



Case Report

Fibromuscular dysplasia: Rare case of secondary hypertension in young male



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ABSTRACT

Introduction: and importance: Renovascular hypertension accounts for 1–2% of all cases of hypertension in the general population, but plays a major role in treatable causes of hypertension in the young. Most of the renovascular diseases are due to atherosclerosis, and fibromuscular disease is thought to be a rare cause of renovascular hypertension.

Case presentation: We present a rare case of a young male patient who was diagnosed with renovascular hypertension due to fibromuscular dysplasia of both renal arteries. Computed tomographic angiogram revealed thinning and narrowing area of both renal arteries. The left kidney looks atrophied. After vigorous antihypertensive medication and hemodialysis, he was discharged with normal blood pressure without the need for angioplasty.

Clinical discussion: Fibromuscular dysplasia (FMD) is a nonatherosclerotic, noninflammatory arterial disease that most commonly involves the renal and carotid arteries. Renal arteries are most commonly involved. Renal FMD may lead to renal failure and secondary hypertension. Multiple stenoses and the 'string-of-beads' appearance seen in renal angiogram are diagnostic for FMD. Females are mostly affected by FMD. Our case was a successfully managed young male patient with renal failure and resistant hypertension due to renal FMD.

Conclusions: Fibromuscular dysplasia causing renal artery stenosis, though a rare cause of renovascular hypertension, is essential to be considered in young hypertensives, even in the absence of a family history of hypertension. A high index of suspicion is necessary for early diagnosis and prompt treatment, which can result in rapid and complete recovery.

1. Introduction

Fibromuscular dysplasia (FMD) is an infrequent systemic vascular disease that mostly affects young women and accounts for 10%–20% of all renal artery stenosis cases worldwide. FMD is a non-atherosclerotic, idiopathic disorder that most typically affects the renal and carotid arteries, but it can affect any arterial segment [1]. Renovascular hypertension as a result of renal artery involvement is the most prevalent clinical manifestation of FMD. Renal artery stenosis caused by FMD can occur at any stage of hypertension; however it is more frequently diagnosed in patients with stage 2–3 hypertension [2,3].

Because of the rarity of renal fibromuscular dysplasia, our aim is to present here an interesting case of a young male patient with

uncontrolled hypertension diagnosed with FMD.

1.1. Case presentation

A 29-year-old male, a known case of renal failure and on hemodialysis, presented with generalized weakness and abdominal pain. He had hypertension for one year. Despite taking Amlodipine 10mg qd and Doxazosin 4mg qd, his blood pressure was not controlled.

He was on regular routine hemodialysis. His GFR was calculated using CKD-EPI and it was 8 ml/min/1.73 m². There was no known family history of chronic disease, including hypertension, diabetes, chronic kidney disease, or fibromuscular dysplasia. On admission, his blood pressure was 188/120 mmHg. Other physical exams were

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unremarkable.

The patient was admitted to the internal medicine ward. He had several sessions of hemodialysis with ultrafiltration. Blood pressure was controlled with furosemide (IV INFUSION OF 20MG LASIX 5 amp, bid) and aldactone 100mg one times per day, in addition to his prior medications (amlodipine 10mg and doxazosin 4mg).

Echocardiography showed circumscribed pericardial effusion with normal left ventricular function without any impact on the overall and segmented heart function and no hemodynamically significant valvulopathy.

His initial renal Doppler assessment was inconclusive. A contrast enhanced computerized tomography (CT Angiography) scan demonstrated thinning and narrowing area of both renal arteries in the arterial phase with 'string of beads' appearance (see Fig. 1). Diethylene triamine penta acetic acid was the contrast agent used for this CT angiogram.

The classic 'string of beads' appearance on renal angiography was suggestive of fibromuscular dysplasia. This patient's intractable hypertension was assumed to be secondary to fibromuscular dysplasia.

After giving multiple sessions of hemodialysis and 4 classes of antihypertensive medications, the patient's blood pressure returned to normal and was discharged with frusemide 20mg bid, Telmisartan 80mgqd, Aldactone100 qd, and doxazosin 4mg bid. This case has been reported in line with the SCARE 2020 criteria [4].

