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CASE REPORT

Fibromuscular dysplasia in an adult male as a cause of renal artery stenosis and secondary hypertension treated with renal artery stenting

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KEYWORDS

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Abstract *Background:* Renovascular hypertension due to fibromuscular dysplasia is an uncommon cause of secondary hypertension and is more common in females. This entity is an important treatable cause of secondary hypertension.

Case presentation: We report the case of a 21-year-old asymptomatic male found to have high blood pressure on routine checkup. Renal angiogram revealed fibromuscular dysplasia involving the right renal artery. He underwent percutaneous angioplasty with complete recovery. The single antihypertensive which he was on was stopped next month.

Conclusion: Fibromuscular dysplasia causing stenosis of renal artery is uncommon. High degree of suspicion is required for the timely diagnosis and treatment of this potentially treatable cause of secondary hypertension

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1. Introduction

Renovascular hypertension is one of the most common treatable causes of secondary hypertension. The incidence depends upon the clinical presentation with ranges from $<1\%$ in cases of mild hypertension[1] to 10 and 45 percent of white patients with severe or malignant hypertension.[2] The mechanism of hypertension is activation of the renin angiotensin aldosterone system.[3] The characteristic features suggesting renovascular hypertension are hypokalemia, young age of onset and renal bruit.[3] The most common cause of renal artery stenosis (RAS) is atherosclerosis of the renal artery (75% of all cases).[4] Among the patients with renovascular hypertension, fibromuscular dysplasia (FMD) constitutes 35 to 50 percent of cases in children and 5 to 10 percent of cases in adults under the age of 60 years.[5,6] This etiological diagnosis is particularly important because the goal of treatment is cure from hypertension. In fibromuscular dysplasia, the hypertension is cured or improved in majority of the patients in contrast to atherosclerotic renal artery stenosis.[7,8]

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2. Case report

We present a case of 21-year-old adult male who was found to have high blood pressure during routine checkup. He did not have any symptoms prior to the hospital visit. There was no family history of hypertension or familial dyslipidemia. His maximal recorded blood pressure was 230/140 mmHg in both the arms. All peripheral pulses were palpable with no radio-radial or radio-femoral delay.

General physical examination did not reveal any abnormality. His body weight was normal with Body Mass Index of 23.5 kg/m^2. The precordial, respiratory and neurological system examinations were normal. There were no features of hypo- or hyperthyroidism and Cushing's disease. On auscultation, there was an audible renal artery bruit. Routine blood investigations including urea, creatinine and serum electrolytes were normal. His chest X-ray, ECG and echocardiogram were normal.

We started him on standard anti-hypertensive medications and did further investigations. As renal artery stenosis was suspected from clinical examination, renal Doppler was performed, which did not reveal any renal artery stenosis but there was significant discrepancy in the sizes of the right and left kidneys. The right kidney was 8 cm and left kidney 10 cm in size with normal cortico-medullary differentiation. Because of low sensitivity of the renal Doppler examination, arterial stenosis was not ruled out. Since clinical examination and different kidney sizes were in favor of renal artery stenosis, we decided to go for invasive renal angiogram.

Renal artery angiogram of right renal artery revealed multifocal renal FMD with angiographic appearance of strings of beads (Fig. 1). The left renal artery was normal (Fig. 2). Subsequently, the patient underwent balloon angioplasty but due to suboptimal dilatation (Fig. 3), stenting of right renal artery was performed (Fig. 4). His blood pressure was controlled with only one antihypertensive medication, which was also stopped after one month. There were no procedural complications and his hospital stay was uneventful.

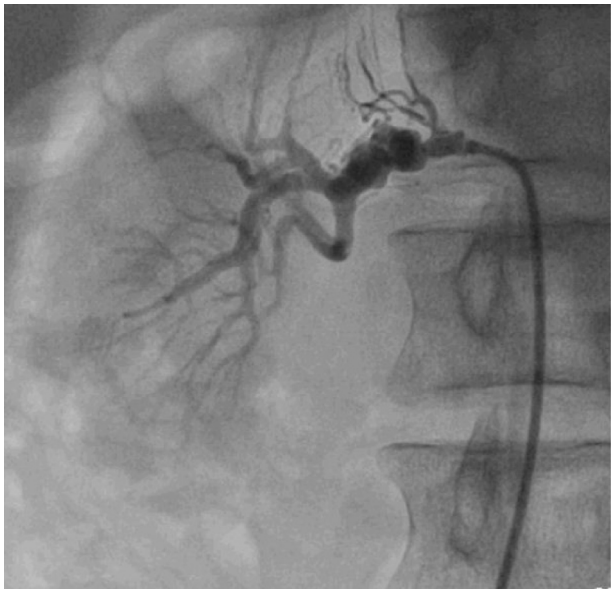

Figure 1 Right renal angiogram.

