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CASE REPORT

Renal fibromuscular dysplasia with malignant hypertension cured by balloon angioplasty with stenting

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Abstract

We presented a 31-year-old female patient with a history of hypertension and severe pulsing headache for about 3 months. The patient had pulsing headaches over the past 3 months with increased very high blood pressure (BP = 220/130 mmHg), sometimes with blurred vision, nausea and vomiting, with no known pathological conditions in her medical history or family background. A digital subtraction angiography confirmed tight stenosis (90%) in the middle segment of the right renal artery. Balloon angioplasty with a stent was the treatment of choice. Blood pressure dropped to normal after treatment.

Key Words: *Balloon angioplasty, dysplasia, hypertension, renal artery stenosis, stent*

Introduction

Renovascular hypertension is the most common cause of secondary hypertension with a 4% prevalence rate in the general hypertensive population. Renal artery stenosis (RAS) is defined as the narrowing of one or both renal arteries, or of their branches. RAS is related to fibromuscular dysplasia (FMD), a non-atherosclerotic, non-inflammatory vascular disease of unknown etiology that causes abnormal growth within the wall of an artery (1). FMD has been found in nearly every arterial bed in the body. However, the most common arteries affected are the renal and carotid arteries (2). Atherosclerosis and FMD differ in terms of presentation and clinical consequences, as well as treatment: balloon angioplasty with stenting proved efficient and to provide positive results in some FMD patients, whereas the best management for atherosclerosis lesions is still controversial.

Case report

We report the case of 31-year-old female patient with a history of hypertension and intermittent pulsing headache for about 3 months prior to her admittance. The patient presented severe, intermittent pulsing headaches over the past 3 months with increased very high blood pressure (BP = 220/130 mmHg), sometimes with blurred vision, nausea and vomiting without obvious related causes. With no other cardiovascular risk factors, the patient was admitted to our department for further evaluation of hypertension, and the patient did not take any medication during the 3 months.

The initial clinical exam showed a normal development female patient with normal cardiovascular, respiratory and central nervous system examinations, and no detectable heart or vascular (including abdominal) murmurs. We recorded a BP of 212/140 mmHg, heart rate of 82 beats/min, pulsating peripheral arteries and no neurological issues. The chest X-ray and electrocardiogram were within normal values and revealed no additional information. The renal function test and liver function test were within the normal range.

Biologically, the patient had normal blood tests, and did not have inflammatory syndrome. Urine analysis revealed no signs of proteins, red cells or cellular elements. We performed an abdominal and

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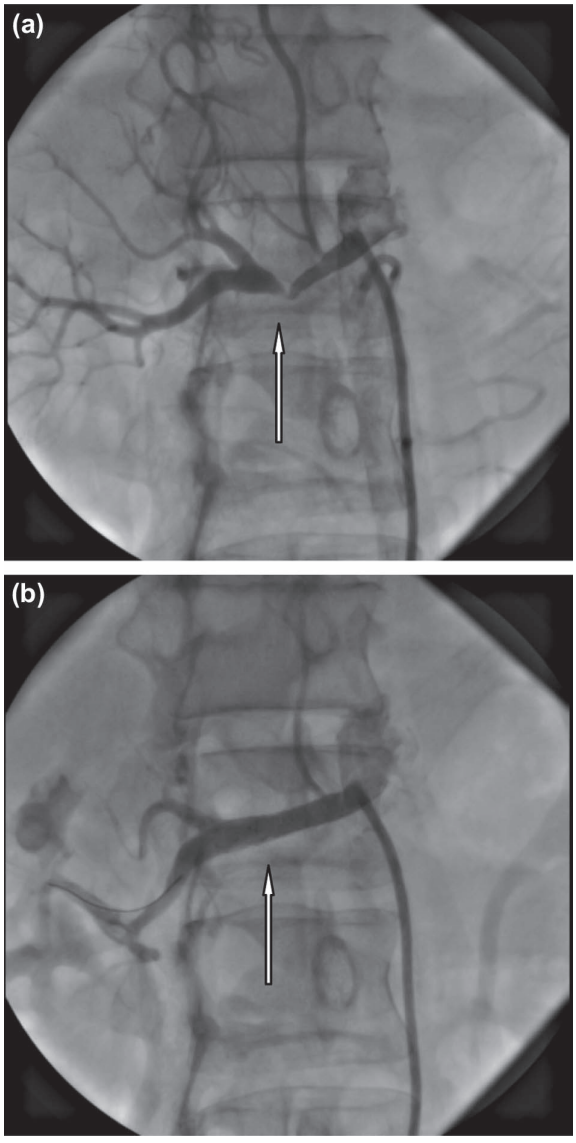

Figure 1. (a) Renal angiogram showing severe stenosis (90%) in the middle segment of the right renal artery (arrow). (b) Subsequent to percutaneous balloon angioplasty with stenting of the right renal artery: with no residual stenosis.

