Systemic vasculitis with dermatomyositis, hearing loss, neuropathy, and multiorgan dysfunction

Figure Nerve and muscle biopsy

Nerve: (A) Hematoxylin & eosin (H&E) stain: inflammatory cells invading an epineurial arteriole; (B) smooth muscle actin immunostain: vessel wall fragmentation; (C) epoxy section: mildly reduced fiber density and epineurial vessel wall infiltration. Muscle: H&E stain: (D) perifascicular fiber atrophy and (F) perimysial perivascular inflammation; (E) myxovirus resistance A (MxA) immunostain: perifascicular sarcoplasmic expression of MxA.¹

A 39-year-old woman presented with 10 months of steadily progressive weakness, myalgia, weight loss, and intermittent feet tingling. During the course of her illness, she developed refractory atrial fibrillation, left followed by right-sided hearing loss, amenorrhea, and hematuria. She had been on rituximab for years for “polyarthralgias,” which was stopped at symptom onset. Examination showed severe proximal weakness and mildly decreased toe proprioception. Deltoid biopsy (figure) was diagnostic of dermatomyositis. Liver and enteric biopsies showed lymphocytosis. Suspicion for vasculitis led to sural nerve biopsy (figure), showing large-arteriole vasculitis. Despite treatment with high-dose prednisone and cyclophosphamide, the patient died. This is a unique case of systemic vasculitis associated with dermatomyositis, hearing loss, neuropathy, and multiorgan dysfunction. The histologic findings and the relentless progression despite stopping rituximab argue against a role for rituximab in the pathogenesis.

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