1.2. Clinical discussion

Fibromuscular dysplasia (FMD) is one of the two main causes of renal artery stenosis, accounting for about 10–20% of these cases. It refers to a group of rare, idiopathic, non-atherosclerotic, non-inflammatory conditions that lead to the narrowing of the small and medium-sized arteries. FMD mostly affects women below the age of 40 and, more specifically, renal arteries in the distal two thirds or even segmental segments [5,6]. Despite the fact that fibromuscular dysplasia is usually seen in females, our patient was a 29-year-old male with uncontrolled hypertension and renal failure on routine hemodialysis. Because of his severe hypertension, he developed renal failure, which made him dependent on renal replacement therapy. He had no family history of FMD. On the search for his resistant hypertension led to the finding of an underlying FMD.

FMD is most often diagnosed in the renal arteries, accounting for 60–75% of cases. Involvement of extra-cranial cerebrovascular arteries accounts for 25–30% of cases, and miscellaneous other arteries (mesenteric or brachial arteries) for up to 30% [7].

Renal artery FMD has been tentatively associated with environmental factors, and it is likely that there is a genetic predisposition. Renal manifestations present as hypertension and acute renal infarct, followed by cerebrovascular manifestations including cervical artery

dissection, stroke, subarachnoid hemorrhage, and TIA [8]. Our patient had chronic uncontrolled hypertension despite multiple antihypertensive medications. He also had accompanying renal failure and was dependent on routine hemodialysis. There was no extra renal involvement of FMD in this case, including carotid stenosis/dissection or cerebrovascular events.

Computed tomographic angiography (CTA) is the preferred diagnostic method for suspected FMD. The gold standard for diagnosis is catheter-based angiography and identification of the pathognomonic "string of beads" sign. However, given its cost, invasiveness, and risks associated with contrast and radiation dye, it is generally reserved for when other imaging studies are inconclusive or if a procedure is required [9,10]. Given the rarity of this disease, in which no previous case has been seen in our hospital, we face a great challenge in dealing with this disease. In our case diagnosis was made through contrast enhanced computerized tomography (CT Angiography) scan which demonstrated thinning and narrowing area of both renal arteries in arterial phase with 'string of beads' appearance. The classic 'string of beads' appearance on renal angiography was suggestive of fibromuscular dysplasia.

No definitive treatment for FMD. The goal of treatment is to reduce the blood pressure and to prevent organ further organ injuries. Resistance hypertension is often seen in internal medicine department and should be suspected in every patient with new onset high blood pressure and below 40 years of age [11]. The patient was managed conservatively managed with multiple antihypertensive drugs (4 drugs combinations) along with routine hemodialysis. After improvement he was discharged. On follow up he was in a better condition after ideal blood pressure control.

2. Conclusion

Though FMD is a histopathologic diagnosis, the clinical diagnosis can be made with a high degree of accuracy on the basis of the angiographic appearance. Our case was the first case of FMD in Somalia and was diagnosed based on CT angiography findings, and the patient's blood pressure was controlled with 4 classes of antihypertensive medications without the need for angioplasty.

Ethical approval

Ethical approval is not required in our institution. However, written informed consent was obtained from the patient for publication of this case report and the accompanying image. Conflict of interest statement: The authors declare that they have no competing interests.



Fig. 1. Axial and Coronal CT angiography with Arterial imaging shows stenoses and dilations, string of beads appearance of the entire right renal artery.

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Author contribution

MAN involved in patient care, collected data, and performed a literature review. MSH performed literature, wrote the manuscript and also contributed to the patient care. MOS involved in patient care. AAS involved research design. AAA involved in literature review. All authors reviewed and approved the final version for submission.

Trial register number

N/A.

Guarantor

Mosab Ahmed Nor, Corresponding Author of the manuscript.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Availability of data and materials

The datasets used and/or analyzed during the current study are available from the corresponding author on reasonable request.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Declaration of competing interest

No conflict of interest was declared by the authors.

Abbreviation

FMD	Fibromascular dysplasia
CKD	Chronic kidney disease
CTA	Computed tomography angiography
GFR	Glomerular filtration rate

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