

It could be in the kidneys: fibromuscular dysplasia and the association with headaches and mood disorders

Dhiran Verghese, Taha Alrifai, Manojna Nimmagadda, Manisha Upadhyay

Internal Medicine, AMITA Health
Saint Joseph Hospital, Chicago,
Illinois, USA

Correspondence to
Dr Taha Alrifai;
tahaalrifai@gmail.com

Accepted 10 December 2019

DESCRIPTION

A 23-year-old woman presented to the emergency department (ED) with complaints of a headache associated with a transient episode of blurry vision lasting for about 5 min on the day prior to presentation, which resolved spontaneously. The patient had a medical history significant for migraine headaches and an 8-year history of depression. Her medications included bupropion and aspirin/paracetamol/caffeine tablets that she used as needed for headache relief. She did not have any significant surgical history. She had a family history significant for hypertension (HTN) in her father.

On presentation to the ED, the patient was found to have a blood pressure (BP) of 211/125 mm Hg, a heart rate of 117 beats/min and a respiratory rate of 18 respirations/min. Physical examination was otherwise unremarkable. Laboratory work-up was significant for a serum troponin-I level of 0.04 ng/mL (reference range 0.0–0.03 ng/mL). Other laboratory reports were significant for haemoglobin 164 g/L, hypokalaemia with potassium of 2.9 mmol/L and serum sodium of 135 mmol/L. The elevation in troponin-I was concerning for the presence of a hypertensive emergency leading to myocardial ischaemia. The patient was admitted to the telemetry unit where she received intravenous labetalol for BP control and was then started on oral antihypertensives including amlodipine, lisinopril and metoprolol and subsequently achieving adequate BP control. Work-up for secondary causes of HTN was pursued and included serum aldosterone, serum renin, aldosterone/renin ratio and serum metanephrines, all of which were within normal limits. There was a suspicion for fibromuscular dysplasia and she underwent renal arterial duplex ultrasound, which was suggestive of bilateral renal

artery stenosis with a lower restrictive index on the right suggestive of spectral broadening, consistent with turbulent flow. CT angiography demonstrated 80% irregular diffuse stenosis of the distal right renal artery ([figure 1](#)). The decision was made to pursue percutaneous angioplasty the next day. Angiography yielded a beaded appearance of the main right and left renal arteries concerning for fibromuscular dysplasia (FMD), subsequently, percutaneous transluminal angioplasty was performed with a 5 mm balloon and post-angiographic images demonstrated a good angiographic result ([figure 2](#)). We proceeded to look for carotid involvement given her headaches to rule out the presence of haemodynamically significant cerebrovascular disease. Bilateral carotid ultrasound was significant for the irregular wall, peak systolic velocity of 173 cm/s and 50%–70% stenosis in the right distal internal carotid artery, and less than 50% stenosis in the left internal carotid artery.

The patient did well after the angioplasty and she was discharged home in a stable condition. At a 6month follow-up her BP was 117/74 mm Hg on amlodipine 10 mg one time daily and lisinopril 5 mg one time daily. Prior to the angioplasty, she had weekly headaches and post-angioplasty her headaches resolved without a single episode in 6 months. She was off of the bupropion and reported a stable mood over the past 6 months.

FMD most commonly affects the renal and extracranial carotid and vertebral arteries, but nearly all arterial beds may be affected, and multivessel involvement is common. Approximately 80%–90% of patients with FMD are women.¹ The US registry for FMD has a total of 1885 registered patients, of which 1681 patients have cerebrovascular imaging. 80.4% of these patients were found to have cerebrovascular FMD



Figure 1 CT angiography of the abdomen and pelvis showing a beaded appearance of the right renal artery.



Figure 2 Angiography of the right renal artery. Left: string of beads pattern consistent with multifocal fibromuscular dysplasia; right: post-angioplasty images demonstrating good angiographic result.



© BMJ Publishing Group Limited 2019. No commercial re-use. See rights and permissions. Published by BMJ.

To cite: Verghese D, Alrifai T, Nimmagadda M, *et al.* *BMJ Case Rep* 2019;**12**:e231322. doi:10.1136/bcr-2019-231322

Learning points

- ▶ Fibromuscular dysplasia (FMD) should be suspected in young patients who present with complaints of chronic migraine headaches, particularly in the setting of pulsatile tinnitus, early-onset hypertension (HTN) or cervical or abdominal bruit.
- ▶ FMD has been recognised to involve the carotid and vertebral arteries as commonly as the renal arteries. A one-time brain to pelvis imaging study (CT or MR angiography) should be performed to assess for different vascular bed involvement.
- ▶ Young patients with new-onset HTN diagnosed with FMD should be treated with angioplasty with the goal of curing HTN.
- ▶ The use of antiplatelet agents (eg, aspirin 75–325 mg) is a class IIa recommendation for patients with FMD.

lesions (1352/1681), 77.1% have extracranial carotid artery involvement (1279/1658), 16.8% have intracranial carotid artery involvement (197/1173) and 36.9% have vertebral artery involvement (495/1342). Of note, cerebrovascular FMD lesions were noted more frequently than renal artery lesions (66.1%, 1076/1628 patients) in the US registry. In the European/International registry for FMD (total 609 registered patients), renal FMD lesions were more frequently noted (91.9%, 509/554) when compared with cerebrovascular FMD lesions (58.6%, 217/370). It is noteworthy that only 60.8% (370/609) patients had Cerebrovascular arterial imaging in the European/International registry when compared with 89.2% (1681/1885) of patients in the US registry.

