

Fibromuscular Dysplasia Presenting as Posterior Reversible Encephalopathy Syndrome in a Young Female

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ABSTRACT

Fibromuscular dysplasia (FMD) can present with renovascular hypertension with or without hypertensive emergency in mostly young patients. A 16-year-old girl presented with uncontrolled hypertension and features of posterior reversible encephalopathy syndrome (PRES). She had no features of large vessel vasculitis. Renal artery bruit could be auscultated on the right side. Blood examination showed hyperreninemic hyperaldosteronism. The USG renal artery doppler study and invasive renal angiogram showed unilateral right renal artery stenosis. She was provisionally diagnosed as a case of FMD and was treated accordingly.

Keywords: Case report, Fibromuscular dysplasia, Hypertensive emergency, Posterior reversible encephalopathy syndrome, Renal artery stenosis, Renovascular hypertension.

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INTRODUCTION

Fibromuscular dysplasia (FMD) can present with renovascular hypertension with or without hypertensive emergency in mostly young patients.¹ Exact epidemiology of FMD in India is not available. We report a case of FMD presented with posterior reversible encephalopathy syndrome (PRES).

CASE DESCRIPTION

A 16-year-old girl with no significant medical or surgical history was admitted with complaints of blurring of vision, non-localizing headache, and two episodes of seizures. On admission, her blood pressure was 210/110 mm Hg. She was unmarried and had no history of regular medication intake, any history of hematuria or claudication. On examination, all her peripheral pulses were palpable, no radio-radial or radio-femoral delay was present, and there was no significant discrepancy between systolic blood pressures of all the four limbs. A bruit could be auscultated 2 cm superior to the umbilicus and slightly right to the midline. No carotid or femoral bruit or any murmur over left scapular angle was audible. Cardiovascular examination showed no significant abnormality.

Her MRI of brain showed multifocal T2 and FLAIR hyperintensities in both cerebral hemispheres with posterior and subcortical predominance, suggestive of PRES. Her routine blood reports, serum calcium, and thyroid profile were apparently normal, except serum potassium level 1.9 mEq/L. The ABG showed metabolic alkalosis. Morning serum cortisol, 24-hour urinary metanephrines levels were normal. Blood for HIV, ANA, ESR, CRP reports were normal. Plasma renin activity was 46 ng/mL/hour (normal 0.6–4.3 ng/mL/hour), plasma aldosterone level was 540 pmol/L (cut off 450 pmol/L), and plasma aldosterone-renin ratio (ARR) was 11.73 (normal cut-off 750 pmol/L:ng/mL/hour). Urine analysis showed no hematuria. Her renal ultrasound showed size of left kidney being 10.7 × 4.9 cm and size of right kidney being 8.4 × 2.3 cm with bilateral normal cortical echogenicity. The USG doppler of B/L renal artery showed right renal artery PSV 240 cm/s, AT 72 ms, AI 2.82 m/s², RI 0.58, PI 0.5. Left kidney parameters were normal. Contrast

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CT scan of abdomen showed small right kidney with grossly stenosed right renal artery. Left kidney size and left renal artery diameter was normal (Figs 1 and 2). Invasive renal angiogram showed left renal artery was normal in diameter and dye filling phase was normal (Supplementary Video 1). Right renal artery showed long smooth narrowing and the stenosis was most prominent in the proximal part, dye filling was obstructed resulting in extensive regurgitation (Supplementary Video 2). No specific beads-on-string pattern was noticed.

The patient was diagnosed as a case of PRES with hypertensive emergency due to unilateral renal artery stenosis (RAS) [most likely etiology of FMD (intimal or medial fibroplasia)]. She was treated with oral Amlodipine and IV infusion labetalol and her symptoms subsided within 24 hours as the BP came down to normal range. As soon as the diagnosis was made, she was put on Losartan. Repeat serum potassium level was 4.1 mEq/L. Repeat MRI brain showed resolution of all previous vasogenic edema. The patient was referred to higher center for percutaneous transluminal renal angioplasty with stenting.

