VOLUME - 13, ISSUE - 03, MARCH - 2024 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Medicine

FIBROMUSCULAR DYSPLASIA: RARE CAUSE OF RENAL ARTERY STENOSIS. A CASE REPORT

Karpovich Y.I	Grodno State Medical University, Grodno, Belarus
Karpovich Y.L	Grodno State Medical University, Grodno, Belarus
Patel Krushikumar Panishkumar	Grodno State Medical University, Grodno, Belarus
A.J.A. Kavindya Kavindi Fernando	Grodno State Medical University, Grodno, Belarus
D. M. P. I Dissanayake	Grodno State Medical University, Grodno, Belarus
Nimavat Anmol Manish	Grodno State Medical University, Grodno, Belarus
Oza Yugam Rajeshkumar*	Intern Doctor, M.P Shah Medical College, Jamnagar, Gujarat, India *Corresponding Author

ABSTRACT This case report details the clinical presentation, diagnostic journey, and management of a middle-aged male, aged 40, who sought medical attention in October 2023 for elevated blood pressure and left lumbar pain. Radiological investigation, including non-contrast and contrast-enhanced computed tomography (CT), unveiled left renal artery stenosis attributed to fibromuscular dysplasia. Subsequent evaluations by urologists and vascular surgeons delineated sub-occluded arterial branches and ischemic changes in the left renal parenchyma. The patient was discharged with directives for regular home monitoring, adherence to a salt-water regimen, and recurring ultrasonography of abdominal aorta and renal arteries every 6 months. Consultations with urologists and neurologists were strongly recommended, underscoring the importance of a multidisciplinary approach and diligent monitoring in the continuum of healthcare management. This case emphasizes the significance of timely diagnosis and tailored intervention in managing fibromuscular dysplasia-associated hypertension.

KEYWORDS : renal artery stenosis, fibromuscular dysplasia, high blood pressure, case report.

INTRODUCTION

Renal artery stenosis (RAS), a clinical entity marked by the narrowing of blood vessels supplying the kidneys, poses a significant challenge to renal hemodynamic and function.[1] Fibromuscular Dysplasia (FMD), the focal point of this case report, is characterized by a non-inflammatory and non-atherosclerotic pathology, contributing to vascular abnormalities.[2] This report intriguingly unfolds in the context of a 40-year-old male, deviating from the conventional demographic profile associated with FMD, which traditionally manifests in young to middle-aged women and is infrequently associated with pronounced declines in renal function.[3,4]

While FMD predominantly affects the renal and cerebral arteries, accounting for approximately 65% to 70% of cases,[2,5] it is essential to underscore that the spectrum of FMD is not confined solely to these vascular territories, exhibiting potential manifestations in diverse arterial beds.[5] The clinical categorization of FMD spans three distinctive classes: intimal, medial, and perimedial, with angiographic classification encompassing multifocal, tubular, focal, and mixed types.[3]

The consequence of RAS induced by FMD is notable for the triad of heightened blood pressure, proteinuria, and a decrement in renal function.[1] While atherosclerosis is a prevailing etiological factor in RAS, FMD assumes significance as a distinctive contributor to this vascular pathology[1]. The diagnostic landscape for renal artery stenosis attributed to fibromuscular dysplasia is intricate, demanding precision for the judicious selection of suitable treatment modalities. While selective renal angiography (SRA) remains the gold standard, alternative non-invasive diagnostic methodologies, such as Doppler ultrasound (DU), MR angiography (MRA), and CT angiography (CTA), have proven to be accurate in assessing RAS, offering valuable alternatives to traditional diagnostic angiography.[6]

Therapeutically, FMD management pivots on sustaining optimal blood pressure levels and mitigating risk factors, including obesity and tobacco smoking, with the overarching goal of preventing ischemic events.[7]

Revascularization emerges as the preeminent surgical intervention, strategically poised to forestall ischemic complications associated with FMD.[7] This case report enriches the current literature by providing nuanced insights into the exceptional presentation of FMD in a male patient, thereby accentuating the necessity for sophisticated diagnostic approaches and tailored therapeutic strategies for this infrequent manifestation of fibromuscular dysplasia.

Table 1 : Timeline of patient

16 October, 2023	NCCT at central district hospital
23 October,2023	Confirmed diagnosis by Multi Spiral CT
30 October,2023	Urologist was consulted
7 December,2023	Admitted to regional hospital and electrocardiogram, oncological examination performed
8 December, 2023	Consultation with doctors and specialist doctors, general urinalysis, blood test, biochemical blood test performed
11 December, 2023	Discharged

Case Report

In October 2023, a middle-aged male aged 40 presented himself for medical consultation due to elevated blood pressure and experiencing sharp pain in the left lumbar region. The patient had a documented history of licarnidipine 5mg intake, and average blood pressure of 120/80.

