A painful lower limb in a patient with a nephrotic syndrome

Case

The patient initially presented with a classic nephrotic syndrome (lower limb oedema, proteinuria and hypo-albuminaemia). Renal biopsy confirmed the diagnosis of primary amyloidosis. There were no other potential aetiologies for the amyloidosis. He was normotensive, non-diabetic and an ex-smoker of 3 years. His management consisted of diuretics to control the oedema symptomatically.

He presented acutely with a 3-day history of pain and unilateral swelling of the left leg. This was accompanied by the progressive development of pallor, coldness and loss of power in the limb. The limb had developed a mottled cyanotic appearance over the lower leg and foot 6 h prior to admission. Clinically all limb pulses were absent.

Question

What is your diagnosis?
Answer to quiz on preceding page

Urgent arteriography confirmed occlusion of the left common external iliac and femoral arteries with thrombosis. The patient was anticoagulated and taken to theatre for urgent thrombectomy. Arterial flow was re-established with satisfactory recovery of limb perfusion and function. Subsequent investigations demonstrated no underlying coagulopathy. Echocardiography and ultrasound of the pelvic vessels demonstrated no source of emboli. There had been no history of any trauma to the femoral region. His renal function was normal at that stage. He remained anticoagulated on warfarin. He subsequently progressed to end-stage renal failure and was successfully treated with CAPD. He died 4 years later of congestive heart failure secondary to amyloidosis. There were no further thrombotic events.

The nephrotic syndrome results in an hypercoagulable state with an increased risk of thromboembolic events. A number of mechanisms have been postulated, including increased levels of plasma procoagulant factors, reduced levels of anti-coagulant proteins, abnormal platelet function, altered endothelial function and decreased fibrinolytic activity [1]. The majority of thromboses are venous in origin with deep vein thrombosis of the lower limbs and renal vein thromboses well documented. There are only a limited number of case reports of arterial thromboses associated with a nephrotic syndrome in the literature, with tendency to occur more frequently in children [2]. However, the associated morbidity can be much more severe, resulting in limb amputation and a higher likelihood of death [3].

Arterial thromboses are rare. In the small number of cases reported there is an increased incidence in males (M:F 10:1), with the renal femoral and aorta the most common sites [3]. The underlying aetiology of the nephrotic syndrome does not appear to influence the rate of occurrence [4]. Arterial thrombi require urgent diagnosis and intervention. Analysis of five previous cases of lower limb arterial thrombosis in nephrotic patients showed an amputation rate of 60% [3]. Immediate thrombectomy and anticoagulation is required. Fortunately, in this case, early intervention resulted in limb salvage. The potential role for thrombolysis is unknown. The role of prophylactic anticoagulation is still debated, but should be considered in high risk patients who present with a nephrotic syndrome [5].

References


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