Vesiculobullous skin lesions on the hands and face

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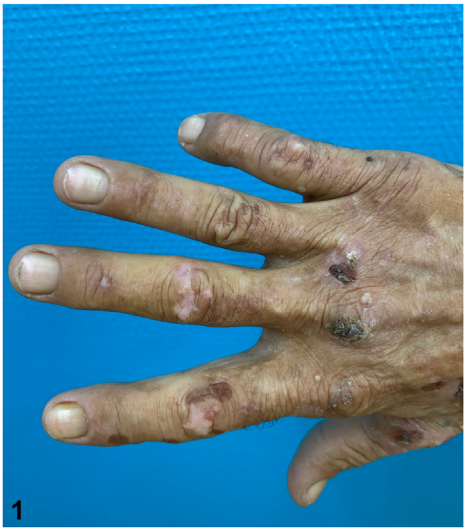

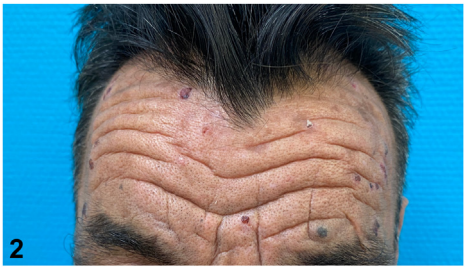

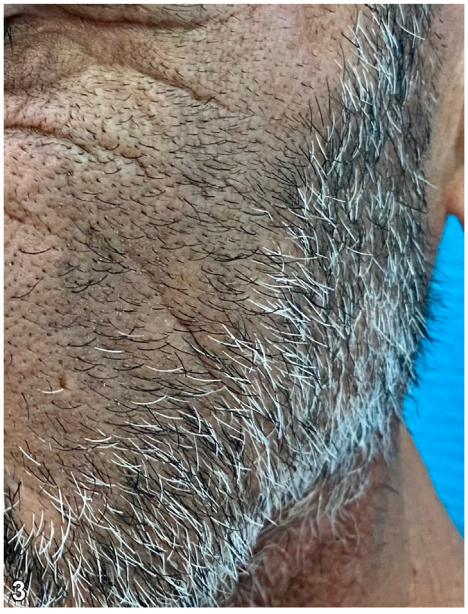

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CASE PRESENTATION

A 58-year-old man presented at the dermatology clinic with bullous lesions on his hands that had appeared 3 months earlier, in early summer. Upon examination, bilateral tightly roofed vesicles on his hands, along with milium-grain scars (Fig 1), crusted erosions on the face and hands (Figs 1 and 2), bilateral malar hypertrichosis (Fig 3), diffuse skin pigmentation, and port-red urine were observed. The skin biopsy showed dermal-epidermal junctional cleavage without alteration in the epidermal roof or dermal inflammatory infiltrate.

Question 1: What is the likely diagnosis?

A. Bullous pemphigoid

B. Porphyria cutanea tarda (PCT)

C. Scabies

D. Systemic lupus erythematosus

E. Contact dermatitis

Answers:

A. Bullous pemphigoid — Incorrect. Bullous pemphigoid typically presents with tense bullae on erythematous skin, without the characteristic features of PCT such as malar hypertrichosis or port-red urine.

B. PCT — Correct. The presented clinical features, including vesiculobullous lesions in photo-exposed areas, milium-grain scars, malar hypertrichosis, skin pigmentation, and port-red urine, are typical of PCT. It is a rare skin disorder resulting from acquired or inherited hepatic deficiency of uroporphyrinogen decarboxylase.[1,2]

C. Scabies — Incorrect. Scabies usually causes intense itching and burrows in the skin, including in interdigital spaces.

D. Systemic lupus erythematosus — Incorrect. Systemic lupus erythematosus may present with cutaneous manifestations on sun-exposed areas, but it is characterized by a prominent rash on the cheeks and nose, commonly known as a "butterfly rash".

E. Contact dermatitis — Incorrect. Contact dermatitis typically presents with erythema, edema, and vesicles in areas of contact with an offending agent, which does not match the presentation described in the case.

Question 2: Which of the following laboratory findings is most commonly associated with this condition?

A. Elevated serum IgE levels

B. Decreased serum ferritin levels

C. Increased plasma uroporphyrin levels

D. Antinuclear antibodies

E. Elevated serum creatinine levels

Answers:

A. Elevated serum IgE levels — Incorrect. PCT is not typically associated with elevated serum IgE levels.

B. Decreased serum ferritin levels — Incorrect. PCT is commonly associated with hyperferritinemia due to iron overload, not decreased levels. In the case presented herein, laboratory tests revealed hyperferritinemia (1035 ng/mL).

C. Increased plasma uroporphyrin levels — Correct. Increased serum uroporphyrin levels are a characteristic laboratory finding in patients with PCT. In the case presented herein, consistent biological findings with increased uroporphyrin in blood, urine, and stool confirmed the diagnosis. Twenty percent of PCT cases are of genetic origin, which justifies a systematic determination of uroporphyrinogen decarboxylase activity (negative for the patient presented herein).[3]

D. Antinuclear antibodies — Incorrect. PCT is not considered a primary autoimmune disease. Therefore, the presence of antinuclear antibodies is not a characteristic feature of PCT. If present, it may suggest the presence of another autoimmune condition rather than PCT itself.

E. Elevated serum creatinine levels — Incorrect. Elevated serum creatinine levels are not typically associated with PCT.

Question 3: Which of the following triggers is NOT commonly associated with exacerbation of this condition?

A. Alcohol consumption

B. Hepatitis C infection

C. Exposure to sunlight

D. Hydroxychloroquine intake

E. Estrogen-based contraception

Answers:

A. Alcohol consumption — Incorrect. Alcohol consumption is a well-known trigger for exacerbation of PCT due to its effect on hepatic metabolism.

B. Hepatitis C infection — Incorrect. Hepatitis C infection is commonly associated with PCT and can exacerbate the condition. In the case presented herein, laboratory tests revealed elevated liver function tests (aspartate transaminase 228 IU/L and alanine transaminase 198 IU/L) with positive serology for hepatitis C. This patient had multiple potential triggers: underlying, and previously unknown, liver disease with hepatitis C infection, hyperferritinemia, and alcohol consumption.

C. Exposure to sunlight — Incorrect. Exposure to sunlight is a known trigger for exacerbation of PCT, as it can induce porphyrin production in the skin.

D. Hydroxychloroquine intake — Correct. Conversely, the treatment of PCT can be achieved using very low doses of synthetic antimalarial drugs, which remove excess porphyrins from the liver and increase their excretion in the urine.[4] For this patient, hydroxychloroquine was deemed unnecessary and treatment for hepatitis C was initiated alongside guidance for photoprotection to manage PCT cutaneous manifestations.

E. Estrogen-based contraception — Incorrect. Estrogens, such as contraceptives and hormone replacement therapies, are a known trigger for PCT and may cause cutaneous signs that may warrant discontinuation during a vesiculobullous episode.[5]

Key words

clinical; coproporphyrins; dermatology; face; hands; hepatitis C; liver; porphyria; porphyria cutanea tarda; risk factors; skin diseases; vesiculobullous

Conflicts of interest

None disclosed.

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