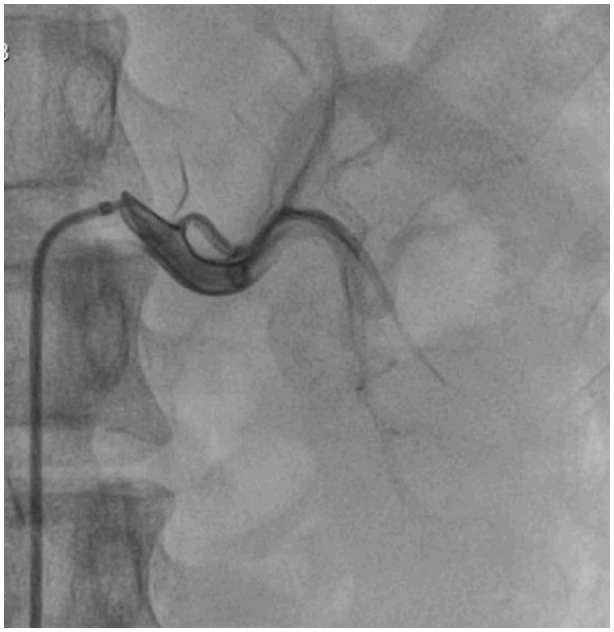

Figure 2 Left renal angiogram.

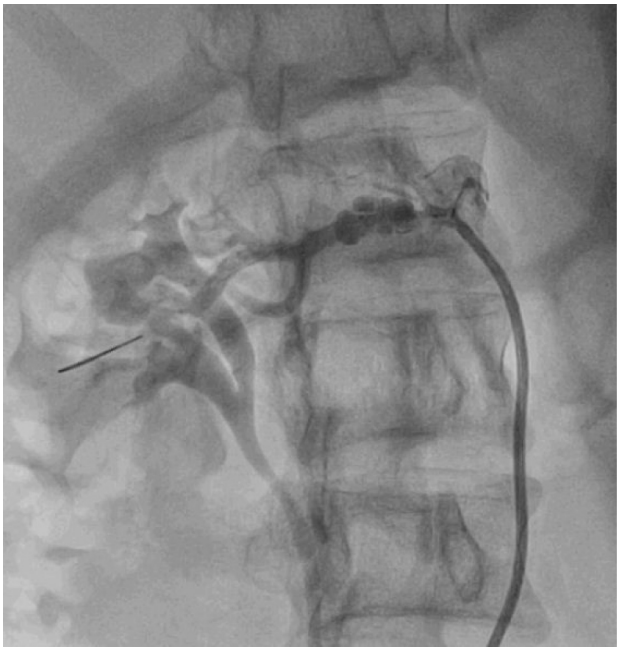

Figure 3 Angiogram post ballooning.

3. Discussion

Fibromuscular dysplasia is an uncommon cause of arterial disease that predominantly affects females.[9] It is a non-inflammatory, non-atherosclerotic disease that mostly involves the renal and carotid arteries but can involve any arterial bed. Renal artery is involved in around 60–75% of patients.[7] Histologically, FMD is classified based on the dominant

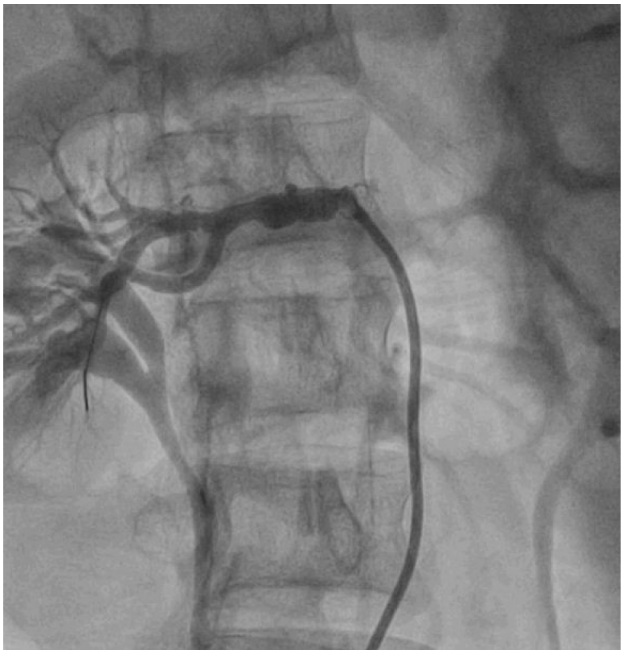

Figure 4 Angiogram post stent deployment.

