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March 2013

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Why is my skin so fragile?

By Elizabeth Satter MD; and
Catherine Maari, MD, FRCPC

Mr. Young, a 67-year-old homeless male, was admitted to the hospital for a course of intravenous antibiotic for severe pneumonia.

In this article:

Mr. Young's Medical History

- Alcoholism (drinks approximately six to eight beers a day).
- Hepatitis C.
- Chronic obstructive pulmonary disease.
- Depression.
- Not taking any medication.

Mr. Young's Stats

- Multiple, small 3 mm to 4 mm, erosions and scars on the dorsum of his hands and face, mostly in areas of sun exposure (Figure 1).
- During the summer, his skin is very fragile and he always gets small blisters on his hands.
- Vesicles were not seen during his admission.
- A few small white papules consistent with milia were also seen on the dorsum of his hands and face.
- Significant increased facial hair growth was noted on the upper cheeks and eyebrows.

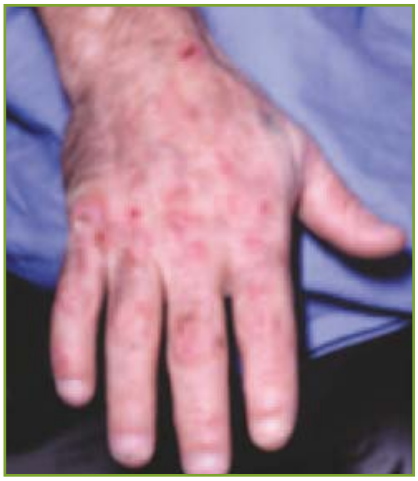

Figure 1. Small erosions, scars and milia can be seen on the dorsum of the hands.

What's your diagnosis?

A. Epidermolysis bullosa acquisita
B. Polymorphous light eruption
C. Porphyria cutana tarda
D. Poison ivy
E. Systemic lupus erythematosus

Answer: Porphyria Cutanea Tarda

The term "porphyria" originates from the Greek term prophyreos, which means "purple" to reflect the burgundy/red colour that can be seen in the urine of some of the patients with porphyrias. Clinically, the patient presents with cutaneous findings associated with excessive photosensitivity and skin fragility, and acute findings. These are reflected by neurologic or gastrointestinal symptoms, or a combination of both.

What is PCT?

Porphyria cutanea tarda (PCT) is the most common of all the porphyrias and occurs as a result of a deficiency of uroporphyrinogen decarboxylase. Although PCT can occur as an autosomal dominant disorder, 80% to 90% of cases are acquired sporadically (Table 1). The inherited form of PCT, the enzymatic defect, is seen universally in all tissues, whereas the acquired form is localised in the liver. Most patients present at the average age of 45, and before oral contraceptives were introduced, most patients were males. Liver disease as a result of alcoholism, hepatitis C, or hemochromatosis, is frequently seen in patients with PCT.[1-4] PCT can also be associated with diseases (Table 2).

Table 1
What Can Exacerbate PCT?

Porphyria cutanea tarda (PCT) can be induced or exacerbated by multiple chemicals and drugs, including:

- Alcohol
- Estrogen
- Chlorinated phenols
- Chlorodibenzo-p-dioxin
- Hexachlorobenzene
- High doses of iron

Table 2
Diseases Associated with PCT

1. Diabetes mellitus can be seen in 15% to 20% of patients with porphyria cutanea tarda (PCT).
2. Patients with the following diseases have a higher incidence of PCT:
    - Lupus erythematous
    - Wilson's disease
    - Renal failure
    - Human immunodeficiency virus

What are the cutaneous symptoms?

The cutaneous symptoms of PCT are related to the accumulation of porphyrins within the skin. Porphyrins cause a phototoxic eruption by forming reactive oxygen species when exposed to light. The maximum absorption of light occurs at the Soret band, 400 nm to 410 nm, which lies within the wavelength of ultraviolet light, specifically ultraviolet A (Table 3).

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What Am I Looking For?

- Patients often complain of fragile skin and experience vesicles and bullae on the dorsal surfaces of their hands, feet, and occasionally the nose.
- When these bullae rupture, they result in scars and milia formation.
- Other manifestations include, hypertrichosis of the cheeks and temples and hyperpigmentation of sun-exposed areas.
- The central face often has a purple/red suffusion, and the accumulation of uroporphyrins can result in waxy yellow/white sclerodermoid papules and plaques on the face, chest, and back of the neck.
- The combination of the above lesions give the patient a premature aged appearance.

