Surgical revascularization of bilateral renal artery stenosis due to fibromuscular dysplasia

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Case Report

INTRODUCTION
Fibromuscular dysplasia (FMD) is a noninflammatory disease affecting small- and medium-sized arteries of the renal and the carotids. It affects the renal arteries in nearly 60%–75% cases. The primary clinical manifestation of renal FMD is hypertension. Medial fibroplasia represents the most common dysplastic lesion. We report two cases who presented with hypertension and renal insufficiency and on evaluation was found to have bilateral renal artery stenosis. Stenting of the renal vessels was not possible due to the narrowed caliber of the vessel and inability to cannulate the renal arteries. They underwent renal artery revascularization with a splenorenal end to end anastomosis. The renal parameters and blood pressure of both the patients stabilized subsequently. Renal revascularization can be a good option for patient having failed angioplasty with stenting.

Key Words: Fibromuscular dysplasia, renal artery stenosis, surgical reconstruction

CASE REPORT
First patient
A 29-year old gentleman had acute episodes of breathlessness, vomiting, headache, and decreased urine output 18 months before presentation. He denied a history of photophobia, fever, fatigue, joint pain, weakness or skin rashes on the face, neck, and upper body. There were no symptoms suggestive of Raynaud’s phenomenon. He did not have palpitation, sweating, syncope, muscle weakness, mood changes, or weight gain. On evaluation, elsewhere, he was found to have bilateral chest crepitation, raised blood pressure (BP), and serum creatinine—11 mg%. Rest of the physical examination were normal. He underwent four sessions of hemodialysis. He required five antihypertensives (torsemide/clonidine/metoprolol/prazosin/nifedipine) for control of BP. His hemoglobin (Hb)-10.9 g%, urea-45 mg%, serum creatinine - 1.92 mg%, 24 h urine protein-745 mg. Additional screening for vasculitis, systemic lupus erythematosus, cushing, and pheochromocytoma were negative.
His BP on five antihypertensives was 200/100 mmHg. Computed tomography angiography showed bilateral renal artery stenosis (RAS) with multiple collateral vessels on both sides. Stenting of the renal vessels was not possible as the ostial lumen was narrowed and guide wire could not be passed on angiography [Figure 1].

He initially underwent left renal revascularization with a splenorenal end to end anastomosis.

Subsequently, he had a right iliac fossa renal autotransplantation 1 month later.

Postoperative renogram showed a good perfusion of both the kidneys [Figure 2].

His serum creatinine was 1.01 mg%, urea: 36 mg%, 24 h protein: 146 mg%, hemoglobin: 16 g%, serum electrolytes: normal, and BP: 140/90 mmHg on two antihypertensives (metoprolol and prazosin) on follow-up.

Second patient
An 18-year old gentleman was diagnosed to have bilateral RAS while being evaluated for progressive vision dimness and hypertension. He had no skin infection, rash, arthritis, hemoptysis, lower urinary tract symptoms, or symptoms suggestive of Raynaud’s syndrome. He underwent right renal artery stenting followed by left renal artery angioplasty and stenting within a gap of 6 months. His serum creatinine was 1.48 mg%, serum urea - 24 mg%, Hb - 10.3 g%, 24 h urine protein - 3.7 g%, and BP - 140/90 mmHg on five antihypertensives. His BP continued rise for which right renal artery stenting was reattempted after a year. This was followed by right laparoscopic nephrectomy for nonfunctioning right kidney. Histopathology showed right hilar renal artery with medial fibroplasia.

His BP progressively rose and had several episodes of generalized tonic–clonic seizures. Imaging revealed worsening of left RAS [Figure 3]. He underwent end to end left lienorenal arterial revascularization under general anesthesia.

Postoperative blood findings serum creatinine - 0.89 mg%, urea - 15 mg%, 24 h urine protein - 271 mg%, Hb - 11.9 g%, S. electrolytes - normal. He was on three antihypertensives (prazosin/cilnidipine/bisoprolol) at time of discharge and his BP was 130/80 mmHg.

