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Porphyria or not porphyria – that is the question...

A Smyth, L Redahan, C Wall, G J Mellotte

Department of Nephrology, Adelaide and Meath Hospital, Dublin, Ireland

Correspondence to A Smyth, andrewsmyth@physicians.ie

DESCRIPTION

A 55-year-old man presented with a 2-month history of blisters affecting his hands. His history was significant for treated multiple myeloma with residual monoclonal gammopathy of undetermined significance, and end stage renal disease on automated peritoneal dialysis. Medications included aspirin, clopidogrel, warfarin, pregabalin, bronchodilators and hydrocortisone. Examination showed widespread scarring on the dorsum of the hands with multiple intact fluid filled blisters (figure 1) in a classic distribution for porphyria cutanea tarda (PCT) (figure 2). He was transitioned to intermittent high-flux haemodialysis for other reasons, but coinciding with this change, his blistering rash became significantly worse. Skin biopsy showed mild hyperkeratosis, minimal dermal inflammation and negative direct immunofluorescence. Urine and faecal porphyria screens were negative, thereby excluding PCT and diagnostic of pseudoporphyria. Although aspirin was identified as

a possible causative agent, this could not be discontinued due to recent coronary metallic stenting. He was treated with n-acetylcysteine (NAC) 600 mg twice daily for 2 months with advice on minimising sun exposure. His rash improved significantly with healing of lesions and few fresh lesions. Pseudoporphyria has been associated with many medications including anti-inflammatory agents, antimicrobials and diuretics. Many cases of pseudoporphyria have been treated with NAC¹ in conjunction with high-flux haemodialysis.^{2 3} The mechanism of action of NAC is poorly understood but is felt to be related to an effect on 8-hydroxy-2'-deoxyguanosine (oh⁸dG) formation which has a role in free radical generation and subsequent tissue damage in association with 5-aminolevulinic acid which accumulates in porphyria. This diagnosis may be under-recognised in the renal population, and we suggest heightened awareness.



Figure 1 Multiple blistering lesions on dorsum of left hand at various stages of formation and rupture.



Figure 2 Large ruptured infection blister on dorsum of right hand.

Competing interests None.

Patient consent Obtained.

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