Fatal Occlusive Vessel Disease in a Patient with Systemic Juvenile Idiopathic Arthritis

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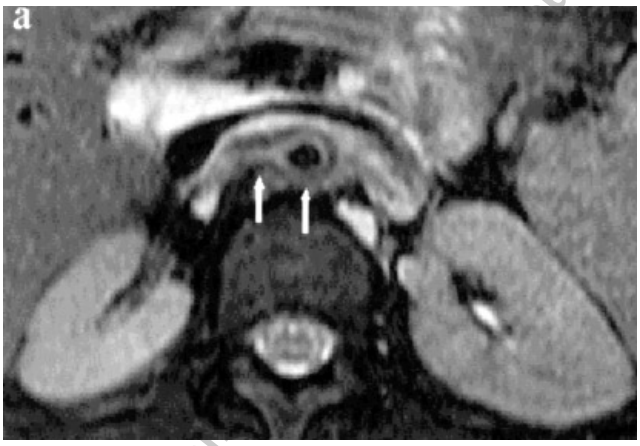
A boy aged 3 years presented with spiking fever and rash, followed by polyarthritis suggesting systemic juvenile idiopathic arthritis (JIA). HLA-B27, rheumatoid factor, and antinuclear antibodies were negative. With therapy of corticosteroids and nonsteroidal antiinflammatory drugs the systemic symptoms subsided within the first year of disease, but the polyarthritis progressed. At the age of 6, low dose methotrexate (MTX) was started, but was ineffective, as was addition of leukerane. At the age of 16, leflunomide led to a sustained remission, but was stopped after 12 months because of side effects. Infliximab (Remicade®) was only transiently effective, and was stopped after 8 months, and MTX was continued. Eight months later, he developed neurologic symptoms with loss of consciousness due to hypertensive encephalopathy.

Baseline inflammatory activity indicated by C-reactive protein was increased. Magnetic resonance angiography revealed segmental high-grade stenoses of the renal arteries and the infrarenal aorta, and occlusion of the common iliac arteries (Figure 1). The superior mesenteric artery, both renal arteries, and the aorta showed segmental concentric wall thickening (Figure 1b). Negative antinuclear cytoplasmic antibodies, normal D-dimer, complement factors C3 and C4, and immune complexes argued against a diagnosis of vasculitis but it could not be ruled out. A rectal biopsy revealed mild amyloidosis.

With intensive care management the hypertension was controlled, and the neurologic symptoms waned. Cyclophosphamide for amyloidosis and interventional dilatation of vessel obstructions were refused, and the patient was discharged. Eight weeks later, aged 19 years, he collapsed at home and died immediately, which may suggest a myocardial infarction. Autopsy was refused by the parents.

To our knowledge this is the first case of fatal obstructive vessel disease in a patient with systemic JIA.



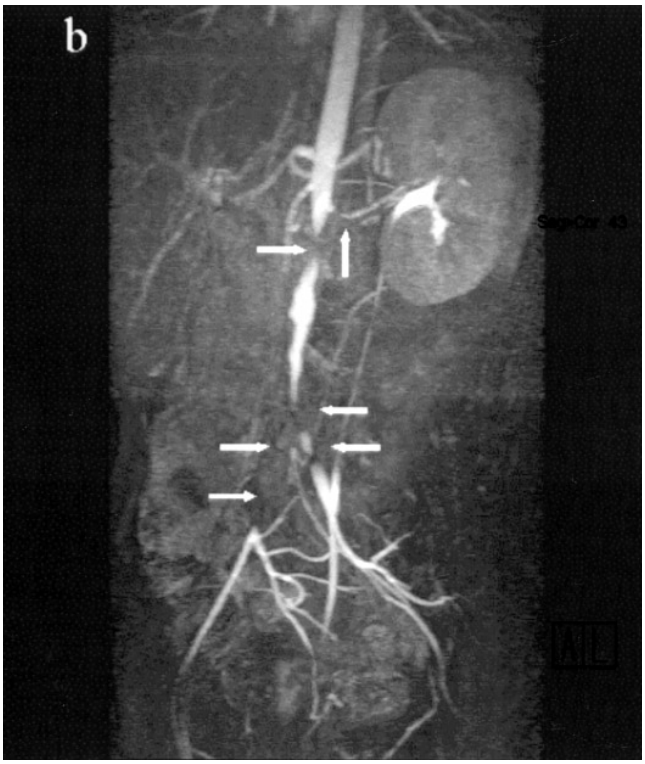


Figure 1. A. Axial T2 weighted magnetic resonance image at the level of the renal arteries. Note the concentric wall thickening of the aorta and the renal arteries (arrows) with a hyperintense inner layer. B. Oblique view of 3D-reformatted contrast-enhanced MR angiography showing segmental high-grade stenoses of the renal arteries and the infrarenal aorta (arrows). The common iliac arteries are occluded bilaterally (arrows).