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Vasculitis Causing Complete Occlusion of Aorta

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Vasculitis Causing Complete Occlusion of Aorta

A 14-year-old boy presented with several months of intermittent abdominal pain and non-bloody diarrhea. He endorsed a 10 kg weight loss in the past year and lower limb claudication. Previously, he had been treated for an unprovoked deep venous thrombosis of uncertain etiology of the left common femoral vein. On physical exam, he had moderate pallor, digital clubbing, oral ulcers, and tender red plaques on his forehead and legs. Pulse was absent below the left popliteal artery and was decreased in the right leg.

Laboratory investigations revealed microcytic hypochromic anemia (Hb 8.1 g/dL), erythrocyte sedimentation rate 90 mm/h, and C-reactive protein 29 mg/dL. Because systemic vasculitis syndromes were being considered, biopsy of the skin lesions was obtained, which showed necrotizing vasculitis involving subcutaneous small and medium-sized vessels (**Figure 1**; available at www.jpeds.com). Magnetic resonance angiogram revealed complete occlusion of the aorta below the renal arteries (**Figure 2**; available at www.jpeds.com), mural thickening of the aortic wall, and collateral vascularization of the lower extremities (**Figure 3**). An extensive infectious

disease and rheumatologic work-up to identify the cause of this vasculitis has remained largely inconclusive.

Complete occlusion of aorta is unusual and suggests that the aortic disease was slowly progressive, allowing collateral vessel development.¹ Arteries of any caliber, ranging from the aorta and its branches to small capillaries, may be affected; however, each vasculitide has a tendency to target specific-sized vessels.^{2,3} He was initially treated with high dose pulse steroids and then transitioned to infliximab based on clinical response.⁴ Two years after presentation, the patient remains relatively asymptomatic with persisting aortic occlusion and resulting mild claudication. ■

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Figure 3. Magnetic resonance angiogram showing complete occlusion of the aorta and collateral vascularization of the lower extremities.

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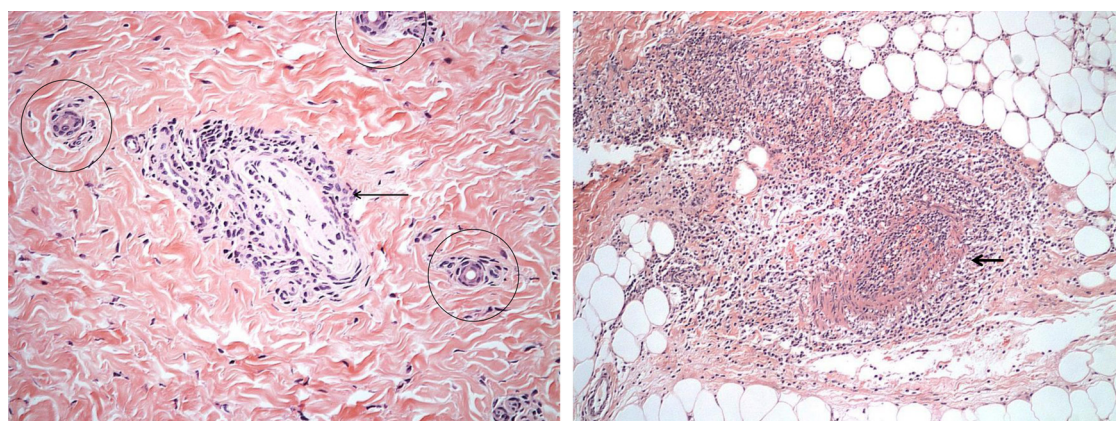


Figure 1. Biopsy of the skin lesions showing necrotizing vasculitis involving subcutaneous small (*circles*) and medium-sized vessels (*arrows*).



Figure 2. Magnetic resonance angiogram showing complete occlusion of the aorta below the renal arteries.