Hong Kong J. Dermatol. Venereol. (2014) **22**, 153-154

Dermato-venereological Quiz

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An 80-year-old Chinese man was admitted for lower limb purpura. He had underlying diabetes mellitus, hypertension, chronic renal failure, hyperlipidaemia and ischaemic heart disease. The eruption was progressive for two weeks, extending from the legs to the buttocks. It was slightly itchy and painful, with some lesions developing into blisters. He was well systemically and remained afebrile. There was no recent change of medication or invasive procedures being performed. Examination revealed multiple purpuric macules, papules and blisters (Figure 1). Urine Multistix® was positive for red blood cells and protein. He had acute-on-chronic renal failure with an increase in creatinine from 215 mmol/dL to 384 mmol/dL, and proteinuria of 4 g daily. Otherwise, his complete blood picture and film, liver function and cryoglobulin levels were normal. His autoimmune markers, hepatitis serology, urine and blood culture were negative. Serum IgA was raised at 7 g/L (two-fold). Herpes viral culture of blister fluid was negative.

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An excisional skin biopsy was obtained on the leg for histological evaluation (Figures 2 & 3). Renal biopsy was obtained subsequently (Figure 4).

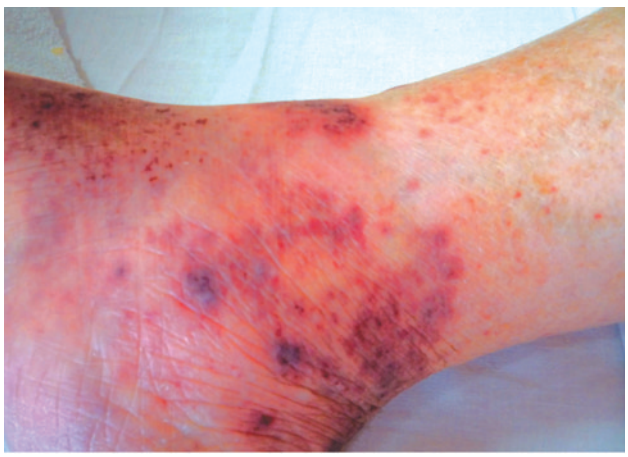

Figure 1.

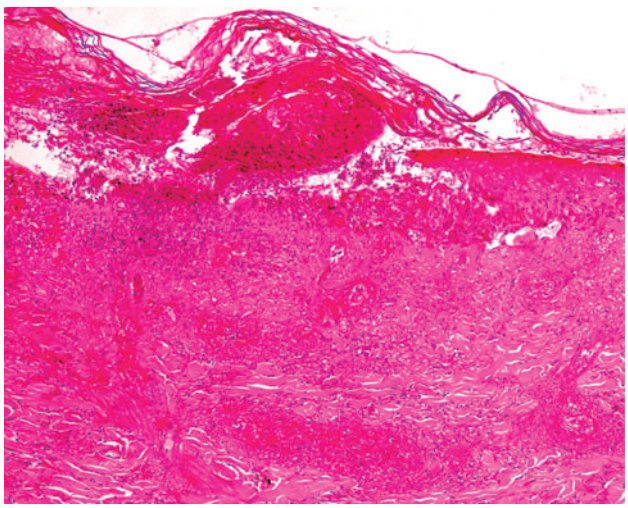

Figure 2. Skin biopsy. Haematoxylin & eosin stain. Magnification x 100.