arterial wall layer involved, namely intimal, medial, and adventitial (perimedial).[10] The most common is the medial type which accounts for 85% of cases and is characterized by "string of beads" appearance. The intimal and adventitial types constitute 5% and 10% respectively.[11] The etiology of FMD has not been established although a variety of genetic, mechanical, and hormonal factors have been proposed.[7]

The common age of renal involvement in FMD is between 15 and 50 years of age. Most commonly, the disease is asymptomatic and is discovered during the routine checkup or while investigating for some other disease.[7] According to the previous reports, renal FMD accounts for less than 10% of cases of RAS.[3] The commonest presentation of FMD is renovascular hypertension. The mechanism depends on whether the stenosis is unilateral (renin-mediated hypertension) or bilateral (volume depended hypertension).[12] Two-thirds of RAS due to FMD is bilateral.[13]

Once FMD as a cause of renal artery stenosis is suspected, and there are various methods of imaging for confirmation of the diagnosis. Biochemical tests for the diagnosis of RAS by using renin lack specificity. Renal duplex sonography for the detection of hemodynamically significant renovascular disease has a sensitivity of only approximately 50%. The major limitation of the duplex sonography is operator dependency.[14] The advantage is that it is noninvasive and has no apparent side effects.[15] Another advantage is that the measurement of resistive index in the cortical blood vessel can be done. Resistive index less than 80 showed favorable outcome in a study done by Radermacher et al.[16] Other methods are magnetic resonance angiogram (MRA) and computed tomographic angiography (CTA) with the use of high-resolution multi-slice detector devices.[17] The CT imaging has its limitations; the quality of image depends upon equipment, technique, and reconstruction of the images, patient-related factors, including the presence of calcium, the presence of stents, and the ability to hold one's breath during imaging.[7] MRA is limited by its frequent association of gadolinium contrast with nephrogenic systemic fibrosis.[18] Despite the invasive nature of investigation the gold standard in diagnosing FMD is intra-arterial angiogram but only as intent to revascularize because of its invasive nature. The characteristic feature is multifocal stenosis with the "string of beads" appearance, which likely indicates the presence of medial type FMD. Other patterns include tubular or focal lesions.[19]

Treatment includes medical management of hypertension and revascularization, both surgical angioplasty and percutaneous angioplasty. Medical management is done in accordance with the guidelines of the Joint National Committee on prevention, detection, evaluation and treatment of high blood pressure.[20] Almost all the patients require at least one antihypertensive and the initial drug class of choice in FMD is an angiotensin-converting enzyme (ACE) inhibitor or angiotensin receptor blocker (ARB).[21–23] Revascularization is the treatment of choice in patients in young patients with hypertension refractory to pharmacological therapy, those who are intolerant to antihypertensive and in those who have lost renal volume due to ischemic nephropathy.[7] There are two modalities of revascularization - surgical angioplasty and percutaneous transluminal angioplasty (PTA). There is a lack of data regarding direct comparison of these two modalities. Surgery used to be method of choice prior to PTA.[24,25] Nowadays, PTA has established itself as a revascularization method of choice with success rates of 60–80%.[24,25] The major advantages are that it is less costly, less invasive, can be performed as an outpatient procedure and has a lower morbidity.[7] In FMD, unlike the atherosclerosis, balloon angioplasty is the preferred therapy.[7] Routine use of stents in FMD is not recommended; however, if the dilatation is suboptimal and if renal artery dissection occurs, stenting can be performed.[26,27] Complications of PTA are around 14% and rarely, renal artery perforation, dissection or segmental renal infarctions may occur.[28,29] In 30–50% of cases, complete resolution of hypertension is achieved.[23,24]

Conflict of interest

The authors declare that they have no conflict of interest.

Ethical approval

All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed consent

Informed consent was obtained from individual in the study.

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