Necrotic lesions of the hands

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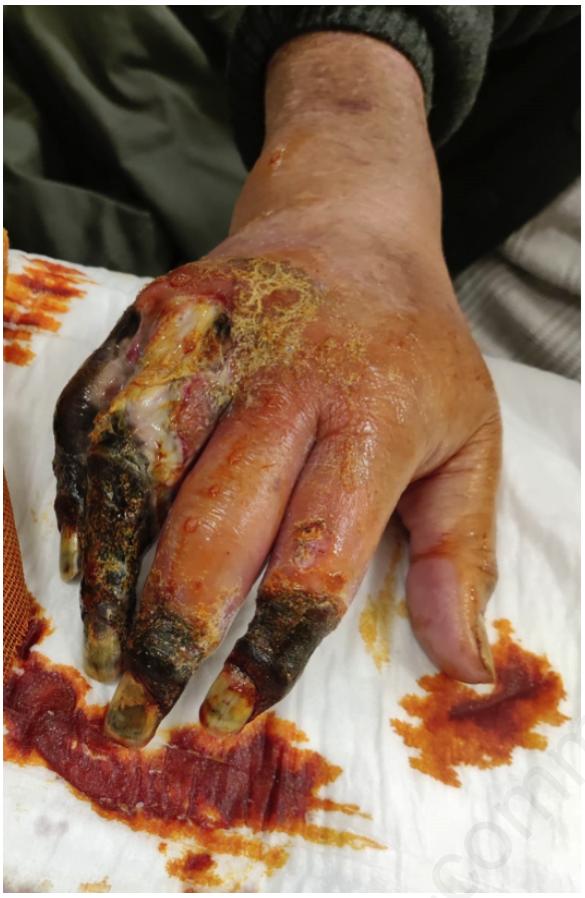

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An 82-year-old man with a history of end-stage renal disease due to glomerulonephritis requiring haemodialysis, hepatitis C-related liver cirrhosis and hypertensive cardiomyopathy presented with painful and necrotic lesions of both his hands. He had no other symptoms. Laboratory results showed leucocytosis (13,000 per cubic millimetre, reference range 4,000 to 10,000) and increased C-reactive protein (5 mg/dL, reference range 0 to 0.5) and procalcitonin values (9.4 ng/mL, reference range < 0.5). Skin lesion culture resulted positive for Klebsiella pneumoniae and Enterococcus faecium. The patient was urgently referred to the vascular clinic and treated with partial bilateral hand amputation and parenteral antibiotic therapy.

Question

Given the patient's history, what is the likely mechanism of his skin lesions?

1. Porphyria cutanea tarda
2. Calcific uremic arteriolopathy (calciphylaxis)
3. Immune Thrombocytopenia Purpura (ITP)
4. Cryoglobulinaemic Vasculitis (CryoVas)

Answer

Crioglobulinaemic Vasculitis (CryoVas) is the most likely. Serum cryoglobulins were detected and a diagnosis of necrotizing CryoVas was done. Cryoglobulins are antibodies that precipitate at low temperatures and dissolve after rewarming. The presence of circulating cryoglobulins is called cryoglobulinemia and it can lead to an inflammatory syndrome of the small and medium-sized blood vessels, characterized by fatigue, arthralgia, purpura, ulcers, neuropathy and/or glomerulonephritis.[1] Cryoglobulinemia is classified into three types (I, II and III) on the basis of immunoglobulin composition.[2] Lymphoproliferative, autoimmune diseases and hepatitis C virus infection are predisposing causes. CryoVas is the most common extrahepatic manifestation in patients with hepatitis C. The diagnosis is based on clinical features and laboratory detection of serum cryoglobulins. The treatment strategy depends on the cause of cryoglobulinemia. Antiviral therapy is indicated in patients with chronic C hepatitis, while immunosuppressive or immunomodulatory therapy, including steroids, plasmapheresis, and cytotoxic agents, is reserved for organ-threatening manifestations.[3] The prognosis is poor, particularly in elderly patients.[4] The main causes of death are renal failure and widespread vasculitis with the involvement of the gastrointestinal system.[5,6] Liver fibrosis at the time of diagnosis is the poorest prognostic factor in patients with C hepatitis.[7]

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