The patient denied any pertinent medical history, including tuberculosis or viral hepatitis, and confirmed no recent travel history or contact with infectious individuals. A meticulous inquiry into his medical background revealed an absence of allergies, hereditary conditions, transfusions, and a lack of symptoms indicative of intestinal infection within the preceding 10 days.

Radiological investigation via non-contrast computed tomography (CT) unveiled a hypodense area in the lower middle third of the left renal parenchyma. Despite maintaining a satisfactory objective status, the patient exhibited an elevated blood pressure reading of 140/90 mm Hg during physical examination.

Notably, peritoneal symptoms were absent, and abdominal palpation indicated softness. Furthermore, the patient demonstrated normal, painless urine output with sufficient diuresis, and the absence of edema, coupled with a negligible risk of venous thromboembolism.

A subsequent contrast-enhanced CT scan conclusively identified left renal artery stenosis, attributing it to fibromuscular dysplasia. Consequently, the patient referred to a urologist and a vascular surgeon for an exhaustive evaluation, where the sub-occluded arterial branch to the lower pole of the left kidney and ischemic changes in the left renal parenchyma were meticulously delineated.

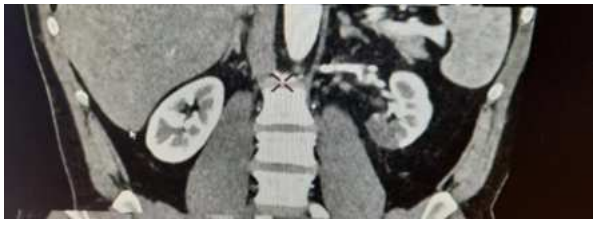

Fig.1 "Left Renal Artery stenosis from FMD causes lower lobe ischemia of the left kidney"

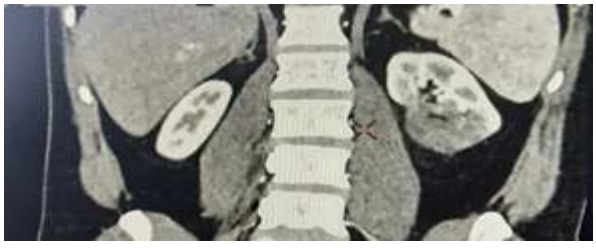

Fig.2 "Left Renal Artery stenosis from FMD causes lower lobe ischemia of the left kidney"

Fig.3 "Right kidney and intact renal artery"

In December 2023, the patient was admitted to the hospital to initiate a comprehensive investigation regimen. Oncological examination and computer electrocardiogram (ECG) were conducted on the day of admission, yielding unremarkable results of normal sinus rhythm, a horizontal electrical axis, and a heart rate of 63 beats/min. Laboratorial exams showed,

Table 2: Full blood count 08.12.2023

WBC (Leukocytes)	6.98 x109/L	(4-9 x109 /L)
RBC (Erythrocytes)	4.85 x 1012/L	(3.9 – 5.1 x 1012/L)
HGB (Hemoglobin)	152 g/l	(130-170)g/L
HCT (Hematocrit)	44.8 %	(35-50)%
MCV (Mean Red Blood Cell Volume)	92.4 Fl.	(82-92)Fl
MCH (Hemoglobin content in erythrocytes)	31.3 pg	(28-32)pg
MCHC (Erythrocyte hemoglobin concentration)	339 g/L	(320-360)g/L
PLT (Platelets)	279 x109/L	(150-450)x109/L
RDW-CV	11.9 %	(11.5-14)%
Neutrophils	66.5 %	(45-70)%
LYMPH (Lymphocytes)	22.5 %	(18-40)%
MONO (Monocytes)	7.9%	(3 - 8)%
EO (Eosinophils)	2.3%	(1 - 5)%
BASO (Basophils)	0.4%	(0 – 1)%
IG	0.4 %	(1-5)%
ESR	5	(2 - 10)

Table 3 General Urinalysis 08.12.2023

Specific gravity	1.030	(1.015-1.025)
Leukocytes	1/hpf (Microscopy)	(<9)number/hpf (Microscopy)
Nitrites (NIT)	Negative	
pH	6	(5.5 - 7)
Color	Yellow	
Transparency	Transparent	
Crystals	1/hpf (Microscopy)	(<=0) number/hpf (Microscopy)
Mucosal cells	5/hpf(Microscopy)	(<9) number/hpf (Microscopy)

