

Porphyria Cutanea Tarda Masquerading as Chronic Hand Eczema

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Sir,

Porphyria cutanea tarda (PCT) affects sun-exposed areas of the skin, and therefore commonly the dorsa of the hands, resulting in skin fragility and atrophic scarring with milia formation. Unless the possibility of this diagnosis is considered, the necessary investigations will not be performed, leading to a delay in diagnosis and treatment. For the patient, this potentially means loss of time at work, further scarring of the skin and liver damage.

Within 2 weeks in March 2000, we saw 2 patients with PCT in the contact dermatitis clinic. Both were referred from dermatologists with a clinical diagnosis of chronic hand eczema.

CASE REPORTS

Case 1

A 42-year-old self-employed bricklayer had noticed a rash on the dorsa of both hands in the summer, which improved during the winter months, only to recur the following year. The skin on his hands was erythematous and scaly with blistering. A course of flucloxacillin had resulted in some improvement. His skin was dry, but there was no personal or family history of atopy, nor any other past medical history of note. His only medication was occasional paracetamol. He consumed an average of 60 units of beer a week.

Because of a diagnosis of hand eczema, betamethasone valerate and fusidic acid cream were prescribed, without improvement. He was therefore referred for patch testing to exclude an occupational contact dermatitis. On examination the presence of milia was noted (Fig. 1).

Patch tests to a standard and medicament series did not reveal any positive reactions. Abnormal results of laboratory investigations confirmed the diagnosis of PCT: ALT 205 IU/l (<35), Ferritin 408 g/l (<220), plasma porphyrins 238 nmol/l (<11.2), urine uroporphyrin I 228.5 nmol/l/nmol creatinine (<3.31), urine uroporphyrin II 128.3 nmol/l/nmol creatinine (<0.67) and total stool porphyrins 324.4 nmol/g dry weight. A liver biopsy showed alcoholic steato-hepatitis with no signs of haemochromatosis or alpha-1-antitrypsin deficiency. The patient had antibody-evidence of past hepatitis B infection and was found to be hepatitis C-positive, also on PCR. Phlebotomy could not be undertaken because of the risk of infection. However, after 2 years he eventually managed to stop all alcohol intake. Within 2 months, he had no further



Fig. 1. The dorsum of the hand of Case 1 showing a milium, erosions and atrophic scarring with pigment disturbance.

blisters or skin problems despite continuing in his job. He remained under follow-up because of his active hepatitis C.

Case 2

A 47-year-old roofer was seen with an 18-months' history of a sometimes blistering eruption on the dorsa of his hands, which was worse during the summer and had started during an episode of stressful life events. He had used diesel to wash his hands for many years. There was no significant past medical history or personal history of atopy, but his daughter suffered from asthma. The patient stated that he recently had reduced his alcohol intake to 70 units of beer per week. A diagnosis of irritant hand eczema was made, but he was referred for patch testing to exclude an allergic element.

On examination he showed atrophic scars on the dorsum of both hands with mottled pigmentation and milia formation on the background of generally hyperpigmented skin. Patch tests to a standard and medicament series were negative. PCT was diagnosed following blood and urine investigation (same as for case 1). He tested negative for hepatitis C. Following the venesection of 12 units of blood, the blisters on the dorsum of his hands disappeared, which was mirrored by biochemical improvement. He was subsequently lost to follow-up.

DISCUSSION

Current textbooks list irritant and allergic contact dermatitis, tinea manuum, psoriasis and atopic eczema in

the differential diagnosis for chronic hand eczema, but PCT goes unmentioned (1–3).

The clinical features of cutaneous porphyria include vesicles and bullae on sun-exposed sites such as the dorsa of the hands. Crusting is followed by healing, which may take weeks and results in atrophic scarring with milia formation. Milia are small, sub-epidermal keratin cysts which appear as tiny, white, millet seed-like papules. Pigment disturbance, most commonly hyperpigmentation, may also occur. Patients may be unaware of the seasonal exacerbation of their condition.

Patients with PCT show an inherited autosomal dominant (20% of patients) or acquired (80%) reduction of the hepatic uroporphyrin decarboxylase activity (4). PCT is the most common porphyria; it occurs worldwide without racial predilection and a mean age of onset of 47 years (5). Variegate porphyria presents with clinically indistinguishable cutaneous signs, but additional systemic symptoms.

Skin fragility with blistering followed by milia is a result of a subepidermal split below the basement membrane. Apart from porphyria, other acquired causes for this include pseudoporphyria due to renal failure or drugs, bullous amyloid and epidermolysis bullosa acquisita (6).

Acquired or sporadic PCT is commonly precipitated by environmental factors such as for example oestrogens or, as in our patients, alcohol and viral hepatitis, which may unmask an underlying enzyme defect. Excess alcohol intake is found in most males with PCT and liver cirrhosis develops in up to one-third of cases (7). Plasma iron is raised in up to 50% of patients (7). On diagnosis of PCT, hepatitis C should be excluded, for which there is a marked regional variation. In Northern Europe, 17% of the patients with PCT were found to be hepatitis C-positive, but studies have described up to 94% (8).

Apart from stopping the exogenous precipitants, treatment options for PCT include removal of iron by phlebotomy or low-dose chloroquine (9).

Our observation of 2 patients with PCT within a 2-week period in our patch-testing clinic was an unusual cluster, but patients intermittently present to our tertiary referral unit in this fashion. Both patients regularly consumed alcohol well in excess of the recommended limit of 21 units a week (10), and one was additionally hepatitis C-positive. They were both in professions with a high risk of irritant and allergic contact dermatitis; in bricklayers, 16.1 cases of hand

eczema, and in builders, 13.5 cases, were reported by Dermatologists and Occupational Physicians per 100,000 workers in the UK (11). However, all their skin problems resolved following biochemical improvement of their PCT, despite continuing in their jobs.

The skin of the hands is readily examined by any dermatologist. We suggest that cutaneous porphyria, in particular PCT, should be added to the list of differential diagnoses for hand eczema. The presence of milia should prompt re-evaluation of the clinical diagnosis of dermatitis. Blood, urine and stool samples should be sent to a biochemical laboratory in light-shielded containers for a porphyrin screen, in order not to miss this important diagnosis.

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