Coexistent Discoid Lupus Erythematosus and Porphyria Cutanea Tarda

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Bullae had recently appeared on the dorsa of the hands and fingers of a 46-year-old man. His medical history was significant for alcoholism, discoid lupus erythematosus (DLE), and hepatitis C. Systemic lupus erythematosus (SLE) was ruled out; the patient’s symptoms did not meet the American Rheumatism Association criteria for SLE.

A few weeks earlier, hydroxychloroquine, 200 mg/d, had been prescribed for DLE. Several days after the medication was started, the patient’s urine turned pink. Pink urine, which indicates elevated levels of urinary porphyrins, is a sign of porphyria cutanea tarda (PCT). Multiple dull red macules with adherent scales and atrophic, depigmented, scarred patches typical of DLE lesions were present on the patient’s scalp, face, back, and arms. Large bullae filled with clear fluid were noted on the normal dorsal skin of the hands and fingers. Drs Scott L. Goffin and Yelva Lynfield of Cedarhurst, NY, identified these lesions as PCT; the clinical diagnosis was confirmed by the finding of high levels of porphyrins in the patient’s serum and stool. The hydroxychloroquine was discontinued; oral prednisone, 60 mg/d, was prescribed. All of the bullous lesions healed. The dosage of systemic corticosteroids was tapered and then discontinued. The patient’s DLE lesions were treated with mid-potency topical corticosteroids. A cosmetic cover cream was recommended to mask the facial lesions. Six months after his initial presentation, DLE lesions recurred on the patient’s face (A) and forehead (B). The lesions extended over the patient’s arms (C); this widespread distribution is characteristic of disseminated DLE. The only remaining evidence of the PCT were the small erosions with hemorrhagic crusts and small scars on the hands (D). The coexistence of lupus erythematosus (LE) and PCT was first reported by Wolfram in 1952; since then, more than 35 cases of coexistent DLE or SLE and PCT have been described in the literature. Is this association coincidental, or are the diseases related? Hypotheses to explain their linkage include:

- The disorders stem from a common genetic abnormality.
- Porphyria triggers an autoimmune response (LE).
- Preexisting LE results in an acquired metabolic fault that leads to porphyria.
- LE precipitates a genetically determined metabolic fault that results in PCT.

We believe that our patient had the latent metabolic defect of PCT from alcoholism and hepatitis C, which manifested clinically when he was given hydroxychloroquine. The porphyrias are characterized by an increase in the intermediate metabolites of hemoglobin synthesis; PCT is the most common form. Each porphyria is associated with an enzyme deficiency in the metabolic pathway of heme synthesis. Cutaneous manifestations of PCT include vesicles, especially on the sunexposed skin of the hands, forearms, ears, and face. These lesions are not surrounded by erythema; they rupture easily to form erosions or shallow ulcers. Skin fragility, hyperpigmentation, facial hypertrichosis, and
sclerodermatous thickening may be seen. Both PCT and LE are exacerbated by sunlight. However, photosensitivity in LE is attributable to UV-B rays, whereas longer wavelengths—specifically the Soret band (UV-A 400 to 410 nm)—are responsible for PCT flares. Caution patients with PCT to avoid sun exposure and to use broad-spectrum sunblocks, such as titanium dioxide and zinc oxide. Sunscreens with high SPF content—which is a measure of protection against UV-B light—may not protect against the long-wave UV-A rays. Other recognized precipitants of PCT include alcohol; drugs, such as estrogens and antimalarials; polychlorinated hydrocarbons; disturbances of iron metabolism; and HIV and hepatitis C virus infections. Since antimalarials trigger PCT, only very low-dose hydroxychloroquine or chloroquine can be used to treat LE in persons with both diseases. These agents liberate the excess porphyrin from the liver; the porphyrin is then excreted in the urine. Because of this patient’s continued alcohol use and history of noncompliance, antimalarial therapy—which requires careful monitoring—was not appropriate. Topical corticosteroids and sunblock creams were prescribed for his skin lesions.

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