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On the Cover

What's the diagnosis?

A 10-year-old girl was incidentally found to have severe hypertension (180/100 mmHg) while undergoing evaluation for right-sided hemihypertrophy. She was otherwise well and had no complaints. Family history was unremarkable. Physical exam was notable for coarse facies, enlarged right lower extremity, and paraumbilical bruit. Echocardiogram demonstrated mild left ventricular hypertrophy. C-reactive protein, serum aldosterone, plasma renin activity, serum creatinine, alpha-fetoprotein, and plasma metanephrines were normal. Kidney doppler sonography demonstrated elevated peak renal artery systolic velocities bilaterally with no parvus et tardus waveform. Abdominal computed tomography angiography confirmed the diagnosis.

Image Description:

Lateral view demonstrates a long segment of irregular stenosis of the mid abdominal aorta extending from below the diaphragm to the level of takeoff of the inferior mesenteric artery with focal stenoses at the origin of celiac trunk, superior and inferior mesenteric arteries. AP view demonstrates left renal artery stenosis, irregular stenosis at the aortic bifurcation, long segment stenosis of right internal iliac artery, and occlusion of left common iliac artery. Large splanchnic, lumbar and pelvic collaterals seen on both views provide compensatory flow.

Teaching Points:

- She was diagnosed with idiopathic midaortic syndrome and treated with 4 anti-hypertensive medications.
- Midaortic syndrome is the acquired or congenital narrowing of the abdominal aorta and associated branches. While congenital narrowing is due to incomplete or overfusion of the embryonic dorsal aortas, acquisition of the vascular anomalies postnatally has been linked to Williams syndrome, giant cell arteritis, mucopolysaccharidosis, fibromuscular dysplasia, neurofibromatosis, Alagille syndrome, and Moya-Moya disease. Many cases are idiopathic.
- Patients often present prior to adolescence with severe hypertension, headaches, claudication, or abdominal angina.
- Treatment involves antihypertensive medications and often requires surgery, including balloon angioplasty, vessel stenting, aorto-aortic bypass, patch aortoplasty, and aortic lengthening.
- Left untreated, patients usually die by their 40s from hypertensive complications.

(Images and text provided by Rachel Herdes, Cullen Clark, Nikita Patel, and Isa Ashoor, Louisiana State University Health Sciences Center, and Ewa Wasilewska, Tulane University School of Medicine)