

Diffuse Capillary Malformation With Undergrowth of a Limb in a Boy

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PRACTICE POINTS

- The term *diffuse capillary malformation with undergrowth* (DCMU) describes a distinct counterpart to diffuse capillary malformation with overgrowth. It can be challenging to distinguish from other vascular malformations associated with congenital structural abnormalities.
- The vascular patterning of DCMU may fade over time, but patients should continue to be monitored for their structural incongruity.

To the Editor:

Capillary malformations (CMs), the most common vascular malformations that can affect the skin,¹ present clinically as macules and patches of various colors, shapes, and sizes. Congenital structural abnormalities are associated with conditions such as Klippel-Trenaunay syndrome (KTS), cutis marmorata telangiectatica congenita (CMTC), and megalencephaly–capillary malformation syndrome.² Diffuse CM with overgrowth (DCMO) of the soft tissue and bones is an established association of CMs; however, *diffuse capillary malformation with undergrowth* (DCMU) is a more recent term that describes the lesser-recognized counterpart to DCMO.³ Herein, we describe a case of CM with left-sided undergrowth.

An 11-year-old boy presented to our clinic with asymptomatic vascular patterning on the left side of the body that had been present since birth. He previously

was diagnosed with congenital right hemihypertrophy. He reported that the areas gradually lightened over time, and he denied any history of ulceration or venous or lymphatic malformations. Additionally, he explained how the left arm and leg have been noticeably smaller than the right extremities throughout his life. Physical examination revealed superficial, violaceous, reticulated patches along the left upper back tracking down the arm, abdomen (Figure 1A), and anterior thigh (Figure 1B) without crossing the midline. A few dilated veins were noted in the same region as the patches. There was no evidence of scarring or depression found in the skin. The right arms and legs were visibly larger compared to the left side (Figure 2A), and there also was macrodactyly of the third digit of the left hand (Figure 2B). Radiography confirmed the limb length discrepancy and showed the right and left legs to measure 73.2 cm and 71.3 cm, respectively. Given the patient's multifocal reticulated CMs and ipsilateral undergrowth, a diagnosis of DCMU was rendered. The superficial vascular pattern is likely to fade over time, which will partially be hidden by his darker complexion. He also was advised to continue to see an orthopedist to monitor the limb length incongruity. Surgical intervention was not recommended.

It ordinarily is thought that vascular anomalies of a limb may result in hypertrophy due to increased blood flow such as in KTS, but there are occasions where the affected limb(s) are inexplicably smaller.^{2,4} Cubiró et al³ observed that in 6 patients with unilateral CMs, all had ipsilateral limb undergrowth. They proposed the term *diffuse capillary malformation with undergrowth* as a distinct counterpart to DCMO. Diffuse capillary

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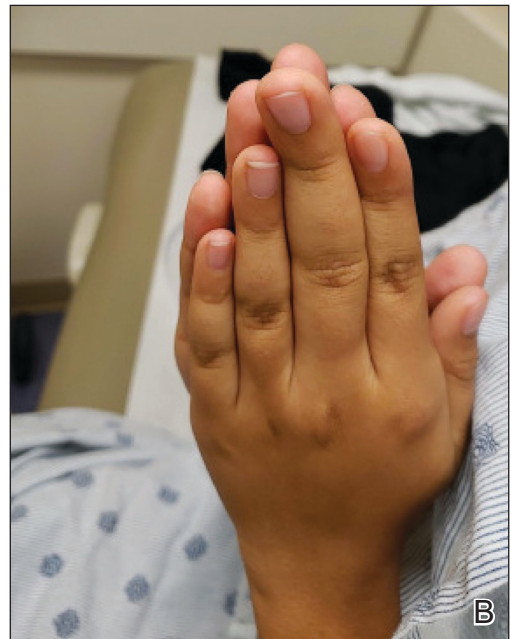
The authors report no conflict of interest.

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FIGURE 1. A and B, Reticulated violaceous patches on the left abdomen and left anterior thigh, respectively.



FIGURE 2. A and B, Hypotrophy of the left arm and hand as well as macrodactyly of the left third digit, respectively.



malformation with undergrowth is most similar to CMTC, as both can present with patchy or reticulated capillary staining with ipsilateral limb hypotrophy, but girth more often is affected than length; CMTC also may be associated with dermal atrophy and ulceration.² The lesions of CMTC typically diminish within the first few years of life whereas those in DCMU tend to persist. Patients with KTS also can exhibit soft-tissue and bony undergrowth, which is termed *inverse Klippel-Trenaunay syndrome*³; however, the lack of the triad of capillary-lymphatic-venous malformation in our patient made this condition less likely. Additionally, it appears that

our patient had left-sided undergrowth rather than the previously diagnosed right hemihypertrophy. The ipsilateral macrodactyly of the third digit of the left hand was an interesting observation and contrasted the undergrowth apparent in the rest of the left limb, which could be caused by increased blood flow specifically to the third digit resembling DCMO.⁴

Of note, genetic mutations have been implicated as a cause of vascular malformations and growth abnormalities. Specifically, mutations in the phosphoinositide-3-kinase-AKT pathway have been reported in these cases likely due its role in cell growth, proliferation, and

angiogenesis.^{3,4} Future studies should investigate genetic associations in patients with DCMU to determine if there is a robust genotypic-phenotypic link.

Although CMs are a common occurrence in pediatric dermatology, CMs with concurrent limb undergrowth are rare. Our patient's unique features included involvement of both an arm and leg as well as the presence of macrodactyly. We agree with the terminology for DCMU to describe multifocal reticulated vascular patterning with ipsilateral undergrowth.³

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