Bullous lesions in a haemodialysis patient

Bullous Lesions in a Haemodialysis Patient

Justin Chan Hsian Loon, Thin Han, and Zaw

A 41-year-old lady on haemodialysis secondary to diabetic nephropathy with a history of untreated hepatitis C pre-sents with a 4-month history of painful bullous lesions on both her forearms, shins, feet, face, neck and left pinna. Figure 1 She had been previously independently mobilizing but now severely debilitated by pain. There were no new medications or evidence of systemic infections. Her workup revealed a ferritin of 4790 ug/L, haemoglobin of 110 g/L and elevated urinary porphyrins, and histology of the lesion was in keeping with porphyria cutanea tarda (PCT). The patient was initially treated with a 4-month course of hydroxychloroquine with no effect. A trial of weekly phle-botomy of 180 ml for 2 months during haemodialysis in combination with an increased dose of erythropoietin resulted in rapid resolution of her lesions and pain, ferritin of 450 ug/L and haemoglobin ranging between 90 and 100 g/L without symptoms of anaemia. PCT is normally provoked when the haem biosynthetic enzyme uropor-phyrinogen decarboxylase activity in the liver is decreased to below 20%, often from multifactorial causes including familial mutations in the uroporphyrinogen decarboxylase gene, iron overload, hepatitis C infection, alcohol, smoking, oestrogen containing oral contraception and failure to excrete porphyrins, especially in anuric haemodialysis patients, where porphyrins are not fully cleared by haemo-dialysis.1,2 Treatment options in haemodialysis patients include small volume phlebotomy with concurrent erythro-poietin, desferoxamine, an iron chelator as well as treat-ment of underlying Hepatitis C. Hydroxychloroquine is usually ineffective in haemodialysis patients and may result in worsening of PCT.3,4

References

1. MarkovaA,LesterJ,WangJ et al. Diagnosis of common dermopathies in dialysis patients: a review and update. Semin. Dial. 2012 Jul; 25
2. (4): 408–418. DOI: 10.1111/j.1525-139X.2012.01109.x. Review.
3. BulajZJ,PhillipsJD,AjiokaRS et al. Hemochromatosis genes and other factors contributing to the pathogenesis of porphyria cutanea tarda. Blood 2000 Mar 1; 95 (5): 1565–1571.
4. RyaliME,WhittierWL. Bullous skin lesions in a patient undergoing chronic hemodialysis. Semin. Dial. 2010; 23 (1): 83–87. DOI: 10.1111/j.1525-139X.2009.00672.x.
5. GisbertJP,García-BueyL,PajaresJM et al. Prevalence of hepatitis C virus infection in porphyria cutanea tarda: systematic review and meta-analysis. J. Hepatol. 2003 Oct; 39 (4): 620–627. Review.