Pseudo porphyria or porphyria cutanea tarda?

¿Pseudoporfiria o porfiria cutánea tarda?

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Dear Editor:

Manuscripts about pseudoporphyria (PP) and porphyria cutanea tarda (PCT) are commented here, emphasizing some gastroenterological relationships and possible diagnostic challenges or misdiagnosis, in special for daily practice of primary health care workers. Batrani *et al*. described PP associated with the use of imatinib and manifested by bullae, erosions, scarring and milia on the dorsum of hands and feet, in absence of photosensitivity [(1)]. Biopsy sample showed subepidermal blister, dermal festooning of papillae and thickened vessels; and the level of urinary porphyrin was within the normal range [(1)]. Imatinib has been also associated with PCT; however, in this patient the diagnosis was consistent with PP [(1)]. Peláez-Castro *et al*. reported the first Peruvian case of PP in a hemodialytic woman with facial hyperpigmentation, bilateral bullae on the fingers, and some crusts on the dorsum of hands [(2)]. Biopsy study revealed sub epidermal blisters, with dermal festooning and mild lymphocytic infiltrate; however, the normal profiles of porphyrins allowed confirm the diagnosis of PP [(2)]. This condition is related to renal failure and hemodialysis, non-steroidal anti-inflammatory drugs, furosemide, retinoids, sulphonamides, tetracyclines, dapsone, and nalidixic acid [(1,2)]. Hypertrichosis, hyperpigmentation, calcifications and sclerodermoid lesions are uncommon in PP; however, clinical, histopathological and immunofluorescence features can mimic PCT [(1)]. Therefore, normal urinary and plasmatic porphyrins are mandatory for diagnosis of PP [(1,2)].

PCT is the most frequent porphyria, due to deficiency of hepatic uroporphyrinogen decarboxylase (UROD), with high levels of uroporphyrinogen, uroporphyrin/ coproporphyrin higher than 3:1, elevation of heptacarboxilate, and photosensitivity are classical findings [(3-5)]. This condition is classified in two types: I or sporadic, and II or familial (20%), and clinical manifestations are recurrent vesicles and bullous lesions in skin exposed to sunlight [(3,5)]. The urine may be red-to-brown on natural light, changing to pink-to-orange under Wood's lamp [(5)]. Risk factors include alcoholism, smoking, hepatitis C (HCV), HIV, estrogen, and imatinib; liver cirrhosis due to HCV and hemochromatosis are described in association with PCT [(3-6)]. Although some authors reported significant improvement of PCT, controversial persists if the treatment of HCV with interferon and ribavirin might precipitate clinical relapses [(3,4,7)]. This may be relevant, if considering that PCT may be associated with HCV in up to 66% of cases [(3)]. Additionally, to HCV, hereditary hemochromatosis can play a role in PCT [(5,6)]. The causal relationship of high levels of serum iron and of ferritin with PCT is well understood [(3,5)]; moreover, both hemochromatosis and PCT are associated with mutations in the HFE gene [(5,6)]. Biopsy data may be indistinct from PP; even the sub epidermal blisters with lymphocytic infiltrates and dermal papillae extending to the cavity of the bulla do not confirm the diagnosis of PCT, which depends on the plasmatic and urinary typical porphyrin profiles [(1,2,5)]. Differential diagnoses include pseudoporphyria, bullous lupus erythematosus, epidermolysis bullosa, hydroa vacciniforme, polymorphic light eruption, scleroderma, and solar urticaria[(5)]. Control of PCT involves photo protection, chloroquine, phlebotomy, andiron chelation [(3,5,7)].

These comments were addressed aiming to enhance the suspicion index of primary health workers about these dermatological conditions involving possible diagnostic pitfalls. Major concerns should include expensive laboratory determinations in low-income countries.

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