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“Cryoglobulinemic vasculitis in the setting of Sjögren's syndrome: a case report and review of the literature”

Introduction: Cryoglobulinemic vasculitis results from the deposition of cryoglobulins, proteins that precipitate in cold temperatures, within the circulation. The resultant vascular occlusion causes widespread end-organ damage. While more commonly associated with lymphoproliferative or infectious diseases such as HCV and HIV, cryoglobulinemia also occurs in patients with Sjögren's syndrome, a chronic autoimmune disorder affecting the lacrimal and salivary glands.

Case: We present a case of a 48-year-old Caucasian male with a past medical history significant for Raynaud phenomenon, recurrent nephrolithiasis, and episodic urticaria over the last year. He presented to the emergency department with complaints of fever, dyspnea, productive cough, severe non-pitting edema of the left lower extremity, and a non-healing ulcer on the right lower extremity. He subsequently underwent an extensive workup and was diagnosed with Sjögren's syndrome with concomitant cryoglobulinemic vasculitis. His hospital course was complicated by septic and cardiogenic shock, with necrotizing fasciitis requiring a left below-the-knee amputation. While hospitalized, he was treated with broad-spectrum antibiotics, systemic steroids, and plasma exchange therapy. At discharge nearly three months later, he began Rituximab infusions which he continues every six months.

Discussion: Our case report and literature review reveals that patients with primary Sjögren's syndrome (SS) who present at diagnosis with cryoglobulinemic vasculitis are at an increased risk for severe multi-systemic complications involving the cutaneous, renal, glandular, hematologic, gastrointestinal, pulmonary, and peripheral nervous systems. Patients with this diagnosis are also at higher risk of malignancies such as lymphomas and have a higher mortality rate than the average population.

Conclusion: This review should instruct the general practitioner and other subspecialists on the rare and potentially catastrophic complications of Sjögren's syndrome, highlighting that it is more than a disease of "dry eyes and mouth." When synthesizing a differential, physicians should consider the severe manifestations of cryoglobulinemic vasculitis and etiologies beyond the more common infectious and lymphoproliferative diseases.