Operative procedure: the patient was placed in right lateral with the operation table flexed. A lumbar incision was made and 11th rib excised. Gerota fascia opened, the renal hilum exposed anteriorly, and the renal vein was identified. The renal artery was spotted posteriorly. The splenic artery discerned and isolated, the distal splenic artery was left untouched. The splenic artery was divided after applying vascular bulldog clamps and spatulated. The renal artery was divided till there was adequate lumen. Splenorenal anastomosis was done with interrupted 7-0 prolene. Hemostasis achieved and abdomen closed in layers.
DISCUSSION

Fibromuscular dysplasia (FMD) is a nonatheromatous disease affecting the small- and medium-sized arteries. It affects mostly the renal and internal carotid arteries. FMD is very often seen affecting the renal arteries, 60%–75% cases. The prevalence of FMD is 4.4%–6.6%, most commonly in females. It is most commonly seen in patients younger than 50 years of age. Medical fibroplasia represents the most common dysplastic lesion. FMD of the renal arteries is bilateral in 35%–50% cases. A variety of mechanical, genetic, and hormonal factors have been proposed, but the exact cause of FMD remains elusive. Manifestation of disease may result due to ischemia resulting from stenosis, spontaneous dissection of arteries, rupture of aneurysms, or embolization of intravascular thrombi from aneurysmal segments. The FMD progresses in up to 37% of patients. The primary clinical manifestation of renal FMD is hypertension; however, it accounts for <10% of cases of renovascular hypertension. Nearly 63% of patients with RAS presents with renal failure. Other symptoms such as fever, flank pain, vomiting, and oliguria also are seen. Intraarterial digital subtraction angiography remains the gold standard for exclusion or confirmation of RAS caused by FMD.

The principal aim for the treatment of FMD is control of hypertension and its complications. Most of these patients can be managed medically. Revascularization is reserved for those patients with recent onset of hypertension (<1 year), in whom BP control has proved difficult, intolerant of antihypertensive therapy, not compliant with antihypertensive medication and who exhibit loss of renal volume leading to a diagnosis of ischemic nephropathy. The primary mode of intervention is balloon angioplasty, surgery reserved for refractory cases.

It has been showed in the CORAL study that patients with 70%–80% atherosclerotic RAS, stable renal function, and up to two antihypertensive drugs should not undergo renal artery stenting since there is no incremental benefit.

Hypertension cure rate of 14–59% and improvement of BP in 21%–63% have been reported with percutaneous transluminal renal angioplasty (PTRA). With open surgery, cure of hypertension can be obtained in 33%–63% of patients, with improvements in BP in 24% to 57%. Successful outcome seems to be associated with age <50 years, absence of associated carotid and coronary stenosis, <8 years of hypertension.

According to Caring for Australasians with Renal Impairment guidelines correction of RAS, either by revascularization surgery or percutaneous methods, including stenting, has been shown to be effective in treating hypertension associated with RAS. They advocated procedures to correct RAS in patients who fail medical therapy with refractory or poorly controlled hypertension, dialysis-dependent renal failure resulting from RAS, recurrent flash pulmonary edema, chronic renal insufficiency, bilateral RAS or RAS to a solitary functioning kidney.

Patients who undergo revascularization should undergo duplex ultrasound regularly. Imaging should be performed soon after revascularization then 6 monthly or whenever there is recurrence or worsening of hypertension.

Surgical revascularization procedures such as aortorenal bypass, branch artery reconstructions, and autotransplantations are reserved for patients with complications or failure of PTRA. Complications of surgery such as urinary tract infections, postoperative pneumonia, postoperative occlusion of the graft vessels, and pseudoaneurysms can occur. In a systemic review and meta analysis conducted by L. Trinquart et al. in which they reviewed 23 surgery series (1014 patients) and concluded that angioplasty or surgical revascularization yielded moderate benefits in patients with fibromuscular dysplasia renal artery stenosis. This highlights the relative rare incidence of this procedure being performed.

CONCLUSION

Renal revascularization (lienorenal anastomosis) is a good option for patient having failed angioplasty with stenting on the left side.

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Conflicts of interest
There are no conflicts of interest.

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