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The Case | An unusual cause of renovascular hypertension

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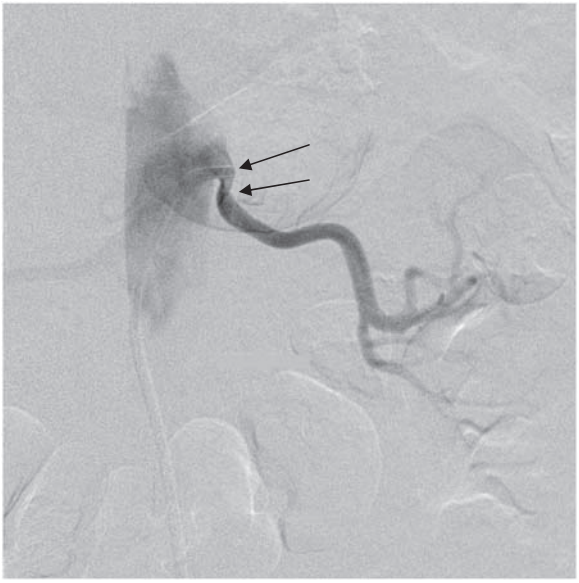

Figure 1 | Conventional renal angiography: high origin of the left renal artery (RA; 15 mm higher than the right) that was 50% stenosed; the course of the left RA along the aorta (arrows).

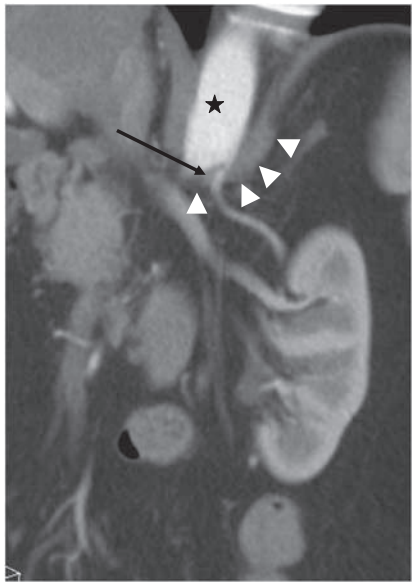

Figure 2 | Computed tomography scan: coronal curved multiplanar reconstruction image of the left RA (black arrow), which shows arterial entrapment between the aorta (black star) and the left diaphragmatic crus (white arrowheads).

A 48-year-old obese male (body mass index, 33), who was an active smoker, was admitted to our department to undergo percutaneous angioplasty of a markedly stenosed left renal artery. Six months earlier, severe hypertension (blood pressure, 200/100 mm Hg) associated with hypokalemia of renal origin (serum potassium level, 3.1 mmol/l; urinary potassium, 75 mmol/day) and stage III chronic kidney disease (serum creatinine level, 146 μmol/l; estimated glomerular filtration rate, 48 ml/min per 1.73 m^2) were discovered. Doppler ultrasound showed 90% stenosis of the ostium and the first segment of the left renal artery. The right renal artery was normal. Both kidneys were 100 mm in length. Magnetic resonance angiography confirmed the Doppler results and the decision to perform a percutaneous renal angioplasty was made.

On admission, the patient's blood pressure was 135/75 mm Hg on three medications including nebivolol (5 mg/day), amlodipine (5 mg/day), and spironolactone (50 mg/day). Physical examination was unremarkable. Laboratory findings were as follows: hypertriglyceridemia at 7.5 mmol/l, hypercholesterolemia (HDL cholesterol at 0.61 mmol/l and LDL cholesterol level at 4.97 mmol/l), serum potassium at 3.5 mmol/l, and serum creatinine at 147 μmol/l. Urine analysis was normal. A renal angiography was performed (Figure 1). An unusual finding led us to perform a computed tomography scan angiography (Figure 2).

What abnormality do you see in the left renal artery and in the CT angiography? What is the cause of this patient's hypertension?

SEE NEXT PAGE FOR ANSWERS

The Diagnosis | Renal artery entrapment by the diaphragmatic crus

Conventional renal angiography findings (Figure 1) supported the diagnosis of extrinsic renal artery compression. Multidetector computed tomography (MDCT) angiography (Figure 2) was performed and revealed that diaphragm muscle fibers caused a verticalization of the root of the left renal artery and lead to a stenosis at its origin. The artery follows an unusual course at an acute angle, which gives it a sigmoid course. MDCT showed compression of the left renal artery by the diaphragmatic crus. This is known as renal artery entrapment (RAE) syndrome.

Renovascular hypertension is the cause of hypertension in 5% of cases. The most common causes of renovascular hypertension are atherosclerotic stenosis (65% of cases) and fibromuscular dysplasia (16% of cases).[1] Renal artery aneurysm, arteriovenous fistula, and extrinsic compression are rather rare causes of renovascular hypertension.[1] Renal artery compression leading to hypertension has been attributed to abdominal aortic aneurysm, tumor, hypertrophic adrenal tissue, psoas muscle band, sympathetic nerve fibers, and compression by a diaphragmatic crus.[2]

Since the first case reported by D'Abrieu, several cases of RAE by the diaphragmatic crus have been reported in the literature.[2] Abnormalities such as abnormal musculotendinous fibers, high ectopic renal artery origin, and hypertrophic diaphragmatic crus were found to be responsible for these entrapments.[2] They occur because of the compression of the renal artery by fibers from the crus of the diaphragm and can be easily diagnosed by helical or MDCT angiography.[2] The diaphragmatic crura arise from the anterior surface of the L1 to L4 vertebral bodies on the right and from the first L2 or L3 vertebral bodies on the left. These muscle fibers can, in some cases, cause a verticalization of the root of the renal artery and lead to a stenosis, usually at the origin of the artery. Subsequently, the artery follows an unusual course at an acute angle, which gives it a sigmoid course.[2,3]

Doppler ultrasound permits the visualization of renal blood flow during a complete respiratory cycle. Doppler ultrasound evaluating the blood renal flow during both inspiration and expiration could help in the diagnosis. Flow demodulation over the arterial ostia and increase of flow velocities during expiration and normalization in inspiration provided hemodynamic information highly suggestive of RAE, which could confirm the diagnosis. These findings must be corroborated by CT angiography.[4] CT angiography, especially one using high spatial resolution, permits visualization of the vascular anatomy (aorta and its branches) and diaphragm relationship.

There is no established protocol for the treatment of the RAE syndrome. Although surgery and stenting have been used, both were associated with surgical morbidity and stent-related complications such as stenosis and occlusion. Botulinum toxin type A injection has shown to be useful in one human case[5] and in several rabbit studies. Treatment options were discussed between the nephrology, vascular interventional radiology, and surgery teams and were presented to our patient. Options discussed included medical treatment, stent placement, and aortorenal bypass. The decision made was to continue medical treatment and monitor the patient's blood pressure closely. If the case had involved prolonged uncontrolled hypertension, more invasive treatment would have been considered.

In summary, renal artery may be entrapped by diaphragmatic crus and cause renovascular hypertension. RAE should be suspected if angiography shows a renal artery parallel to the aorta in the proximal part of its course. Thin fibrous bands from the diaphragm insertion are well shown by the non-invasive MDCT angiography. This pathology, unlike common renal artery stenosis, requires specific therapeutic management, including renal artery decompression and/or aortorenal bypass.

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