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Clinical spectrum of porphyria cutanea tarda

VALERIA BRAZZELLI, MARIA GRAZIA CHIESA, CAMILLA VASSALLO, MARCO ARDIGÒ, GIOVANNI BORRONI
Dept. of Human and Hereditary Pathology, Institute of Dermatology, University of Pavia, IRCCS Policlinico S. Matteo, Pavia, Italy

The term porphyria cutanea tarda (PCT) refers to a group of disorders biochemically charcaterized by reduced activity of uroporphyrinogen decarboxylase in liver and, in familial cases, in a number of tissues and cells including erythrocytes with subsequent accumulation of uroporphyrin and other porphyrins in organs and tissues.[1,2] The presence of oxidized porphyrins in the skin adsorbing long ultraviolet and visible light, is responsible for the main clinical features of PCT. Clinically, PCT is a photosensitivity disorder without the neurologic signs of precursor porphyrias diseases.[3]

The most common variety of PCT is the sporadic or acquired form in which there is a predisposition to acquire the liver enzymatic defect which develops in response to different triggering exogenous factors. The patients are usually adults and they have no family history of PCT.[4]

The most frequent factor implicated in precipitation and aggravation of PCT is ethanol, although only a few alcoholics with chronic liver damage develop PCT. Other common exacerbating factors are therapeutic estrogenic hormones, different drugs (griseofulvin, vitamin B12, sulphonamides, barbiturates, hydantoins), iron excess, viral infections (such as hepatitis B and C viruses, HIV infection), and all factors affecting liver function PCT has also been associated with chronic renal failure, long-term hemodialysis, diabetes mellitus, and, more rarely, with other systemic diseases.[4]

Acute attacks of PCT are not common, while a history of chronic photosensitivity is usual. Liver involvement is always present even if clinical manifestations are not evident. Urinary porphyrin assays are increased and it is possible to observe pink-red fluorescence with a Wood's lamp (Figure 1).

Clinical features

Cutaneous features of PCT[1-5] consist mainly of bullous lesions and increased skin fragility. Bullae are tense (Figure 2), clear-filled first, then cloudy or sero-hemorrhagic, and they usually arise on sun exposed skin and less frequently are surrounded by an erythematosus hue. Moreover, erosions, ulcers and scale-crusts are present on sun-exposed sites such as dorsa of hands (Figure 3), fingers, forearms, face and neck.[6] Generally, these lesions develop following minor trauma or spontaneously. In fact, the increase of porphyrin concentration in the skin is the cause of both cutaneous fragility and photosensitivity.[5] The lesions heal slowly with atrophic, sometimes depressed, scars (Figure 4). Within scars post-bullous milia may be observed.

Sclerodermatous features can occur and they are more common in females. Preauricular calcification is a common complication.[7]

Photosensitivity induces severe erythema and ede-

Figure 1. Urinary pink-red fluorescence induced by Wood's lamp.

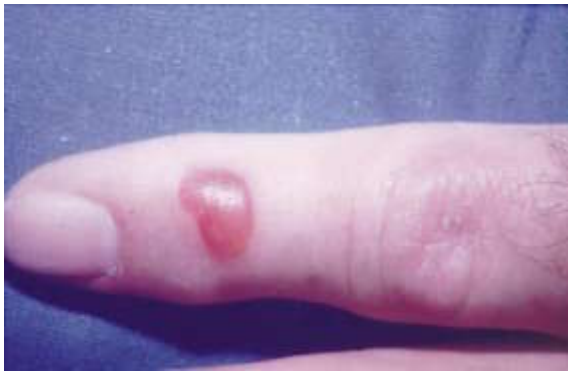


Figure 2. Tense, sero-hemorrhagic bulla on the dorsal surface of the second finger of the left hand.

Correspondence: Giovanni Borroni, Clinica Dermatologica, IRCCS Policlinico S. Matteo, 27100 Pavia, Italy.
Phone: international +39-0382-503494 – Fax. international +39-0382-526379.