What tests can be done?

The diagnosis of PCT can be strongly suspected on clinical grounds, since other disorders typically lack the hirsutism, dyspigmentation and sclerodermoid changes. However, a useful confirmatory test that can be easily done in the office is a urine test. The urine is either left on a windowsill, exposed to a wood's light, or a small amount of acetic acid can be added, and a characteristic burgundy/red colour is seen. More extensive testing can be done by submitting 24-hour collections of urine and stool and quantifying the amount of porphyrins present. In PCT, there is a high con-

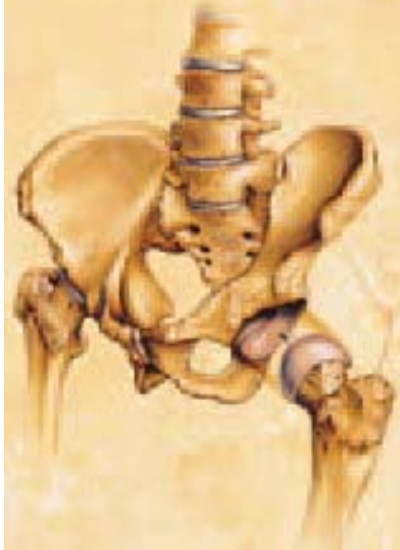

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Table 3
Differential Diagnosis

1. Epidermolysis bullosa acquisita
2. Photosensitising drugs including:
   - furosemide
   - nalidixic acid
   - naproxen
   - sulfonamides
   - tetracycline
3. Polymorphous light reaction
4. Connective tissue disease.

centration of uroporphyrin found in the urine and isocoproporphyrin in the stool. Since the prevalence of hepatitis C and hemochromatosis can vary from 17% to 94% in patients with PCT, dependent upon the population base, both diseases should be tested and if found to be positive, treated appropriately.

What are the treatment options?

1. The first treatment option consists of the **elimination of all precipitating factors.** Both alcohol and estrogen have been shown to induce the hepatic enzyme, aminole vulinic acid synthetase, the rate-limiting enzyme in porphyrin biosynthesis, and thereby, increase the amount of porphyrins produced.

Practice pointer

A useful confirmatory test that can be easily done in the office is a urine test. The urine is either left on a windowsill, exposed to a wood's light, or a small amount of acetic acid can be added and a characteristic burgundy/red colour is seen.

2. **Strict sun avoidance,** using clothing as a physical barrier as well as barrier sunscreens, such as titanium dioxide and zinc oxide, is invaluable in minimising the amount of photosensitivity.

3. **Phlebotomy** is also an effective form of treatment, since it reduces the iron load. Iron inhibits uroporphyrinogen decarboxylase and exacerbates the accumulation of porphyrin precursors. Typically 500 mL of blood every two weeks is removed until the hemoglobin reaches 10 g/dL or a serum iron of 50 g/dL to 60 g/dL.

4. **Antimalarials** are an alternate form of treatment in a patient who already has a low blood count, or in conjunction with phlebotomy in difficult cases. However, be aware that full doses of antimalarials can lead to a severe hepatoxic reaction so chloroquine should only be given in low doses usually 125 mg twice a week.

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Although PCT is relatively uncommon, with an estimated 1 in 25,000 cases reported in North America, it can be easily diagnosed by its characteristic clinical presentation and laboratory findings. Most patients with PCT can be successfully managed with elimination of eliciting factors, sun avoidance, phlebotomies and low-dose chloroquine with a mean remission time of 14.9 months. Dx

References
1. Habif, TP ed: *Dermatology Pearls and Pitfalls.* Clinical Communications, 1995, pp. 78-80.
2. Provost TT and Flynn: *Cutaneous Medicine. Cutaneous Manifestations of systemic diseases.* BC Decker Hamilton, London, 2001, pp. 487-500.
3. Odom RB, James WD, Berger TG: *Andrew's Diseases of the Skin: Clinical Dermatology.* Ninth Edition. WB Saunders Company, Philadelphia, 2000, pp. 652-56.
4. Scarlett, Y and Brenner DA: Porphyrias. J Clin Gastroenterol 1998; 27(3):192-98.

Suggested Reading
1. Roelandts, R: The Diagnosis of Photosensitivity. Arch Dermatol, 2000; 136(9):1152- 57.

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