Renal Infarction Due to Spontaneous Renal Artery Dissection in Ehlers-Danlos Syndrome Type IV

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To the Editor:

Ehlers-Danlos syndrome (EDS) type IV is an autosomal dominant disorder caused by mutations in the gene for type III procollagen (COL3A1). The phenotype is characterized by thin translucent skin with visible veins, spontaneous rupture of arteries, uterus or intestines, characteristic facial features, and easy bruising. Median survival is 48 years, with spontaneous arterial rupture the commonest cause of death. We describe a case of renal infarction due to spontaneous renal artery dissection in EDS, highlighting the clinical presentation and management difficulties.

A 25-year-old man presented to an outlying facility in our hospital network with a 6-h history of acute severe right flank pain associated with 2 episodes of vomiting. Medical history was notable for a diagnosis of EDS type IV, diagnosed by dermal fibroblast culture at the age of 20 years. He dislocated his left shoulder at the age of 21 years and had a pneumothorax at age 22 years. His mother and brother had also been diagnosed with EDS type IV. He was a nonsmoker and denied cocaine use. Examination revealed the characteristic facies of his condition: translucent skin with visible veins, multiple intradermal pressure-point bruising, and mild hypermobility. Abdominal examination revealed tenderness in the right iliac fossa without guarding or rebound tenderness. Laboratory tests showed an elevated white blood cell count at 13.1 x 10^9/l, with neutrophilia 11.2 x 10^9/l. C-reactive protein was 8.5 mg/l. Hematology and biochemistry were otherwise normal. Autoantibody and thrombophilia screens were normal, as were chest radiograph, electrocardiogram, transthoracic echocardiogram, and urinalysis. Computed tomography of the abdomen demonstrated multiple areas of renal infarction in the right kidney, predominantly in the lower pole but also involving the upper pole. Cortical involvement was estimated at 30%. Dimercaptosuccinic acid scan demonstrated minimal if any uptake in the lower pole of the right kidney, with faint uptake in the upper pole. Relative uptake was estimated at 23% for the right kidney and 77% for the left kidney. Magnetic resonance angiography confirmed the renal infarctions (Figure 1A) and demonstrated right renal artery dissection (Figure 1B, Figure 2). Initial treatment consisted of analgesia, intravenous antibiotics, and unfractionated heparin. Following transfer to our center, anticoagulation was added to the differential diagnosis. To our knowledge this is the first report of a case of renal artery dissection presenting with renal infarction in EDS. The optimum management of renal infarction in Ehlers-Danlos syndrome type IV remains to be determined.

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Figure 1. Coronal magnetic resonance renal angiogram demonstrating nonenhancing renal parenchymal infarcts (A, arrows) and right renal artery dissection (B, arrow).
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REFERENCES


Figure 2. Magnetic resonance imaging abdominal aortic angiogram showing right renal artery dissection (arrow).