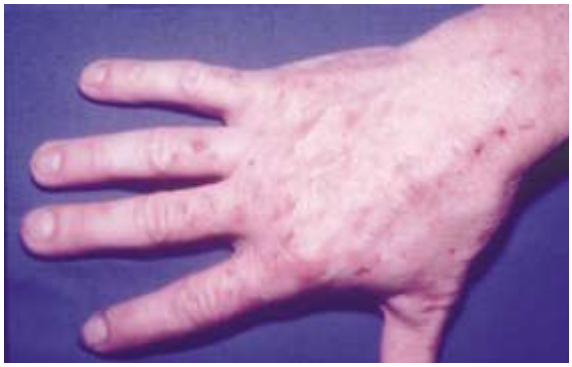


Figure 3. Erosions, ulcers and scale-crusts on the dorsum of hand.

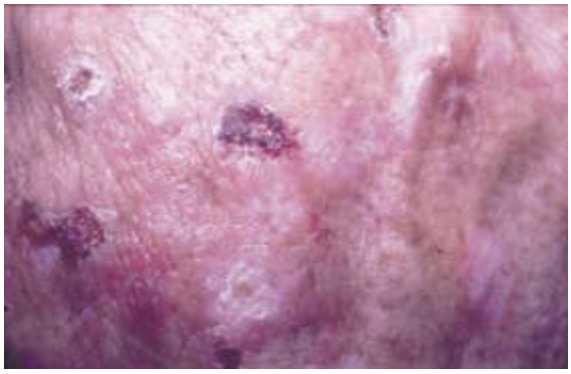
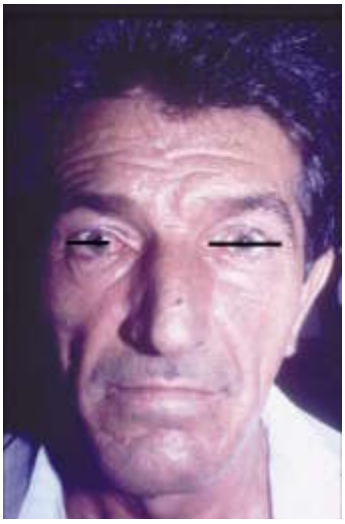
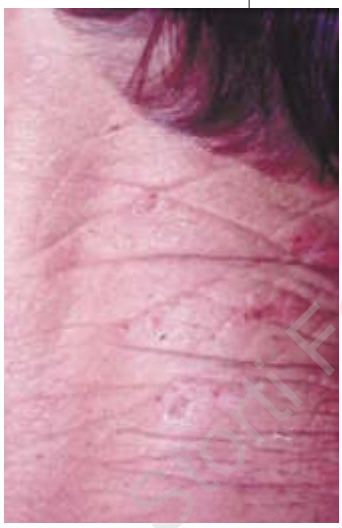


Figure 4. Atrophic and depressed scars.





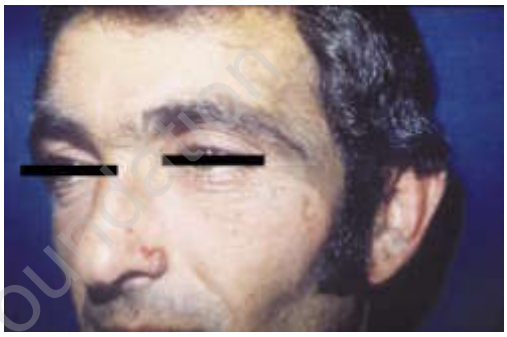


Figure 5 (left). Erythema and edema on the face accompained by hyperemic conjunctivitis.
Figure 6 (center). Cutis rhomboidalis nuchae.
Figure 7 (above). Hypertrichosis on periorbital, temporal and malar areas.

ma on sun-exposed sites accompanied by hyperemic conjunctivitis (Figure 5). No other mucosa other than conjunctiva is involved. Chronic actinic damage is responsible for nodular elastosis with cysts and comedos, thickening of the skin and cutis rhomboidalis nuchae, even in younger patients (Figure 6).

Hyper-hypopigmentations are also present with reticulate, spotty or diffuse patterns, especially localized on temporal regions, cheeks, V-shaped area and arms. Hypertrichosis is a frequent feature and consists of long, dark hair on periorbital, temporal, malar areas (Figure 7), eyebrows and, less frequently, arms and trunk.

Uncommon cutaneous manifestations of PCT include alopecia, affecting the fronto-parietal, temporal and occipital regions and centro-facial papular lymphangiectases. Darkening of hair color is not a common, albeit highly diagnostic, sign.[8]

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