

## Sclerodermatous Changes Revealing Porphyria Cutanea Tarda

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**We report a patient with sclerodermatous changes secondary to porphyria cutanea tarda, who responded to venesection treatment. The clinical characteristics of cutaneous sclerosis secondary to porphyria cutanea tarda and the degree to which it can mimic generalized morpheas are discussed. Key words: morpheas; scleroderma; venesection.**

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Cutaneous sclerosis of porphyria cutanea tarda (PCT) can mimic generalized morpheas, resulting in misdiagnosis and delay of effective treatment. We report such a case in a dark-skinned man who presented also with a rapidly progressive dermatoheliosis.

### CASE REPORT

A 74-year-old man presented with inflammatory sclerodermatous plaques and dermatoheliosis that had started 2 years earlier. He denied any systemic symptoms such as Raynaud's phenomenon, esophagitis or dyspnea. A 6-month course of systemic corticosteroids had failed in controlling the dermatosis. His past medical history consisted only in an excessive alcohol ingestion for many years. On examination, he had numerous erythematous, inflamed, indurated patches of the skin on sun-exposed areas and on the anterior chest and back, but not on the nipples (Fig. 1). They were associated with a scarring alopecia with extensive areas of permanent hair loss. In addition, he had severe signs of dermatoheliosis, with deep wrinkles and comedones on his face. They had occurred 2 years earlier, according to the patient, and were surprisingly severe in this dark-skinned mediterranean man (Fig. 2). Finally, he had pigmentary disorders that were difficult to evaluate, ranging from a brown mottled hyperpigmentation in sun-exposed skin to loss of pigment in scleroderma-like plaques. There was no atrophy of the nose or lips, no telangiectases and no radial furrowing of the mouth. The examination was otherwise normal. In particular, there were no bullae, milia, or skin fragility. Initial routine laboratory tests revealed a mild hepatic cytolysis (ALAT, 62 U/l; ASAT, 47 U/l), an iron overload, 33.5 µmol/l and an elevated ferritin at 510 µmol/l (39–340). Antinuclear antibodies, double-stranded DNA, ENA, ANCA and anti-Scl 70 were all negative, and complement levels were normal. Hepatitis B, hepatitis C and human immunodeficiency virus serology were negative. A chest X-ray and ultrasound of the liver were normal. Histopathologic examination of a biopsy specimen from his trunk confirmed the sclerodermatous changes, with thickening of the dermis by swollen collagen bundles and a mild perivascular mononuclear infiltrate. Direct immunofluorescence was negative. Porphyrin analysis in urine and faeces confirmed the diagnosis of PCT: total urinary uroporphyrin I+III: 725 nmol/l (normal <24), coproporphyrin I+III: 182 nmol/l (normal <115) and faecal coproporphyrin I+III: 75 nmol/g dry stool (normal <46). The patient was instructed to avoid alcohol and sunlight, and repeated venesections were performed. The reduction of urinary uroporphyrin and ferritin levels to within normal was obtained with 20 phlebotomies. The sclerodermatous skin changes simultaneously improved (Fig. 3).

### DISCUSSION

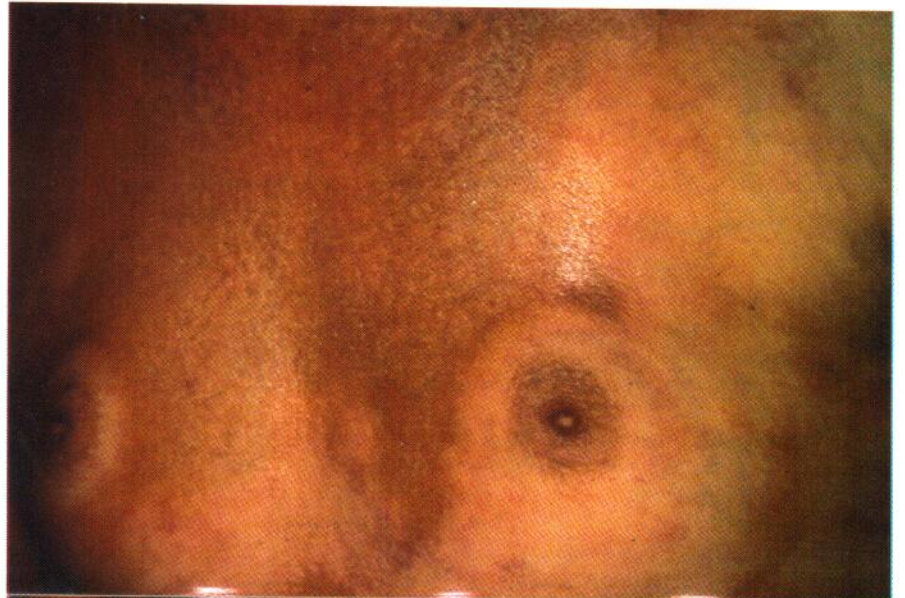
Cutaneous sclerosis is a well-known feature of PCT, occurring

in up to 18% of patients with PCT (1). In the majority of cases, other more characteristic cutaneous features of PCT are present, especially skin fragility of the dorsa of the hands with bullae, scars and crusts. In rare instances, like our case, cutaneous sclerosis and lack of skin fragility can be the major manifestations of PCT (2). The clinical and histological resemblance between sclerodermatous changes of PCT and morpheas can then be so close that it results in misdiagnosis and delay of effective treatment. However, we would like to draw attention to some particular features of sclerodermatous PCT which might allow clinicians to suspect the diagnosis and encourage them to perform porphyrin analysis. The presence of sclerodermatous changes may be an indication of PCT when plaques occur in certain zones, especially the V-shaped area of the neck and presternum. In the literature, the involved areas are described as hypopigmented, pale-yellow, indurated plaques, usually surrounded by atrophic hyperpigmented skin rather than by the typical lilac ring (3, 4). Yet, our patient developed inflamed, sclerotic plaques with erythematous borders. Surprisingly, there were no lesion on the nipples. Such a clinical picture has already been described (5). In view of our case, the diagnostic value of pigmentary changes and actinic damages must also be emphasized. Finally, despite negative immunologic laboratory tests, an association between morpheas and PCT may be difficult to rule out (6). It is only the effect of PCT therapy on the sclerodermatous changes which allows conclusion – as in our case, improvement of the morpheiform changes with venesection proved their link with PCT. Indeed, during PCT, the degree of improvement of the sclerodermatous changes is generally proportional to the reduction of the urinary uroporphyrin levels toward normal (7).

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*Fig. 1.* Erythematous, inflamed, indurated patches of skin except on the nipples.



*Fig. 2.* Severe dermatoheliosis.



*Fig. 3.* Improvement of the sclerodermatous changes.

