Radiology Case Reports 19 (2024) 5785–5787

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Case Report

Fibromuscular dysplasia: An underrated cause of chronic kidney disease in developing countries: A rare case report ☆

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ARTICLE INFO

Article history:
Received 3 August 2024
Revised 13 August 2024
Accepted 15 August 2024

Keywords:
Fibromuscular dysplasia
Bilateral renal artery stenosis
Chronic kidney disease

ABSTRACT

Renal fibromuscular dysplasia (FMD) is one of the rare cardiovascular conditions affecting the kidneys at very young ages. The exact pathophysiology is still not known and is one of the causes of resistant hypertension in young patients. Severe forms of FMD such as those involving bilateral renal arteries are very few reported in the literature. In this case, we report a severe form of FMD resulting in the rapid progression of chronic kidney diseases in a young patient which results in requirement of renal replacement therapy.

Introduction

Fibromuscular dysplasia is a rare form of systemic vascular disease that affects the young population, the majority being women, and accounts for more than 10% of all renal artery stenosis [1]. The prevalence is estimated to be 12 per 100,000 individuals as per a review done in 2021 by Rana et al. [1]. The most affected artery is the right renal artery with approximately (60%-75%) of all cases, although this can happen elsewhere involving major vessels such as the carotid and vertebrae arteries [2]. Hypertension secondary to FMD is the most common clinical presentation in young patients, particularly females. Herein we present a case of FMD which resulted in rapid progression to chronic kidney diseases in a young female of African descent with involvement of both the right and left renal arteries.

Case presentation

A 29-year-old female patient of African descent presented to the outpatient clinic with a complaint of nausea accompanied by loss of appetite for the past 1 week. These symptoms were preceded by on-and-off headaches and easy body fatigue a month before presentation. She has a positive history of hypertensive heart disease in first-degree relatives of the family including her parents. She does not report any history of other chronic illnesses but reports being kept on anti-hypertensives

☆ Competing Interests: The authors declare no conflicts of interests.

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https://doi.org/10.1016/j.radcr.2024.08.079

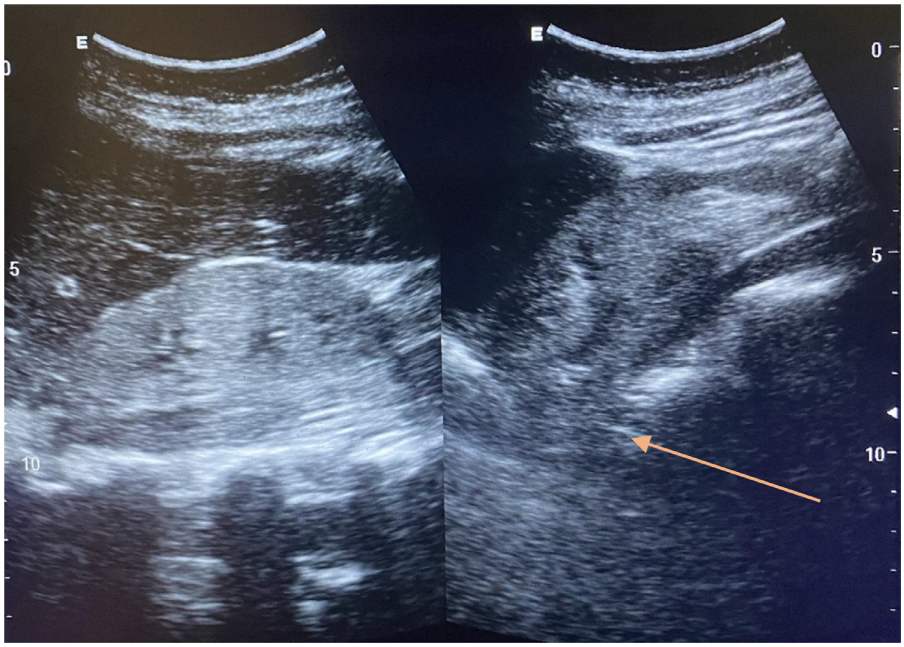

Fig. 1 – Ultrasonography of both kidneys showing poorly differentiated cortex and medulla (orange arrow).

a year ago but had poor adherence to drugs. On examination pale, with mild lower limb swelling, on auscultation decreased air entry in the right infra-mammary region other examination findings were unremarkable. Her blood pressure was 198/110 mmhg, heart rate 88 bpm. Laboratory investigations showed creatinine of 2000 micromols/l (60-120 micromols/l), urea of 45 micromols/l (2.5-6.5 mmol/l), hemoglobin count (Hb) 6.9 g/dL(12-16 g/dL), autoimmune profile including antinuclear antibody, cyclic citrullinated peptide, and serum metanephrine were unremarkable. Results showed severe anemia with poorly differentiated corticomedullary differentiation Fig. 1. Contrasted abdominal-pelvis Computed tomography was ordered to assess for any structural abnormalities of the kidneys and surrounding vessels, which showed chronic kidney disease with bilateral renal artery beading Fig. 2. Fig. 3 shows a cross-sectional view of the right and left affected renal arteries. The patient was started on hemodialysis as was not fit for other modalities of treatments due to the kidney loss.

