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# Images in vascular medicine

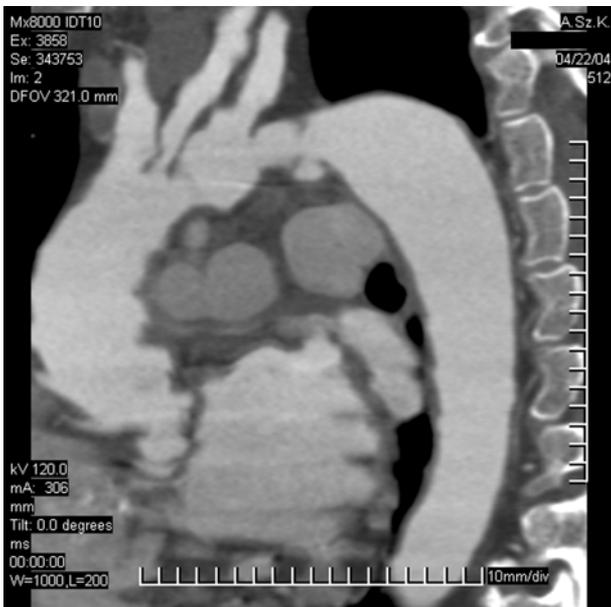
## Atypical aortic coarctation

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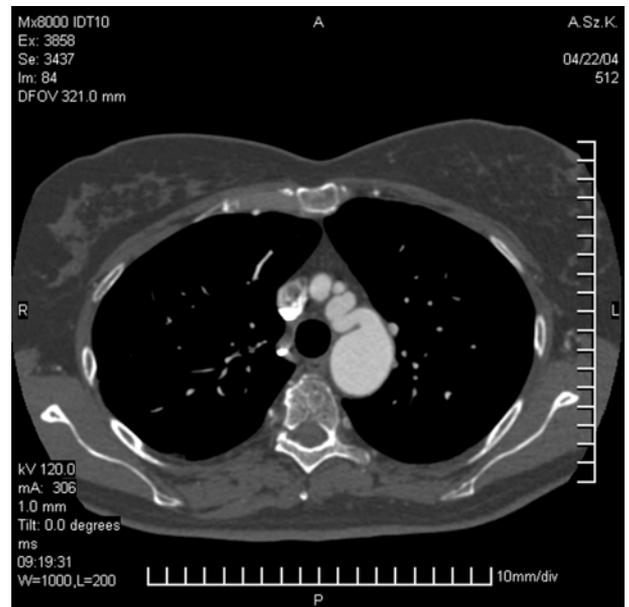
A 67-year-old woman with a 30-year history of hypertension was referred to our department for poorly controlled blood pressure and unstable angina (CCS II/III). A physical examination revealed no significant abnormalities. The systolic arterial pressure on the left arm was 15 mmHg lower than that measured on the right arm. The patient underwent coronary angiography in our institution. Cardiac catheterization from femoral access was impossible due to an inability to cross the aortic arch. Successful catheterization was achieved through the right radial artery. Coronary angiography revealed disseminated atherosclerotic lesions with multiple stenoses up to 50%. Simultaneous aortography demonstrated an atypical long narrowing of the aortic arch. An angio-CT revealed the normal proximal part of the ascending aorta. Behind the origin of the brachiocephalic trunk, atypical constriction of the arch of aorta

with winding course and numerous angular bends was seen (Panels A and B). The constriction involved the whole length of the arch behind the take-off of the brachiocephalic trunk. The Botall duct was not visualized. Post-stenotic dilatation of up to 3.5 cm of descending aorta was present (Panel A). The classic pattern of collateral circulation was not visible. The patient did not consent to surgical reconstruction.

Coarctation of the aorta may be present in about 0.2% of cases of hypertension. Atypical aortic coarctation is very rare and found in 0.5–2% of patients with aortic coarctation. Causes of atypical coarctation of the aorta include developmental vascular abnormalities, genetic disorders (Williams syndrome, neurofibromatosis type I, Turner's syndrome), fibromuscular dysplasia, large vessel vasculitides (especially Takayasu disease) and atherosclerosis. Diagnosis of neurofibromatosis type I,



Panel A



Panel B

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Williams or Turner's syndromes is unlikely (lack of cutaneous neurofibromas and normal physical examination). Atherosclerotic disease of the aorta can be a cause of aortic coarctation; however, published reports usually describe atherosclerotic coarctation being due to heavily calcified, 'coral reef'-type lesions. Morphological features of fibromuscular dysplasia in CT angiography (beads on a string appearance, a long-segment tubular

stenosis), absence of clinical signs and symptoms of inflammatory process, lack of aortic calcifications and smooth lumen of the aorta may support the diagnosis of fibromuscular dysplasia rather than aortitis (eg Takayasu disease) or atherosclerosis. However, without histopathological verification we cannot rule out atherosclerotic disease, aortitis or developmental abnormality.

'Images in vascular medicine' is a regular feature of *Vascular Medicine*. Readers may submit original, unpublished images related to clinical vascular medicine to: Mark A Creager, Editor in Chief, *Vascular Medicine*, Brigham and Women's Hospital, 75 Francis Street, Boston, MA 02115, USA.