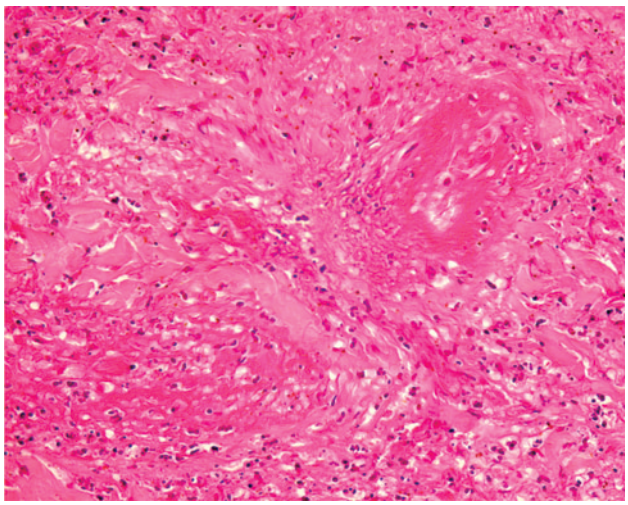

Figure 3. Skin biopsy. Haematoxylin & eosin stain. Magnification x 400.

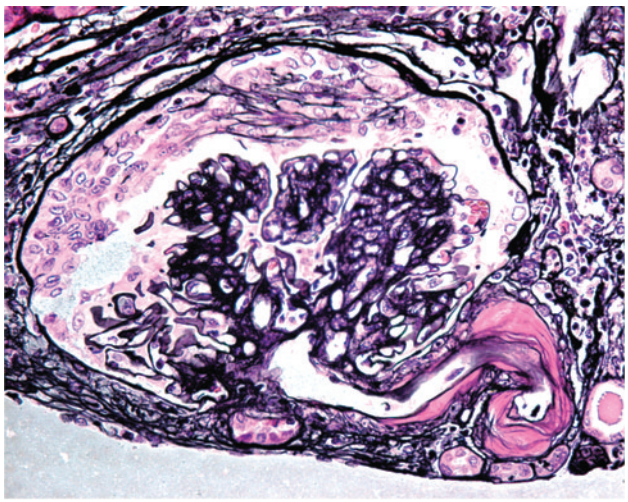

Figure 4. Renal biopsy. Periodic acid silver methenamine stain. Magnification x 400

Questions

1) What are the differential diagnoses?
2) What do the biopsies show?
3) What is the diagnosis?
4) What is the treatment and prognosis?

(Answers on page 163)

Answers to Dermato-venereological Quiz on pages 153-154

1. The clinical differential diagnoses include bullous leucocytoclastic vasculitis due to Henoch Schönlein purpura (HSP) or autoimmune causes, bullous insect bite reaction and bullous pemphigoid.

2. Histopathological examination of the skin biopsy showed a detaching necrotic epidermis and suggestion of subepidermal bulla with marked fibrinoid necrosis of small and medium vessel walls and surrounding nuclear dusts in the dermis. Immunofluorescence study showed vascular deposition of C3.

   Histopathological examination of the renal biopsy showed cresenteric glomerulonephritis with cellular cresents, mesangial expansion and hyaline afferent arteriole of glomerulus. Immunofluorescence study showed mesangial staining of IgA, C3 and lamda.

3. The diagnosis is HSP manifested as bullous leucocytoclastic vasculitis and crescentic glomerulonephritis. More commonly known as IgA vasculitis amongst nephrologists, HSP is a subgroup of idiopathic cutaneous small vessel vasculitis which predominantly occurs in children following recent streptococcal upper respiratory tract infection. It can involve the skin, synovia, gastrointestinal tract and kidneys and usually the recovery is spontaneous. If occurring in adults, it confers a worse prognosis with an increased risk of renal involvement and evolution to chronic IgA nephropathy. In elderly, other causes of small vessel vasculitis, such as gammaglobulinaemia, occult infection, malignancy and drug use have to be excluded.

4. In view of the renal involvement with nephrotic syndrome, the patient was treated with a tapering course of prednisolone (0.5 mg/kg) over six weeks, with close monitoring of subsequent hyperglycaemia. The cutaneous lesions resolved, followed by gradual recovery of the renal function. Dapsone may be used for bullous leucocytoclastic vasculitis without renal involvement. Antihistamines, colchicine and NSAIDs are usually used for mild, non-bullous lesions.