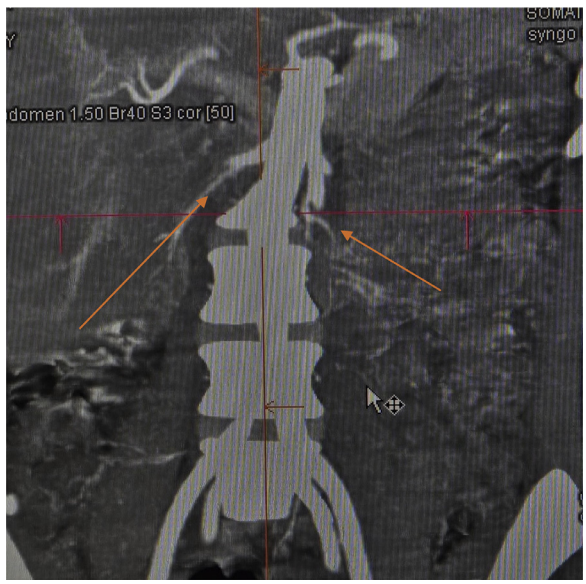

Fig. 2 – Shows evidence of bilateral renal artery beading (orange arrow).

Discussion

Fibromuscular dysplasia involving the renal artery presents with symptoms of hypertension, which will include elevated blood pressure, headache, dizziness, and occasionally tinnitus [3]. These patients are known to have a long-standing history of elevated blood pressure readings which are extremely high and very alarming [2,4]. Currently, literature has shown the physiology of FMD leading to chronic hypertension, but still sporadic cases or none have been reported to the best of our knowledge which have complicated to chronic kidney disease in the second decade of life.

The diagnosis of FMD is complicated and involves the clinician to have a high index of suspicion together with history of illness and the clinical presentation of the patient [5]. Currently, imaging remains one of the most reliable investigation which uses CT- angiography to study renal blood flow [6]. Other superior investigations like magnetic resonance imaging are used, but due to availability and expensiveness contrasted computed angiography remains the gold standard for the diagnosis of FMD of the kidney. Contrasted CT-angiography will show a sign called “string of beads” which is diagnostic for this condition, such as seen by our patient.

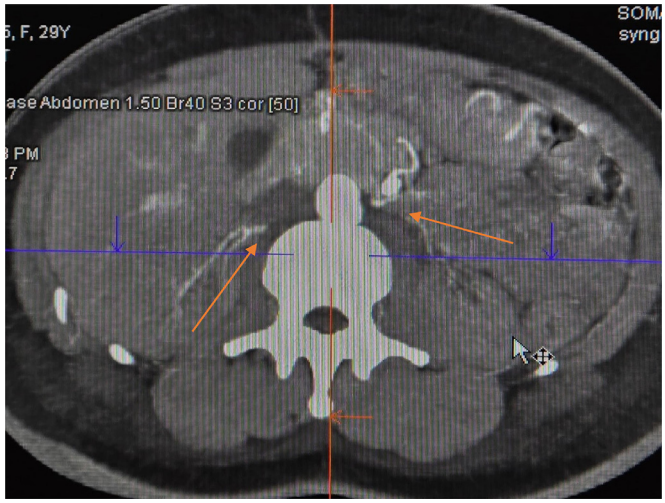

Fig. 3 – Cross-sectional view showing bilateral renal artery stenosis (orange arrow).

The treatment is aimed at the following goals, control patients' blood pressure, preservation of renal function, and prevention of complications resulting from this [2]. Three modalities of treatment have been used to date which include medical management, percutaneous intervention (angioplasty and stenting), and surgical revascularization [2,5,7]. Medical therapy with the use of anti-hypertensives like ACEI inhibitors has been seen as the primary modality of treatment before other advanced modalities [2]. Although patient compliance with the use of these antihypertensives regularly has been seen to be poor [2]. Revascularization is indicated in patients with hemodynamically significant renal artery stenosis, such as bilateral stenosis and resistant hypertension on medical treatment [8]. Conventional balloon angioplasty is the therapy indicated for renal artery stenosis caused by FMD with a cure or improved hypertension in 60%-90% of cases [8,9]. Despite all these, literature shows that the nature of progression of the disease has minimal effects on renal function hence pharmacotherapy is the initial treatment of choice for patients with FMD [9]. Our patient had resistant hypertension even though being on medications that were poorly adhered to, she rapidly developed a severe form of bilateral FMD which resulted in chronic kidney disease requiring renal replacement therapy.

Conclusion

In resource-limited settings, there is lack of diagnostic tools for these challenging medical conditions. We have seen a case of severe FMD at a young age leading to a significant morbidity to the patient and affecting her quality of life at a younger age. This condition is still rare and its exact pathophysiology is not known, clinicians in resource-limited settings should always screen for hypertension in patients at young ages as still possess the risk of having such cardiovascular emergencies. Early diagnosis is life-saving and will prevent social-economic effects on the patients.

Patient consent

The authors have obtained written informed consent for publication from the patient.

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