Fibromuscular dysplasia presenting as macroscopic hematuria and radiologically sporting a 'clover leaf appearance'

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Fibromusular dysplasia can present with renovascular hypertension in the young.[1] Data from our geographical area on this are limited. We report two patients of fibromuscular dysplasia with uncommon features: Both of them were male patients, presented with recurrent macroscopic hematuria, and had multiple saccular aneurysms of renal artery, with clover-leaf appearance in one.

First case is a 29-year-old male patient diagnosed to have accelerated hypertension when he presented with recurrent episode of headache in a local hospital. He was referred to our department for further evaluation of hypertension. His family history was non-contributory. His blood pressure was 200/110 mmHg in right upper limb in supine posture. His systemic examination was unremarkable and epigastric bruit was absent. Urinalysis and renal functions were normal. Ultrasound examination showed asymmetric kidneys (Rt 8.5 cm and Lt 12.5 cm) and Doppler studies did not show any signs of renal artery stenosis. He was treated with telmisartan, indapamide, amlodipine, and clonidine. In view of uncontrolled blood pressure, we proceeded with computed tomography (CT) renal angiogram, and Tc diethylene triamine pentaacetic acid (DTPA) captopril renogram. CT renal angiogram showed short segment narrowing of the mid third of the right renal artery with a clover leaf saccular aneurysm in the distal third near the hilum [Figure 1]. Digital subtraction angiography (DSA) confirmed the CT angiogram pictures, and showed clover leaf aneurysmal outpouchings at distal right renal artery [Figure 2a and b]. Captopril renogram showed prolonged time to max in right kidney and significant reduction in split renal function of right kidney, following captopril. The patient underwent surgical excision of aneurismal segment along with great saphenous vein interposition grafting. Histopathology showed medial fibroplasias. He is now on regular follow-up, and his blood pressure is currently under control with single antihypertensive Telmisartan.

Second case is a 36-year-old male patient, who presented with history of three episodes of macroscopic hematuria. On examination, his blood pressure was 160/120 mmHg. His systemic examination was unremarkable. Ultrasonogram showed right kidney 8.5 cm and left kidney 9.3 cm with normal echogenicity. Doppler studies of renal artery showed peak systolic velocity (PSV) >180 cm/s with parvus tardus pattern in right main renal artery, which is suggestive of renal artery stenosis. The CT angiogram showed bilateral string of beads appearance of main renal arteries [Figure 3a]. Right renal artery showed multiple saccular aneurysm with significant stenosis about 22 mm from the ostium [Figure 3b]. These findings were consistent with fibromuscular dysplasia involving both renal arteries. Tc DTPA renogram showed prolonged time to max in right kidney, and significant reduction in split renal function of right kidney following losartan. The blood pressure was controlled with two antihypertensive hydrochlorothiazide and nifidipine. Though surgical correction was advised, the patient was not willing. He is now on regular follow-up with medical management.

Hypertension is the most common clinical manifestation of renal FMD. Renal FMD is associated with microscopic hematuria in about 50% of cases[2] Notably, both our patients are males. The usual age group is between 20 and 60 years. Renal artery is involved in 60-75% of cases of FMD, followed by internal carotid arteries and less

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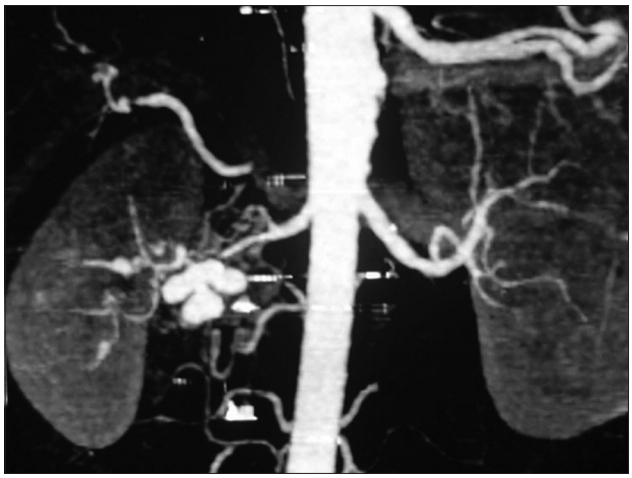

Figure 1: Computer tomography angiogram showing short segment narrowing of the mid third of the right renal artery with a clover leaf saccular aneurysm in the distal third near the hilum

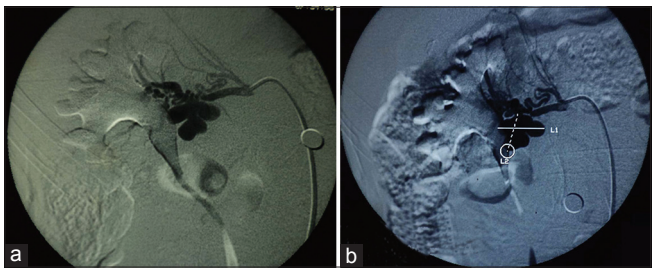

Figure 2: Digital subtraction angiography showing clover leaf aneurysmal outpouchings at distal right renal artery

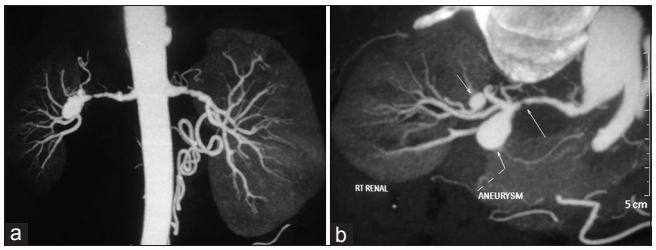

Figure 3: (a) Bilateral string of beads appearance of main renal arteries. (b) Multiple saccular aneurysms of right renal artery

often vertebral, mesenteric, brachial, and very rarely coronary artery. About 30% of patients have multiple artery involvement[3] Most common type is medial fibroplasias which accounts for about 75-80% of cases. It is characterized by classic 'string of beads' appearance due to alternating fibromuscular webs and aneurysmal dilatation. In perimedial fibroplasias, the beads are less in number and will not be greater than the caliber of the original vessel. In medial hyperplasia, there will be concentric hyperplasia of muscles without fibrosis. Intimal fibroplasia is due to deposition of collagen in intima. In adventitial fibroplasias, the normal loose connective tissue of adventitia is replaced by dense collagen. The angiographic appearance of medial hyperplasia, intimal fibroplasias, and adventitial fibroplasias are similar and show smooth tubular stenosis. Even though Doppler is considered as an initial screening modality, normal Doppler studies may not exclude renal artery stenosis as documented in our first patient.

DSA is currently the gold standard for evaluating renal artery FMD.[4] The classical angiographic sign is 'string of beads appearance'. Both our patients had multiple saccular aneurysms of renal artery, with 'Clover Leaf Appearance' appearance in one. The optimal management of hypertension with medical versus revascularization either by surgery or angioplasty depends on patient's clinical condition.

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