CD34+ connective tissue nevi: Are they unusual?

To the Editor: I read with interest the article by Shah, Wells, and Stetson1 (“CD34+ connective tissue nevi: Are they unusual?”). The authors present a case of CD34+ connective tissue nevus (CTN) in a 1-year-old boy and describe 6 additional cases found in their records which affected children from 4 months to 13 years of age. They conclude that CD34+ CTN is probably not rare. I fully agree with them on this point since I personally have seen 7 cases of CD34+ CTN in children. I also agree that some cases probably remain undiagnosed; however, I am surprised that they were unable to find any other such cases published in the literature, despite their Web-based literature search.

They seem to have missed a number of articles published between 1995 and 20092-7 that described “dermal dendrocyte hamartoma,” which is composed of CD34+ spindle-shaped cells. Whereas in some of these cases the lesion presented as a nodule, the majority of patients were found to have “medallion-like” infiltrated plaques. In our article published in 2009, we pointed out that the main diagnostic pitfall in these medallion-like congenital cases is congenital atrophic dermatofibrosarcoma.5 Kutzner et al7 recently published a series of 5 medallion-like dermal dendrocyte hamartomas which they preferred to re-name “plaque-like CD34-positive dermal fibroma” and emphasized the usefulness of fluorescence in situ hybridization analysis for the accurate distinction of dermal dendrocyte hamartoma from dermatofibrosarcoma protubersans.

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REFERENCES

Reply

To the Editor: We appreciate Dr Fraitag’s interest and comments about our manuscript. Dr Fraitag mentions that medallion-like dermal dendrocyte hamartomas should have been included in our review. Our manuscript was accepted in October 2008. Only 3 of the articles mentioned were published before mid-2008,1-3 which is when data for this article were collected. This very rarely reported entity is described as erythematous atrophic wrinkled medallion-shaped patches or nodules resembling anetoderma (macular atrophy), aplasia cutis, and atrophoderma.1-3 Shah et al3 reported that there were only 3 occurrences of this entity reported before their paper was published in 2007. Because of this, many might not include this entity as a connective tissue nevus (CTN). Our cases did not resemble nor do we believe any of our cases represented this entity. A CTN is composed of excessive amounts of one or more dermal components.4 Most commonly we use this term synonymously with collagenoma, but recognize that there are many forms of CTN. Our cases all represent the more classic collagen type (collagenoma). Because of this, medallion-like dermal dendrocyte hamartomas are likely a subtype of CTN, but the first 3 articles do not specifically categorize this entity as such. Internet-based review of the literature
Clas tic vasculitis and is second- and third-line disorders, including Behc¸et’s disease and leukocyto-

also first-line treatment for a number of dermatologic conditions, including psoriasis, pyoderma gangrenosum, relapsing polychondritis, scleroderma, and Sweet’s syndrome. It is often an alternative to dapsone for patients intolerant of dapsone in the treatment of neutrophilic dermatoses.

Colchicine was one of the “unapproved” drugs targeted by the Food and Drug Administration (FDA) for safety and efficacy while “avoiding undue burdens on consumers”. URL Pharma was the only manufacturer to complete the testing required to meet FDA approval of colchicine. The FDA approved the URL Pharma version of colchicine (Colcrys) for the treatment of familial Mediterranean fever and gout and granted the manufacturer 3 years of market exclusivity. During its application process, it was determined that colchicine toxicity was potentially exacerbated by drugs that inhibit cytochrome P450 3A4 (CYP3A4), an enzyme involved in the metabolism of colchicine, and p-glycoprotein, a protein involved in its oral absorption. Drugs that inhibit CYP3A4 and p-glycoprotein include clarithromycin, erythromycin, cyclosporine, verapamil, diltiazem, ketoconazole, and itraconazole. Sixty of the 117 reported deaths (51%) in patients receiving colchicine were concomitantly using clarithromycin.

Generic drugs are those evaluated and approved by the FDA to demonstrate bioequivalence to a brand name reference product. The FDA was aware of 21 firms that manufactured and distributed oral colchicine before Colcrys. These colchicine products were not evaluated and approved by the FDA, which considers these products unapproved drugs, not generics of Colcrys. URL Pharma brought a lawsuit seeking to remove other versions of colchicine from the market and raised the price of a tablet from $0.09 to $4.85 per pill.

Removal of unapproved colchicine increases medication cost from approximately $6 per month to $300 per month for dermatologic disorders treated with colchicine. URL Pharma has both a patient assistance program for the uninsured and a copay coupon program to help eligible patients. This program does not apply to patients in Massachusetts or prescriptions paid in whole or in part by state or federal programs, such as Medicare part B or Medicaid.

Nearly all patients in the United States will face an increase in price for colchicine, and some patients may need to seek alternate treatments even with patient assistance programs. Dermatology patients treated with colchicine will benefit from updated data on drug interactions with colchicine but may not benefit proportionately from

Law of unintended consequences

To the Editor: Colchicine has been used for more than 200 years as an inexpensive treatment for gout. The tablet form has been widely available in the United States since the 19th century. Colchicine is also first-line treatment for a number of dermatologic disorders, including Behc¸et’s disease and leukocytoclastic vasculitis and is second- and third-line treatment for other dermatologic disorders, such as aphthous stomatitis, calcinosis cutis, dermatitis herpetiformis, epidermolysis bullosa acquisita, erythema nodosum, linear IgA bullous dermatosis, palmoplantar pustulosis, psoriasis, pyoderma gangrenosum, relapsing polychondritis, scleroderma, and Sweet’s syndrome. It is often an alternative to dapsone for patients intolerant of dapsone in the treatment of neutrophilic dermatoses.

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