A 'catastrophic' fall!

Authors: Farishta Khan,[A] Rona Applewaite[A] and Anthony Abdullah[A]

Catastrophic antiphospholipid syndrome (CAPS) is a fatal autoimmune disease characterised by accelerated and widespread thrombosis resulting in multiorgan failure. Diagnostic criteria for CAPS include involvement of three or more organs or tissues, rapid development of manifestations, histological evidence of intravascular thrombosis and presence of antiphospholipid antibodies.[1] We present a rare case of a young patient who developed CAPS following a fall and the pivotal role of a cogwheel multidisciplinary team in aiding diagnosis and management.

A 35-year-old man with primary antiphospholipid syndrome, on warfarin, presented to the emergency department with a headache, fever and left foot pain. He reported falling from a height several weeks prior sustaining an injury to the left foot but was 'limping along'. Initial clinical examination revealed photophobia, diplopia, neck stiffness and an asymptomatic blotchy erythematous rash over the buttocks. Promptly, meningitis and subarachnoid haemorrhage were excluded. Blood tests revealed persistent deranged liver function tests, raised inflammatory markers and worsening thrombocytopenia. Imaging during admission unmasked a left calcaneal fracture. He continued to deteriorate with fluctuations in the Glasgow coma scale scores and repeat imaging revealed a parafalcine subdural haematoma in addition to new focal infarcts in the brain. While warfarin was paused and cautiously maintained on thromboprophylaxis dose of enoxaparin and broad-spectrum antibiotics for infection of unclear origin, the skin manifestations vented. Tender plaques and papules developed over the flanks (Fig 1) which progressed to palpable purpura with subcutaneous component and overlying early blisters (Fig 2). Skin histology confirmed thrombotic damage. Intravenous immunoglobulins, glucocorticoid therapy and low molecular weight heparin demonstrated a reassuring response and rituximab was commenced to enhance those effects. Patient stabilised from an impending death.

Although rare, CAPS can be missed and misdiagnosed as sepsis or heart failure due to the multiorgan involvement. Pathophysiology is not fully understood but dysregulation of the complement system from triggers including infection, trauma, surgery, pregnancy and malignancy causes C5a activation of leukocytes and endothelium and C5b-9 causes endothelium to secrete tissue factor causing microvascular thrombosis. The necrotic tissue activates the complement system in an alternative way and a vicious spiral ensues, leading to a thrombotic storm.[2]

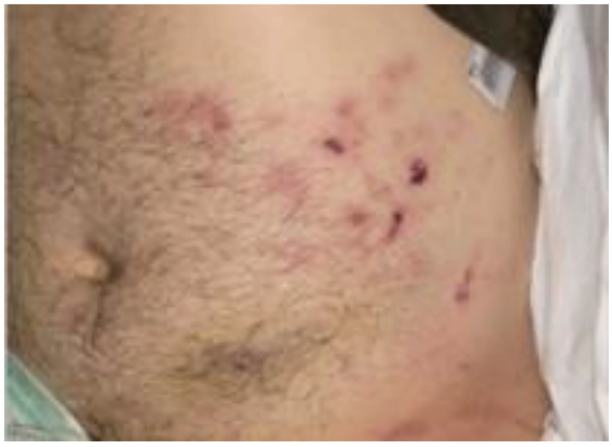

Fig 1. Day 1: Left flank. Evolving dusky changes with tender plaques and few purpuric patches.

In our case, trauma was likely the trigger for developing CAPS; however infection and withholding warfarin were contributing factors. Dedicated input from a range of specialities was paramount in this situation. Current literature suggests rituximab may provide long-lasting remission given its role in preventing

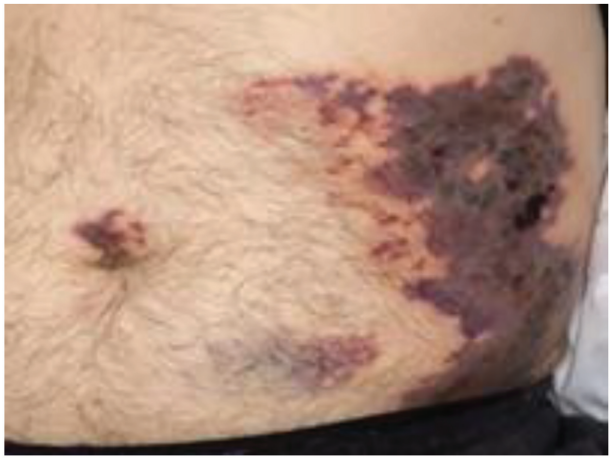

Fig 2. Day 8: Left flank. Gross purpura with necrotic bullae.

Authors: [A]Russells Hall Hospital, Dudley, UK

further production of B-cell driven autoantibodies via CD20 binding; however, more longitudinal research is needed.[3] ■

References

1 Nayer A, Ortega L. Catastrophic antiphospholipid syndrome: a clinical review. *J Neuropathol* 2014;3:9–17.

2 Chaturvedi S, Brodsky R, McCrae K. Complement in the pathophysiology of the antiphospholipid syndrome. *Front Immunol* 2019;10:449.

3 Rymarz A, Niemczyk S. The complex treatment including rituximab in the management of catastrophic antiphospholipid syndrome with renal involvement. *BMC Nephrology* 2018;19:132.