

Paraprotein and Cryoglobulin-Associated Medium-Vessel Vasculitis

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The occurrence of a severe medium-vessel vasculitis in association with Type 1 cryoglobulinemia is rare. Even after a decade of monoclonal gammopathy, paraproteinemia and cryoglobulinemia may contribute to medium-size arteritis and gangrene.

A 50-year-old man with a monoclonal gammopathy of undetermined significance (MGUS) of 10 years' duration developed mild bilateral foot paresthesias. One year later, he presented with crescendo, incapacitating right foot pain. Intense livedo reticularis and dry gangrene of the right forefoot were evident (Figure 1A).

A hypercoagulability laboratory investigation, including antiphospholipid antibodies, was negative. C3 and C4 complement levels were depressed; hepatitis B and C serologies and viral loads were negative. Rheumatoid factor (RF) was similarly negative. Cryoglobulins were detected, with a cryocrit of 4%; serum viscosity was normal. Serum, urine, and cryoglobulin immunofixation electrophoresis revealed 2 dis-

tinct monoclonal IgG- κ bands (Figure 2). Bone marrow examination revealed an IgG- κ -predominant plasmacytosis of insufficient quantity to satisfy criteria for multiple myeloma.

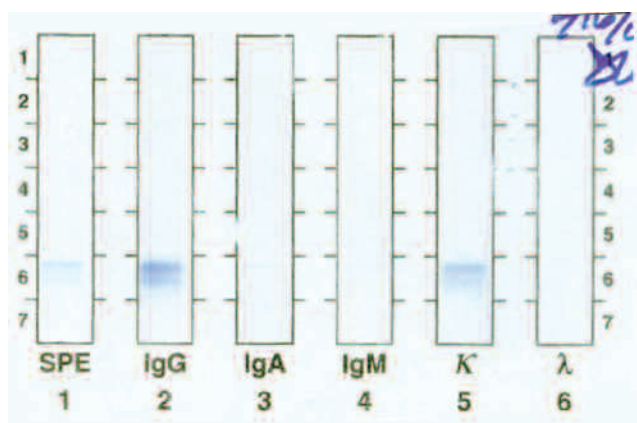


Figure 2. Electrophoresis revealed 2 distinct monoclonal IgG- κ bands.



Figure 1. A. Intense livedo reticularis and dry gangrene of the right forefoot. B. Right limb arteriography reveals smooth tapering and occlusion of the distal anterior tibial and posterior tibial arteries.

Right limb arteriography revealed smooth tapering and occlusion of the distal anterior tibial and posterior tibial arteries (Figure 1B), suggesting an inflammatory process. Consequently, corticosteroid, plasmapheresis, and cyclophosphamide therapy was initiated, although a below-knee amputation was ultimately required. Surgical pathology revealed a medium-vessel arteritis. Postoperatively, the clinical course improved, culminating in resumed ambulation with a prosthetic limb.

Small-vessel disease predominates with cryoglobulinemic vasculitis. In contrast, a medium-vessel phenotype, presenting with digital and extremity gangrene, is distinctly uncommon¹. Moreover, polyarteritis nodosa-like syndromes are reported with multiple myeloma^{2,3}. In this context, this case illustrates the rare occurrence of a severe medium-vessel vasculitis in association with Type 1 cryoglobulinemia (monoclonal, lacking RF ability) and an IgG- κ oligoclonal gammopathy⁴. The case further illustrates how a decade into the course of a

known MGUS, paraproteinemia and cryoglobulinemia may contribute to the extreme phenotype of medium-size arteritis and gangrene.

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