Possible symptoms of cerebrovascular FMD include headaches (migraine and non-migraine type), tinnitus (pulsatile/whooshing or non-pulsatile type), unilateral headache or neck pain or focal neurological findings (partial Horner's syndrome with ipsilateral ptosis or miosis) suggestive of a cervical artery dissection, dizziness/lightheadedness, amaurosis fugax,¹ depression and anxiety.²

The prevalence of anxiety and depression is nearly double in patients with FMD when compared with that of the general population,³ and more than half of the patients with FMD experience headaches, usually migraine headaches.⁴ The possibility of FMD should be suspected in young patients who present for evaluation of chronic migraine headache, particularly in the setting of pulsatile tinnitus, early-onset HTN, or cervical or abdominal bruit.⁵

It has been recently recognised that FMD involves the carotid and vertebral arteries as commonly as the renal arteries.⁶ Therefore, a one-time brain to pelvis imaging (CT angiography or MR angiography) should be performed regardless of the initial site of diagnosis to look for other vascular bed involvements.⁷

The younger the patient, and the shorter the duration of HTN, the higher the likelihood of curing or improving BP control with percutaneous angioplasty.⁸ Angioplasty may be considered as first-line therapy in young patients with recent onset of HTN with the goal of cure of the HTN.⁹

The use of an antiplatelet agent is a class IIa recommendation for patients with FMD of the carotid arteries, regardless of symptoms.¹⁰ It is reasonable to use aspirin 75–325 mg to prevent platelet adhesion to the multiple fibrous webs noted in FMD in areas of arterial dilation which could serve as a nidus for platelet deposition.

Contributors DV contributed to writing the manuscript, image selection, conception of the work, critical review of the article and final approval of the version to be published. TA contributed to writing the manuscript, image selection, critical review of the article and final approval of the version to be published. MN contributed to the conception of the work, critical review of the article and final approval of the version to be published. MU contributed to the conception of the work, critical review of the article and final approval of the version to be published.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Gornik HL, Persu A, Adlam D, *et al*. First international consensus on the diagnosis and management of fibromuscular dysplasia. *Vasc Med* 2019;24:164–89.
- 2 Harriott AM, Zimmerman E, Singhal AB, *et al*. Cerebrovascular fibromuscular dysplasia. *Neurology: Clinical Practice* 2017;7:225–36.
- 3 Bumpus SM, Heidt ST, Krallman R, *et al*. Single site lifestyle survey in FMD patients suggests improvement over time. *Circ Cardiovasc Qual Outcomes* 2017;10.
- 4 Swan K, Gu X, Kline-Rogers E, *et al*. Prevalence of headaches in patients with fibromuscular dysplasia. *Circ Cardiovasc Qual Outcomes* 2017;10.
- 5 O'Connor SC, Poria N, Gornik HL. Fibromuscular dysplasia: an update for the headache clinician. *Headache* 2015;55:748–55.
- 6 Olin JW, Froehlich J, Gu X, *et al*. The United States Registry for fibromuscular dysplasia: results in the first 447 patients. *Circulation* 2012;125:3182–90.
- 7 O'Connor SC, Gornik HL. Recent developments in the understanding and management of fibromuscular dysplasia. *J Am Heart Assoc* 2014;3:e001259.
- 8 Trinquart L, Mounier-Vehier C, Sapoval M, *et al*. Efficacy of revascularization for renal artery stenosis caused by fibromuscular dysplasia: a systematic review and meta-analysis. *Hypertension* 2010;56:525–32.
- 9 Olin JW, Gornik HL, Bacharach JM, *et al*. Fibromuscular dysplasia: state of the science and critical unanswered questions: a scientific statement from the American heart association. *Circulation* 2014;129:1048–78.
- 10 Brott TG, Halperin JL, Abbara S, *et al*. 2011 ASA/ACCF/AHA/AANN/AANS/ACR/ASNR/CNS/SAIP/SCAI/SIR/SNIS/SVM/SVS guideline on the management of patients with extracranial carotid and vertebral artery disease: executive summary. A report of the American College of Cardiology Foundation/American heart association Task force on practice guidelines, and the American stroke association, American association of neuroscience nurses, American association of neurological Surgeons, American College of radiology, American Society of Neuroradiology, Congress of neurological Surgeons, society of atherosclerosis imaging and prevention, Society for cardiovascular angiography and interventions, society of interventional radiology, society of NeuroInterventional surgery, Society for vascular medicine, and Society for vascular surgery. *Circulation* 2011;124:489–532.

Copyright 2019 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <https://www.bmj.com/company/products-services/rights-and-licensing/permissions/>
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

Customer Service

If you have any further queries about your subscription, please contact our customer services team on +44 (0) 207111 1105 or via email at support@bmj.com.

Visit casereports.bmj.com for more articles like this and to become a Fellow