

Images in Nephrology
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Pseudoporphyria in a haemodialysis patient

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A 72-year-old female with end-stage renal disease due to chronic glomerulonephritis had been treated with conventional haemodialysis for 6 years. She was referred three months ago for management of blisters and cutaneous fragility on the dorsa of both hands (Figure 1a and b). The patient denied extensive sun exposure, sun beds, or use of medications other than calcium carbonate, vitamin B complex, folic acid or erythropoietin. Haemodialysis was

performed tri-weekly, 4 h per session, via an arteriovenous fistula over the left forearm, using a cellulose acetate dialyser (FB170, Nipro, Japan). The blood and dialysate flow rates were 200 and 500 cm³/min, and delivered Kt/V (Daugirdas) and urea reduction rates were 1.89 and 0.79, respectively. Blood tests were negative for anti-nuclear, anti-intercellular substance and anti-basement membrane zone antibodies. Urine was also negative for porphyrin and coproporphyrin (spectrophotometry). Skin biopsy (Figure 1c) found a subepidermal vesicle with tombstone-like protruding papillary dermis, whereas immunofluorescence analysis revealed smooth linear vascular staining

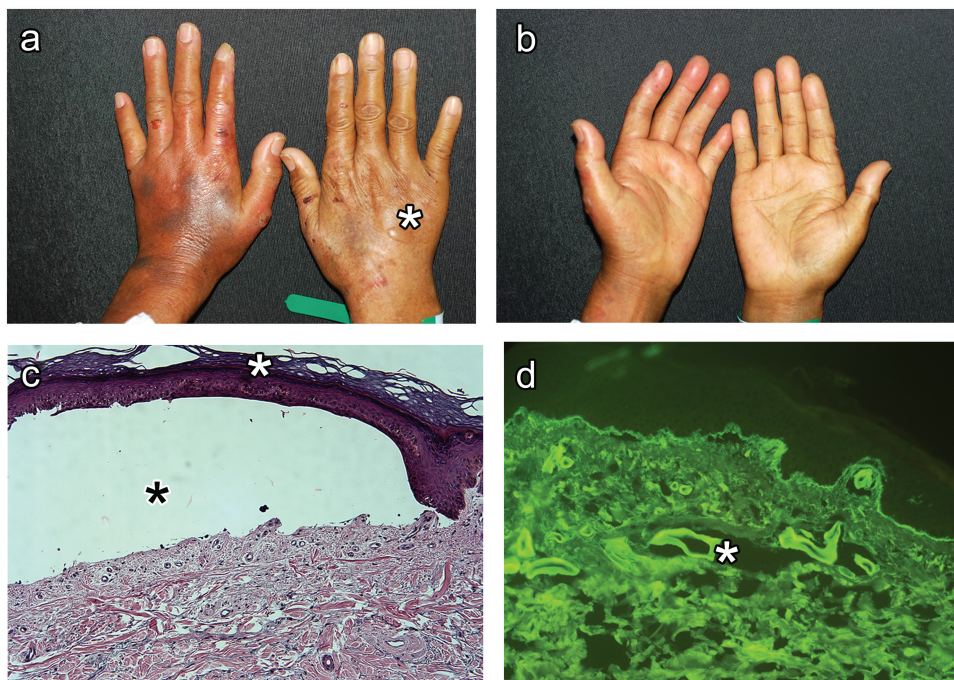


Fig. 1. Pseudoporphyria in a haemodialysis patient. (a, b) Blisters (white asterisk) and cutaneous fragility found on the dorsa of both hands. (c) Haematoxylin and eosin staining found a subepidermal vesicle with tombstone-like protruding papillary dermis and focal extravasation of red blood cells in papillary dermis. No significant inflammatory cells were seen (white asterisk: epidermis, black asterisk: vesicle, magnification $\times 400$). (d) Direct immunofluorescence analysis revealed smooth linear vascular staining by IgG (white asterisk, magnification $\times 400$).

staining by IgG (Figure 1d) and IgA. Pseudoporphyria was diagnosed. The blistering lesions gradually improved after initiation of high-flux haemodialysis with a polysulphone dialyser (F80, Fresenius, Germany). At the time of follow-up, the blood and dialysate flow rates were 280 and 500 cm³/min, and delivered Kt/V (Daugirdas) and urea reduction rates were 2.40 and 0.83, respectively.

Pseudoporphyria is an uncommon blistering disorder with clinical and histological similarities to porphyria cutanea tarda, but lacks urine and serum porphyrin elevations [1]. Pseudoporphyria has been linked with numerous aetiologies including renal failure, ultraviolet A radiation and photosensitizing medications (naproxen, furosemide, nalidixic acid, bumetanide, tetracyclines, amiodarone, etc.) [1]. It is possible that when multiple contributing factors are present in haemodialysis patients, including commonly used photosensitizing medications, the risk of pseudoporphyria may be further enhanced [2]. The mechanism for induction of disease is not understood. Discontinuation of the suspected photosensitizing medication and strict ultraviolet A

protection are the mainstay of treatments. Successful treatment of pseudoporphyria following *N*-acetylcysteine and high-flux haemodialysis has been reported previously [3].

Conflict of interest statement. None declared.

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