Table 4: Biochemical blood test 12.08.2023

Total protein	67 g/l	(65 - 85) g/l
Albumin	42 g/l	(35-53)g/l
Urea	4.7 mmol/L	(1.7-8.3)mmol/L
Creatinine	96 μmol/L	(62-124)μmol/L
Uric acid	0.38 mmol/L	(0.2-0.42)mmol/L
Cholesterol	4.2 mmol/L	(3.12-5.2)mmol/L
Transferrin	2.04 g/l	(2-3.6) g/l
C-reactive protein	0.9 mg/L	(0-6)mg/L
Total bilirubin	18.5 μmol/L	(5-20.5)μmol/L
Blood glucose	4.2 mmol/L	(3.5-6.2)mmol/L
Aspartate aminotransferase	24 U/L	(5-37)U/L
Alanine aminotransferase	30 U/L	(5-42)U/L
Potassium	4.2 mmol/L	(3.2-5.6)mmol/L
Iron	29.3 μmol/L	(11.6-31.3)μmol/L

Subsequent laboratory investigations, including a general blood test, disclosed a marginal increase in mean erythrocyte volume (MCV=92.4 Fl) and a slightly reduced Ig level (0.4%). In contrast, general urine analysis indicated an elevated specific gravity (1030) with crystals.

Biochemical blood tests returned within the normal range, paving the way for a tailored pharmacological intervention. The prescribed regimen encompassed Losartan 50 mg once daily in the morning and Aspirin 75 mg once daily at noon.

Moreover, the patient received directives for regular monitoring of lipid profiles and blood pressure at home, adherence to a salt-water regimen, and a recurring schedule

for ultrasonography of the abdominal aorta and renal arteries every 6 months. Consultations with both a urologist and a neurologist were strongly recommended.Patient condition was satisfactory on discharge.

DISCUSSION

This case report describes the unique presentation of fibromuscular dysplasia (FMD) in a middle-aged male patient. The patient experienced elevated blood pressure and left lumbar pain, which were caused by renal artery stenosis (RAS). RAS occurs when the blood vessels supplying the kidneys narrow, leading to challenges in renal hemo dynamics and function. FMD is a rare condition that contributes to this vascular pathology.

The presented case highlights the importance of early diagnosis and treatment of FMD-associated hypertension. The findings underscore the need for sophisticated diagnostic approaches and tailored therapeutic strategies for this rare manifestation of fibromuscular dysplasia.

As per medical recommendations, revascularization is not typically recommended for patients with unilateral RAS and normal renal function, as in the case of our patient who had a serum creatinine level of 96 μmol/L (GFR 92 ml/min//1,73м^2). However, the decision to pursue revascularization should be made based on the patient's individual characteristics, medical history, and other relevant clinical factors. Healthcare providers should closely monitor the patient's renal function and adjust the dosage of ARB to minimize the risk of complications in patients with renal ischemia, who are at risk of acute renal failure.[10]

The activation of the Renin-Angiotensin-Aldosterone System (RAAS) is often associated with decreased kidney perfusion and the subsequent elevation of various hormones, leading to the development of long-term complications such as chronic kidney disease (CKD), cardiovascular disease (CVD), hypertension, cardiovascular mortality, and heart failure. Proper regulation of the RAAS is crucial in reducing the risk of such complications and improving patient outcomes.[11]

CONCLUSION:

This case report underscores the atypical presentation of Fibromuscular Dysplasia (FMD) in a middle-aged male, manifesting as controlled hypertension and left lumbar pain. The diagnostic journey, encompassing non-contrast and contrast-enhanced computed tomography, revealed left renal artery stenosis attributed to FMD. The patient managed comprehensively with Losartan and Aspirin, and demonstrated a favourable response, highlighting the efficacy of this therapeutic approach in mitigating hypertension and reducing cardiovascular risk.

The importance of a multidisciplinary approach and diligent monitoring, as emphasized by consultations with urologists and neurologists, remains paramount in managing this infrequent manifestation of FMD. This case report adds valuable insights to the literature, emphasizing the significance of timely diagnosis and tailored intervention in addressing Fibromuscular Dysplasia-induced renal artery stenosis and its associated complications.

Acknowledgement :

The authors of this case report would like to express their gratitude to the patient who consented to share their medical history for the benefit of others. We would also like to acknowledge the medical professionals who worked tirelessly to provide comprehensive care to the patients and contributed to the management of their condition.

Finally, we would like to thank the medical community for their continued efforts to advance our understanding of rare and complex medical conditions like fibromuscular dysplasia and provide better care for patients.

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