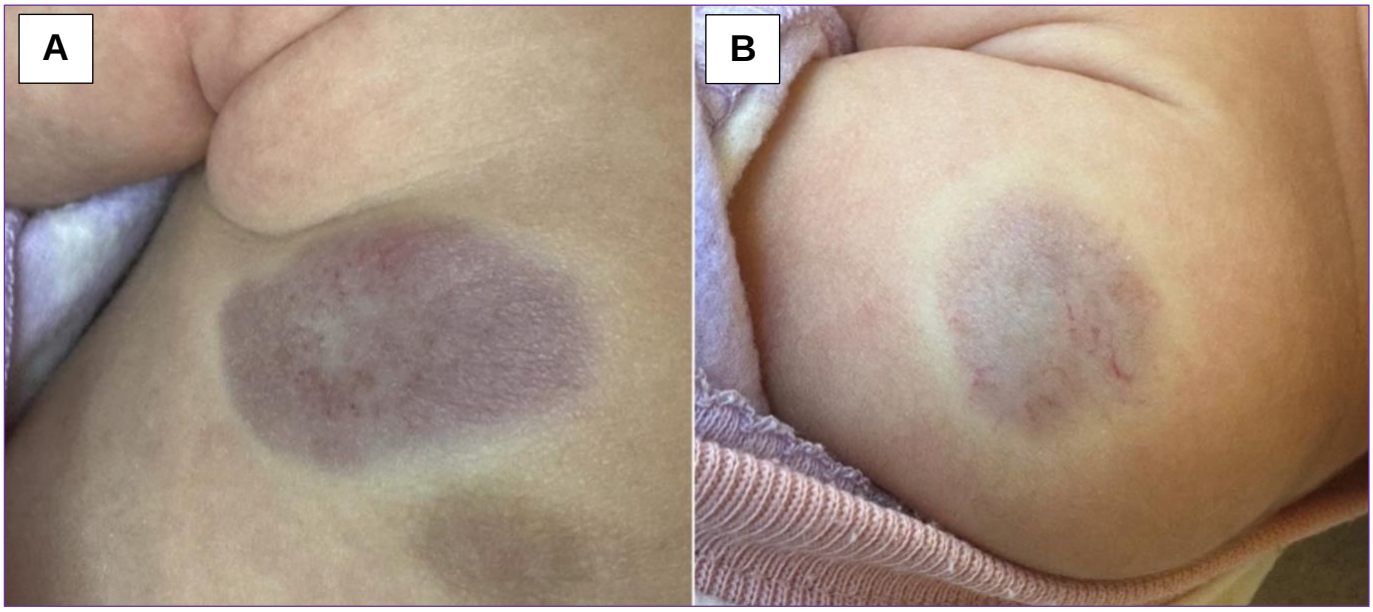

Figure 1. Right chest with 5 cm x 3 cm blue subcutaneous nodule with peripheral pallor (A). Left shoulder with 3 cm x 3 cm blue nodule with prominent telangiectasias and rim of peripheral pallor (B).

A 1-month-old ex-term male presented with two blue lesions, which were unchanged since birth. On physical examination, the right chest (**Figure 1A**) had a 5 cm x 3 cm blue subcutaneous vascular nodule with peripheral circumferential pallor, with a similar 3 cm x 3 cm lesion on the left shoulder (**Figure 1B**). Several dark red-purple telangiectasias were visible within the lesions, which were highlighted using Dermoscopy, along with well-circumscribed blue subcutaneous nodules, peripheral circumferential pallor, and central telangiectasias (**Figure 2**). The remainder of the examination was normal. The lesions exhibited telangiectasias and homogeneous, blue-purple blanches, so duplex ultrasound and biopsy were not needed. Kaposiform Hemangioendotheliomas (KHE) are ruled out due to the absence of Kasabach-Merrit phenomenon, infiltrative growth, and poorly circumscribed lesions; sarcomas and teratomas are ruled out as they typically have irregular borders, rapid growth, and heterogenous features; deep infantile hemangiomas (IH) are ruled out because

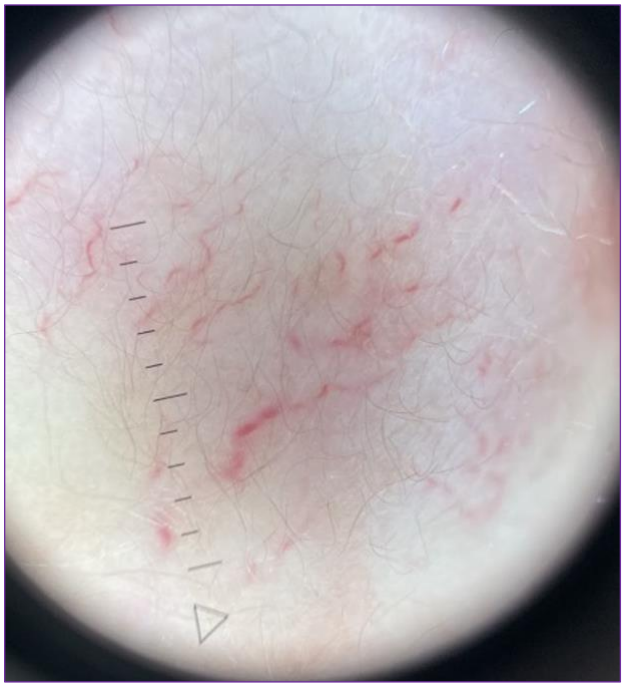

Figure 2. Dermoscopy of left shoulder lesion shows telangiectasias.

they typically appear postnatally and proliferate; a clinical diagnosis of multiple congenital hemangiomas (CHs) was made.

