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# Fibromuscular Dysplasia – A Rare Cause of Reno-Vascular Hypertension

**Manjunath F V[1], Taruni Ng[2], Pranab Kanti Datta[1], Sanjiv Kumar Sharma[1]**

[1]Post Graduate Trainee, Dept of Medicine, Regional Institute of Medical Sciences, Imphal – 795004, Manipur, India

[2]Professor, Dept of Medicine, Regional Institute of Medical Sciences, Imphal – 795004, Manipur, India

**Abstract:** ***Renal artery stenosis (RAS) is associated with increased cardiovascular mortality and morbidity and may constitute a treatable cause of secondary hypertension. Fibromuscular dysplasia is frequently affecting children as the main cause of RAS, but is very rare in adults. We present the case of a 20-year-old female, with no known pathological conditions in her medical history or family background, admitted for severe headache during the past 3 days and sustained increased blood pressure (BP) (maximum BP 220/120 mmHg). The initial clinical exam and first-line imagistic methods did not provide a high suspicion for RAS. However, the invasive methods established the diagnosis of right renal artery medial dysplasia.***

**Keywords:** Renal Artery Stenosis, Dysplasia, Hypertension, Cardiovascular

## 1. Introduction

Reno-vascular hypertension is the most common curable cause of secondary hypertension with a 4% prevalence rate in the general hypertensive population. The renal artery stenosis (RAS), defined as the narrowing of one or both renal arteries, or of their branches, is frequently caused by atherosclerosis (75% of all cases). More seldom, RAS is related to fibro-muscular dysplasia (FMD), while the remaining etiologies occur very rarely in medical practice. Atherosclerosis and FMD differ in terms of presentation, clinical consequences as well as treatment: the balloon angioplasty proved to be efficient and to provide positive results in FMD patients, whereas the best management for atherosclerosis lesions is still controversial.

## 2. Case Report

We present the case of a 20-year female patient, with no known pathological conditions in her medical history or family background, presented with severe headache during the past 3 days, and sustained increased blood pressure(BP at first checkup was 220/120 mmHg).

The initial clinical exam showed normal cardiovascular, respiratory, and central nervous system examinations and no detectable heart or vascular (including abdominal) bruits. The chest X-ray and the electrocardiogram, urine analysis, USG whole abdomen and ANA revealed no additional information, ranging within normal values.

We tried to identify the other neurologic causes eventually accounting for the severe headaches. To this end, we conducted a cerebral computer tomography (CT), with normal results and no suggestive modifications.

Under these circumstances, we started to suspect a renovascular etiology. Consequently, renal artery Doppler and CT renal angiography was performed, which diagnosed a right renal artery stenosis. In CT renal angiography there was evidence of tubular stenosis >60% involving 1.6cm long mid portion of right renal artery without significant post stenotic dilatation. Moreover, the right kidney was slightly smaller than the left one (right was 9.3x3.8cm and left was 12.2x5.3cm) (Fig. 1).

PRA(Plasma renin activity) was also higher(2.6ng/ml/hr) which suggests of Reno vascular cause of hypertension.

Based on the above-mentioned facts and as BP was not controlling with antihypertensives, balloon angioplastyfor the stenotic lesion was suggested, and patient opted to undergo procedure at higher centre(PGI Chandigarh).

## 3. Discussion

Fibromuscular dysplasia (FMD) is one of the two main causes of RAS, accounting for less than 10% of these cases [1]. It refers to a group of rare, idiopathic, non-atherosclerotic, non-inflammatory conditions, which lead to the narrowing of the small and medium size arteries. FMD mostly affects women below the age of 40 and more specifically, renal arteries in the distal two thirds or even segmental segments; bilateral occurrence is quite frequent (60% of cases). Despite various hypotheses linking it to genetic, mechanic or hormonal factors that are being suggested, the pathogenesis of this disease remains unknown.

There is no generally accepted classification. The histological classification includes 3 types of fibro-muscular dysplasia. The most frequent type is the medial multifocal dysplasia, characterized by the “string of beads” appearance (elastic tissue causing multiple stenosis, separated by aneurisms) [2,3]. Radiologists describe 3 aspects which can be observed: multifocal (string of beads) specific to the medial fibro-dysplasia, focal (less than 1 cm long) and tubular (longer than 1 cm).

A recent and sudden onset of severe arterial hypertension in a young female patient with negative pathological personal and family history may raise the possibility of renal artery stenosis. Likewise, refractory hypertension to aggressive antihypertensive treatment is most likely to indicate RAS,

and this pathology needs to be investigated (class I, evidence level B) [4]. In the present case report, subsequent to the RAS diagnosis, the fibro-muscular dysplasia was suggested by female gender and age.

As far as our case was concerned, we considered the patient's age as well as the negative elements for other causes of RAS the absence of plaque, atherosclerotic risk factors, inflammatory syndrome or thickening of the arterial walls and the lack of family history of the disease/syndromes.

Several imagistic methods are useful in diagnosing fibro-muscular dysplasia. When there is suspicion of RAS, duplex ultrasonography (DUS) of the renal arteries should be performed as first-line imaging test [4]. However, in the event of a positive result, the diagnosis shall also be confirmed by other imagistic methods.

The current guidelines for a day-to-day medical practice recommend that the FMD diagnosis should be based on CTA or MRA (class I, evidence level B). Also, it may be determined due to a digital subtraction angiography (the gold-standard), when there is high clinical suspicion and the results of the non-invasive tests are inconsistent (class I, evidence level C) [4].

FMD treatment must follow several goals: renal parenchyma protection while preserving renal function, BP control and the prevention of cardiovascular events. It is imperative to manage aggressively the additional risk factors by lowering lipid levels, smoking cessation and glucose levels [5].

Medical treatment is first indicated for the hypertensive patient. The medical treatment involves angiotensin-converting enzyme inhibitors (ACEI), calcium channel blockers for unilateral lesions, concomitantly aiming to control the BP and to prevent the progression of kidney dysfunction (class I, evidence level A) [4]. ACEI are contraindicated in bilateral severe RAS and single functional kidney. Thiazides, hydralazine, angiotensin II receptor blockers, and b-blockers are also effective in achieving target blood pressures in individuals with RAS [6,7].

The current guideline' recommendations for angioplasty refer to treatment-resistant hypertension, drug intolerance, signs of ischemic nephropathy (kidney function alteration and kidney size' changes) or possible curable hypertension after revascularization.

The prospect of a long-term maximal anti-hypertensive treatment for a young woman with secondary renal impairment was a strong argument for revascularization, especially when considering the eventual BP control and curable hypertension.

FMD responds well to balloon angioplasty, with positive long-term outcomes and low risk of restenosis. If distinctively necessary, the current guidelines recommend stent implantation in FMD patients (class I, level of evidence B). Balloon angioplasty was highlighted by numerous published data as providing significant reduction in BP values up to normal immediately after the procedure as well as during the long-term follow-up [8,9].

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