Remission of nephrotic syndrome in a patient with renal amyloidosis associated with Takayasu’s arteritis after treatment with dimethylsulphoxide

Amyloidosis is a chronic inflammatory disorder characterised by the presence of extracellular deposits of insoluble protein of unknown nature. Renal involvement generally carries a poor prognosis in patients with secondary amyloidosis, who usually display severe proteinuria, nephrotic syndrome and progressive renal failure; uraemia is the major cause of death in this group.1 There is as yet no accepted treatment for amyloidosis and the patient’s life expectancy is short.2 We present a patient with renal amyloidosis associated with Takayasu’s arteritis (TA) (aortitis syndrome) who demonstrated a favourable clinical course.

This 29 year old woman was admitted to our hospital in April 1981 with nephrotic syndrome. Her history revealed that, seven years earlier, she had presented with fever, vertigo, and systolic hypertension. Stenosis of the abdominal aorta was noted. TA was diagnosed; the patient had received prednisolone 5 mg daily since that time. There was no family hereditary disease including amyloidosis.

At admission in 1981, physical examination revealed a systolic heart murmur and murmurs in the neck, subclavicular area, and abdomen. Blood pressures taken in the right arm, left arm, right leg, and left leg were 158/40, 164/40, 110/40 and 104/40 mm Hg, respectively. Laboratory data revealed massive proteinuria of 8-0 g/day. There was no haematuria and renal function was normal, with creatinine 0.56 mg/dl, blood urea nitrogen 6-0 mg/dl, and glomerular filtration rate 123-4 ml/min. Serum albumin concentration was 2.3 g/dl and normal cholesterol concentration 289 mg/dl. C reactive protein concentration was >1 mg/dl and serum gamma globulin concentration 1-69 g/dl. The erythrocyte sedimentation rate (ESR) was 16 mm/h.

Examination of tissue obtained at renal biopsy revealed varying degrees of amyloid deposition in the mesangial areas of all glomeruli. A few arterioles showed mild focal deposits of amyloid. The glomeruli and the interlobular artery were positively stained with Congo red and antibody to amyloid-A protein. Electron microscopy showed amyloid fibrils in the glomerular mesangium and subepithelial region. Deposition of amyloid was
Electron micrograph of the second renal biopsy showing deposition of amyloid in the mesangial (*) and subepithelial (large arrows) regions. Note amyloid fibrils on the glomerular basement membrane (GBM) (arrow heads) and mesangial region (small arrows). Ep = epithelial cell; CL = capillary lumen. Horizontal bar represents 1 μm.

5 Reinmann H A. Recovery from amyloidosis. JAMA 1935; 104:1070–1.

Superficial migratory thrombophlebitis in a patient with reversible protein C deficiency and anticardiolipin antibodies

Superficial migratory thrombophlebitis is a rare manifestation of secondary hypercoagulable states. It has been described in association with infectious diseases, in particular secondary syphilis, in addition to malignancies and vasculitic disorders such as Behcet’s disease or thromboangiitis obliterans.1 We describe here the case of a patient with migratory superficial thrombophlebitis in which increased anti phospholipid antibody concentrations in plasma and a protein C deficiency were found. The clinical symptoms and the pathological laboratory results were reversible by antibiotic treatment.

A 26 year old male medical student was referred to the rheumatology outpatient unit because of migratory arthritis and multiple superficial thrombophlebitis. He had been in good health until one year earlier, when he had worked for two months in Namibia (Africa). Two months after his return to Germany he complained of red and painful nodules at the dorsum of both feet, which persisted for several months. After a further two months he developed migratory arthritis of the joints of the fingers and toes. A biopsy of a cutaneous nodule was performed which showed inflammatory infiltration of the wall of a thrombotic vein. The histology confirmed the clinical diagnosis of superficial thrombophlebitis. The patient received topical therapy (venous compression, local heparin) but was referred to our clinic because of persistence of symptoms.

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