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INTERESTING MEDICAL IMAGE

Porphyria *Cutanea Tarda* in a Patient with Myelofibrosis

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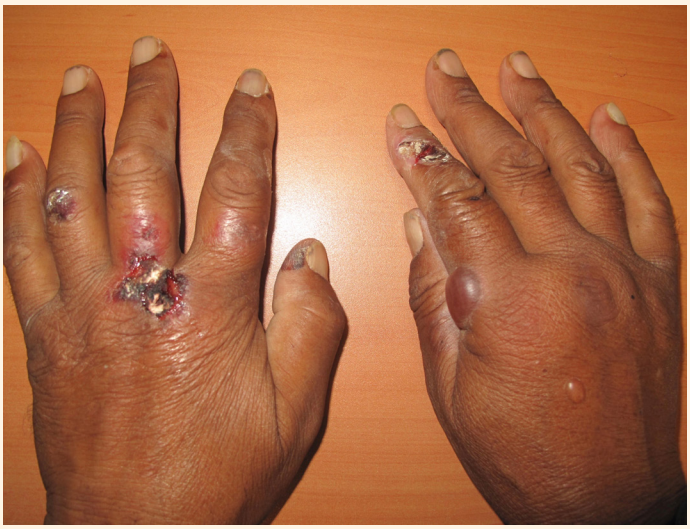

Figure 1: Tense bullae on the dorsal right hand and erosive lesions with *milia* and scarring on the dorsal left and right hand. Marked skin hyperpigmentation.

A 63-year-old male patient presented to a dermatology department with a four-month history of erosions affecting the *dorsum* of the hands. His medical history was remarkable for primary myelofibrosis on treatment with hydroxyurea and supportive transfusions for anaemia and thrombocytopenia. He also reported high alcohol consumption.

Physical examination showed ruptured blisters with signs of superinfection. Bacterial culture was positive for *Staphylococcus aureus* and the patient received treatment with oral antibiotics. One week later, he reported improvement of the erosive lesions but also new blistering lesions on the *dorsum* of the hands [Figure 1]. In addition, the patient also reported generalised skin hyperpigmentation in the last months.

Based on the suspicion of porphyria *cutanea tarda* (PCT), we observed urine through Wood's light, which revealed bright red-pink fluorescence [Figure 2].

Laboratory tests revealed elevated serum ferritin and decreased haemoglobin and platelets. Blood and urine porphyrines determination were compatible with PCT. Skin histology showed subepidermal blister formation and deposits of IgG in the dermal-epidermal junction.

The patient was treated with periodic phlebotomies, with a good clinical response. Patient consent was obtained for the publication of these images.

Comment

PCT is the most common human porphyria, due to a deficit in or inactivation of the uroporphyrine decarboxylase (UROD) enzyme, which leads to accumulation of photosensitive metabolites. The accumulated uroporphyrines and intermediates spill over into the blood circulation and are excreted in the urine.[1,2]

It usually happens in adulthood and there are several known triggers, such as viral hepatitis, alcohol, hormone therapies, haemodialysis and condition leading to iron overload. It has also been reported to be associated with haematological disorders, mainly with thalassaemia, multiple myeloma, hairy cell leukaemia, acute myeloblastic leukaemia, chronic lymphocytic leukaemia and chronic myeloid leukaemia.[3] There are a few reported cases of PCT in association with myelofibrosis, yet the mechanism by which this occurs

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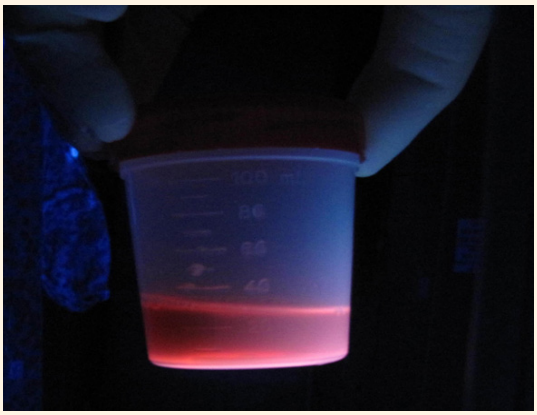

Figure 2: Urine sample fluorescing bright red-pink under Wood's light examination.

is unknown. One hypothesis suggests that myelofibrosis could induce hepatotoxicity leading to liver disfunction and PCT.[3] In the current case, the patient required multiple blood transfusions for treating anaemia caused by myelofibrosis. This situation provoked an iron overload and he finally developed PCT. Urine examination through Wood's lamp is a simple, fast and economic method that supports the diagnosis and can be performed immediately in the consultation room.

Treatment consists of removing probable secondary disease triggers, photoprotection, phlebotomy as an effective treatment of iron removal, resulting in improvement of hepatic UROD activities and the use of hydroxychloroquine.[1–3]

Here we report a case of PCT in a patient with idiopathic myelofibrosis and we suggest the use of Wood's light as a tool to aid diagnosis of such cases.

AUTHORS' CONTRIBUTION

All authors have contributed equally to the conception and design of the work, in drafting the manuscript and revising it critically. All authors approved the final version of the manuscript.

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