CHs are benign vascular tumors that are fully formed at birth. They present as well-demarcated dome-like or subcutaneous nodules with blue-purple pigmentation that may include a white halo around the entire lesion[1,2]. In contrast, infantile hemangiomas (IHs) are usually absent at birth, then rapidly grow at 1-2 months with slower growth over 1 year then involute. IHs are also responsive to β-blockers (propranolol, atenolol, timolol), but CHs are not. CHs are usually solitary. The first histologically-confirmed case of multifocal, miliary CHs was described by Funk *et al.*[3], followed by Blumenthal ***et al.'s*** report of two additional cases with a similar phenotype[4]. Our case is unique in having two large lesions but not the small miliary pattern of the prior reports[3,4].

Clinical history and examination are typically sufficient for diagnosis. Differential diagnoses include tufted angioma (TA), Kaposiform Hemangioendothelioma (KHE), sarcoma, teratoma, or deep IH[5]. Skin biopsy is diagnostic, as IHs are GLUT1-positive on immunohistochemical staining while CHs are GLUT1-negative. CHs are associated with *GNAQ* and *GNA11* gene variants and rarely *PIK3CA*, but lesion genetic testing is not routinely performed[5]. Duplex ultrasound of CH shows dense vascularity, high blood flow velocity, and occasionally calcifications[1,5].

CHs have a favorable prognosis, typically involuting over time without intervention and are not generally associated with significant risk if involution does not occur. Three subtypes exist: rapidly involuting CH (RICH) the most common and likely subtype, partially involuting CH (PICH), and non-involuting CH (NICH). Within weeks-months, it will become apparent if the lesion is a RICH. In our patient, the lesion is most likely PICH or NICH, but confirmation will be apparent after one year of follow-up. Both RICH and PICH have a risk of ulceration[5]. A rare complication of ulceration in RICH and PICH is consumptive coagulopathy with elevated D-

dimer and thrombocytopenia, known as Kasabach-Merritt phenomenon[5]. This phenomenon occurs in up to 70% of KHE, is far less common in tufted angiomas[6], and is only rarely reported in CHs[7]. It is important to check a complete blood count and coagulation tests (PT/INR, PTT, D-dimer, fibrinogen) in patients with involuting or ulcerating RICH/PICH[5]. Very large lesions or those associated with high output cardiac failure may require surgical excision, but most CH lesions should be treated with watchful waiting as they may involute over the first year[5]. If the CH does not involute after the first year, elective surgery can be considered for removal or cosmetic improvement.

If lesions ulcerate, wound care should include keeping the lesion covered with a barrier ointment such as petrolatum (Vaseline) and non-stick dressing. After partial or full involution of lesions, surgery may be performed to remove scar and fibrofatty tissue. Vascular lasers may be considered for treatment. Unlike IHs, propranolol and timolol drops are not effective, so the only treatment is time or surgery/laser[5]. Referrals to pediatric dermatology or a vascular anomalies center should be considered.

Conflict of Interest Disclosures: None

Funding: None

Corresponding Author:
Kristina M. Derrick, MD, ScM
450 Clarkson Ave, Box 46
Brooklyn, NY 11203
Phone: (718) 270-1229
Email: kristina.derrick@downstate.edu

References:

1. Braun V, Prey S, Gurioli C, et al. Congenital haemangiomas: a single-centre retrospective review. *BMJ Paediatr Open*. 2020;4(1):e000816. doi:10.1136/bmjpo-2020-000816
2. Wassef M, Blei F, Adams D, et al. Vascular Anomalies Classification: Recommendations From the International Society for the Study of Vascular Anomalies. *Pediatrics (Evanston)*. 2015;136(1):E203-E214. doi:10.1542/peds.2014-3673
3. Funk T, Lim Y, Kulungowski AM, et al. Symptomatic Congenital Hemangioma and Congenital Hemangiomatosis Associated With a Somatic Activating Mutation in GNA11. *JAMA Dermatology*. 2016;152(9):1015-1020. doi:10.1001/jamadermatol.2016.2365
4. Blumenthal S, Stefanko N, Cossio ML, et al. Multifocal congenital hemangioma: Expanding the pathogenesis of "neonatal hemangiomatosis". *Pediatr Dermatol*. Sep 2019;36(5):720-722. doi:10.1111/pde.13814
5. Bolognia J, Schaffer JV, Cerroni L. *Dermatology*. Fourth edition. ed. Elsevier; 2018.
6. Ji Y, Chen S, Yang K, Xia C, Li L. Kaposiform hemangioendothelioma: current knowledge and future perspectives. Orphanet J Rare Dis. 2020;15(1). doi:10.1186/s13023-020-1320-1
7. Peng YH, Lin MC, Liu WL, Jan SL. Case report: Transcatheter arterial embolization in a newborn with cervical rapidly involuting congenital hemangioma and Kasabach-Merritt phenomenon. *Front Pediatr*. 2023;11:1073090. Published 2023 Feb 24. doi:10.3389/fped.2023.1073090