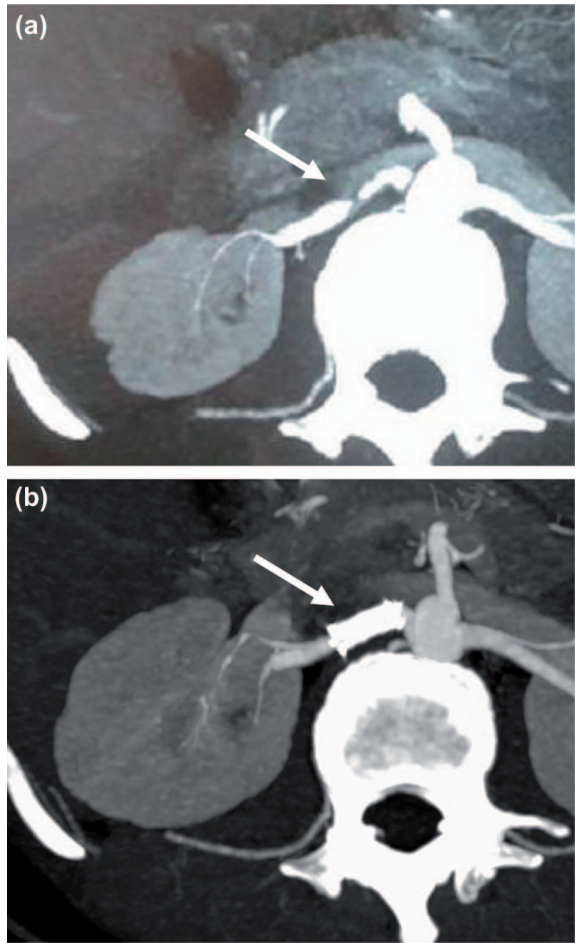

Figure 2. (a) Abdominal angio-CT scan showing the presence of severe stenosis (90%) in the middle segment of the right renal artery (arrow). (b) Abdominal angio-CT scan showing subsequent to percutaneous balloon angioplasty with stenting of the right renal artery: with no residual stenosis.

pelvic ultrasound that did not show any renal or adrenal masses, and revealed no major size difference between the two kidneys. The echocardiography investigation showed no abnormalities and no hemodynamically significant valvulopathy.

The definitive diagnosis was made by percutaneous selective renal angiography and abdominal angio-CT scan, which showed severe focal stenosis (90%) in the middle segment of the right renal artery (Figures 1a and 2a). The left renal artery, the aorta and the other abdominal arteries had a normal configuration. Percutaneous transluminal renal balloon angioplasty was partially successful and included a stent placement (RaraMount) (Figures 1b and 2b). The patient's blood pressure dropped to normal after the procedure and maintained in normal range (120/80 mmHg) with no antihypertensive medications for more than 1 year.

Discussion

FMD is an idiopathic, segmental, non-inflammatory, non-atherosclerotic disorder, which affects arterial wall resulting in arterial stenosis (3). FMD was first described by Leadbetter & Burkland in 1938 (4). McCormack and coworkers first accurately described the pathological characteristics of this entity (5). It is estimated that about 4/1000 in the general population suffer from FMD, which most commonly affects the renal arteries (60–75%) and can lead to hypertension and progressive renal atrophy (6).

FMD mostly affects women below the age of 40 and more specifically, renal arteries in the distal two-thirds, as seen in the present case. Unfortunately, the pathogenesis of this disease remains unknown despite various hypotheses linking it to genetic, mechanic or hormonal factors. Some experts mentioned there were three major types of FMD: medial dysplasia, intimal fibroplasia and adventitial (periarterial) fibroplasia (3). The most frequent type is the medial multifocal dysplasia, characterized by the "string of beads" appearance (6).

Clinical presentation depends on the arterial segment involved, length of stenosis and type of FMD. Clinical presentation can range from being asymptomatic to a necrotizing vasculitis. The most common manifestation of renal arterial FMD is renovascular hypertension. The cause remains unknown although genetic, mechanical and hormonal factors have been proposed in the pathogenesis of FMD.

Several imaging methods are used for evaluation including conventional contrast angiography (CCA), Doppler ultrasonography, renal scintigraphy, computed tomography (CTA) and magnetic resonance angiography (MRA). The catheter-based CCA is the gold standard for detecting RAS, although it is not a first-line screening tool.

The primary goal of treatment of renal artery FMD is control of hypertension. In some cases, the blood pressure can be medically managed. Further treatment options beyond medical therapy involve renal artery revascularization, which can be accomplished surgically or percutaneously. de Fraissinette reported that of 68 patients who underwent percutaneous renal artery angioplasty, 88% had improved blood pressure with 14% cured (7).

Follow-up after revascularization is typically accomplished with duplex ultrasonography. The patient's blood pressure stayed in the normal range (around 120/80 mmHg) with no antihypertensive medications and with no symptoms during the 1-year follow-up, and ultrasonography showed no abnormalities.

Conflict of interest statement: None.

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