

**Coexistence of a systemic lupus erythematosus and porphyria cutanea tarda: case successfully improved by avoidance of sun exposure**

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Keywords: systemic lupus erythematosus, porphyria cutanea tarda

## *CASE REPORT*

A 46-year-old Japanese woman presented with vesicles on her nose and the dorsal aspect of her hands since 3 months. Physical examination revealed tense blisters and atrophic erythematous plaques on her arms and the dorsal side of her hands (Fig. 1a). Erosions were also scattered on the face (Fig. 1b). She also had hypertrichosis on her face. The patient was first diagnosed as having systemic lupus erythematosus (SLE) in 1997 at the age of 41 years, when she presented with alopecia, fever, arthralgia, Raynaud's phenomenon, hemolytic anemia and lymphadenopathy. Liver transaminase levels were slightly elevated. She had no sign of hepatitis including autoimmune hepatitis, drug-related hepatotoxicity, alcoholic hepatitis, or viral infection. Systemic lupus erythematosus was well-controlled with a daily 5-mg dose of prednisolone (PSL).

At age of 43 years she developed erythematous plaques with scaling on the back of her hands. Clinically the skin eruption at that time appeared to be discoid lupus erythematoses. She was treated with a steroid ointment and her eruptions gradually improved, but new skin lesions repeatedly appeared on her hands.

Laboratory investigations at age 46 years while taking PSL 5mg disclosed the following results: white cell count  $3.9 \times 10^9/l$  with lymphopenia of  $0.78 \times 10^9/l$ , red cell count  $3.35 \times 10^{12}/l$ , hemoglobin of 11.4g/l, and a

platelet count of  $165 \times 10^9/l$ . Liver function profile included total bilirubin was 1.7 mg/dl, GOT 46 IU/l, GPT 82 IU/l,  $\gamma$ -GTP 142 IU/l and alkaline phosphatase 456 IU/l. Renal function and electrolytes were normal. Antinuclear antibody (ANA) titers were positive at a dilution of 1/80 (normal value, 1/40) with a homogeneous pattern. Histopathological examination of the back of the hands demonstrated a subepidermal blister with slight inflammatory infiltrate. No eosinophilic deposit around the vessels and little edematous change was present in the dermis. (Fig. 2). Direct immunofluorescence revealed a granular fashion at the dermoepidermal junction (DEJ) with IgG, IgM, and C3. Deposition of C3 around the blood vessels and several clusters of colloid bodies associated with IgM were also observed in the upper dermis. Laboratory value of fractionated porphyrins in her urine revealed the following: uroporphyrin 1080  $\mu\text{g/l}$  (normal: <20), coproporphrin 110  $\mu\text{g/l}$  (normal <100), porphobironogen 1.3 mg/l (normal <2), and  $\delta$ -aminolevulinic acid 3.5 mg/dl. The value of porphyrins in her serum was within the normal range, confirming the diagnosis of PCT. Subsequently, she was treated with topical steroid ointment, and avoidance of sun exposure was implemented. Three months later the value of uroporphyrin and coproporphrin in her urine was almost normal and no further blisters appeared.

## DISCUSSION

Since the association of SLE and PCT was first described by Linden in 1954 (1), approximately 50 cases of LE (all variants) have been described in association with PCT, including 15 cases reported by Gibson and McEvoy (2). PCT results from decreased activity of uroporphyrinogen decarboxylase (UROD). Alcohol, oral contraceptives, polychlorinated hydrocarbons, disturbances of iron metabolism, hepatitis C and infection with HIV are recognized as precipitant factors for PCT (3). Approximately 80 % of PCT patients have the sporadic form in which UROD deficiency is restricted to the liver and the remainder has the familial type in which mutations in the UROD gene are inherited in an autosomal dominant pattern (4).

It is not known whether the association between PCT and LE is coincidental or represents some common link. Harris *et al.* have suggested possible explanations for the coexistence of LE and porphyria: a common genetic abnormality, porphyria triggering an autoimmune response, preexisting LE resulting in an acquired metabolic fault leading to porphyria, and LE precipitating a genetically determined metabolic fault resulting in porphyria (5). It is interesting that the gene for UROD is located on chromosome 1 (6), and that the 1q41-q42 region of that chromosome is probably linked to SLE (7).

Our patient did not drink much alcohol, take oral contraceptives, nor had

any blood transfusions. She did not have HCV or HIV infections. Her liver function test was abnormal, and therefore dysfunction of her liver caused by SLE may have been a factor in precipitating the PCT. She has suffered from hemolytic anemia since 1997, so an overload of iron may have caused liver dysfunction and could also have precipitated the PCT. The histological findings of the subepidermal blistering are compatible with PCT, and positive immunofluorescence staining at the DEJ may have been caused by the SLE and sun-exposed skin.

Association of SLE and PCT is not common, and this combination usually poses practical problems and occasionally therapeutic dilemmas. Phlebotomy is one treatment for PCT, but it is not advisable in LE patients who have anemia. Antimalarials are used for the treatment of LE and PCT. Severe toxicity can result from using high dose of chloroquine(8), and hydroxichloroquine may also be associated with abdominal crises in some cases. (9). However, low-dose chloroquine therapy has been shown to be effective without causing serious hepatotoxicity(10). Therefore, it is important to avoid high-dose chloroquine therapy in cases of joint LE and PCT.

Our patient's skin lesion and the quantity of porphyrines in her urine have been improved since she started the avoidance of sun exposure together with the topical steroid ointment treatment. Strict total sun protection should

be recommended to patients with PCT and SLE because both conditions can be activated by long wavelength light bound in sunlight.

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## LEGENDS FOR FIGURES

**Fig 1.** (a) Right hand with a tense blister, erosions and several pigmented lesions. (b) Numerous erosions on the face.

**Fig 2.** Subepidermal blister with slight inflammatory infiltrate. No eosinophilic deposit are observed around the vessels and few edematous change were present in the dermis.(H&E x 40)





**Fig. 2**

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