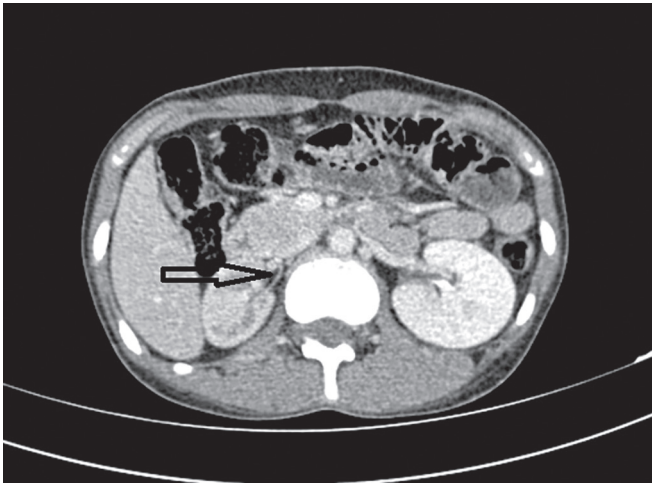


Fig. 1: Contrast CT of abdomen shows small right kidney with thin right renal artery

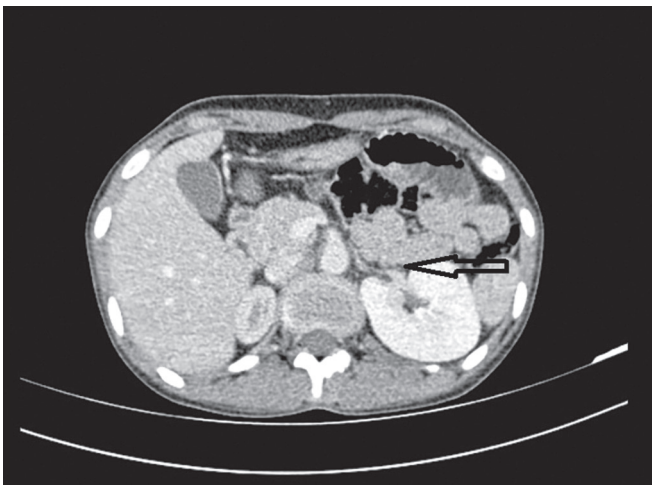


Fig. 2: Contrast CT of abdomen shows normal left kidney and left renal artery

DISCUSSION

Hypertension presents as the most common clinical manifestation of FMD of renal artery.² It is most commonly found in young females. The usual age-group is between 20 and 60 years. In 60–75% of cases of FMD, renal artery involvement can occur. This is followed by internal carotid arteries and less often vertebral, mesenteric, and brachial arteries. Coronary artery involvement is rather rarer than others. Around one-third patients have stenosis in multiple large arteries with clinical signs and symptoms.³ The commonest cause of renal artery stenosis in Indian adolescents is Takayasu arteritis, in USA it is FMD (rare in India). The FMD includes 10% of all cases of renal artery stenosis worldwide.⁴ The prevalence of clinically significant renovascular FMD is estimated at 4 in 1000 in the world.⁵ According to the histologic classification of FMD, the medial involvement is the commonest variety, which presents

with a typical “string of beads” appearance in renal angiography. The intimal type consists of circumferential deposition of collagen intima with fragmentation or doubled internal elastic lamina, and shows concentric smooth stenosis with long smooth narrowing in renal angiography, involving mainly the proximal part of renal artery. The adventitial type is the rarest one with angiogram picture of stenosis with a smooth lumen or diffuse attenuation of vessel adventitia.⁵

CONCLUSION

Patients with RAS present with uncontrolled hypertension with features of secondary hyperaldosteronism with hypokalemia. Digital subtraction angiography (DSA) is the investigation of choice due its noninvasive property, but invasive renal angiogram has more sensitivity and specificity. Even though the classical angiographic sign is “string of beads appearance,” angiographic appearance of medial and intimal fibrotic hyperplasia, and adventitial fibroplasias show smooth tubular stenosis. The optimal management includes medical control of hypertension and renal artery revascularization either by surgical grafting or angioplasty with stenting, considering patient’s clinical condition and available resources.

Clinical Significance

The FMD is the commonest worldwide cause of renal artery stenosis in young, but it is very rare in Indian subcontinent. As the disease primarily presents with hypertensive emergencies and renal failure, early diagnosis and management can prevent undesired morbidity and premature renal jeopardy.

SUPPLEMENTARY MATERIAL

The supplementary videos 1 to 2 are available online on the website of <https://www.apibpj.com/journalDetails/BPJ>.

Video 1: Renal angiogram shows normal caliber of left renal artery.

Video 2: Renal angiogram shows critical stenosis (>50%) of right proximal renal artery.

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