

[EDITORIAL]

Fibromuscular Dysplasia: Another Paradigm Shift in Renovascular Hypertension?

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Renovascular hypertension (RVH) is one of the most common forms of secondary hypertension. It is defined as a syndrome of elevated blood pressure induced by a variety of conditions that interfere with perfusion to the kidney. This most commonly includes atherosclerotic renal artery disease (ARAD) and fibromuscular dysplasia (FMD) but also includes renal artery (RA) aneurysm, arterial embolism, arteriovenous fistula, aortic coarctation, vasculitis (Takayasu's disease, Middle aortic syndrome) and type 1 neurofibromatosis.

The article by Sakuma et al. (1) in this issue of *Internal Medicine* describes the case of a young patient with RVH due to FMD in an extra-renal artery. They detected segmental impairment of renal perfusion and a "string-of-beads" appearance on computed tomography angiography (CTA), and the resolution of hypertension was achieved by successfully restoring vessel patency with percutaneous transluminal renal angioplasty (PTRA). This case underscores the need for screening for FMD with multi-row detector CTA in young hypertensive patients.

FMD is a group of idiopathic, non-inflammatory, non-atherosclerotic vascular diseases of medium-sized arteries leading to arterial stenosis, aneurysm, dissection and arterial tortuosity. Sakuma et al.'s case also raises three important issues related to the current concept and management of FMD. First, contrary to the traditional view that FMD predominately affects the renal arteries, vascular changes were observed in every artery. The most frequently involved arteries are the renal and internal carotid arteries, followed by the vertebral, visceral, external iliac arteries and even intracranial arteries (2). Second, FMD is more common than previously thought. Young and healthy renal donor candidates had an FMD prevalence of 2.3-3.9% detected by CTA (3). In fact, FMD is an incidental finding in the every-day practice of diagnostic imaging. Furthermore, in a prospective

registry of 469 FMD patients, in which patients with symptomatic renal arterial FMD underwent CTA or magnetic resonance angiogram (MRA) other than for the RA and those with cervical FMD underwent CTA or MRA other than for the cervical artery, 225 (48.0%) had multisite stenoses. At least 2 vascular beds were affected by stenoses, aneurysms and dissections in 66.1% of cases. Although the disease presentation of FMD may vary widely, depending on the artery involved and the severity of disease, RA imaging should be proposed for hypertensive patients with a cerebrovascular presentation. Furthermore, cervical artery imaging should be considered in patients with a renal presentation and bilateral RA lesions (4). Third, FMD most frequently occurs in women of child-bearing age. PTRA, as in this case (1), and selective renal embolization (5) are important options to consider in order to eliminate the need for renin-angiotensin system inhibitors to control blood pressure during pregnancy.

In conclusion, FMD is a systemic arterial disease that may be underdiagnosed, possibly due to traditional misconceptions. Eliminating such misunderstandings will pave the way to a better diagnosis and management of FMD.

The authors state that they have no Conflict of Interest (COI).

References

1. Sakuma I, Saito J, Matsuzawa Y, Omura M, Matsui S, Nishikawa T. A unique case of renovascular hypertension due to fibromuscular dysplasia in an extra-renal artery. *Intern Med* 57: 2689-2694, 2018.
2. Olin JW, Gornik HL, Bacharach JM, et al.; American Heart Association Council on Peripheral Vascular Disease, American Heart Association Council on Clinical Cardiology, American Heart Association Council on Cardiopulmonary, Critical Care, Perioperative and Resuscitation, American Heart Association Council on Cardio-

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- vascular Disease in the Young, American Heart Association Council on Cardiovascular Radiology and Intervention, American Heart Association Council on Epidemiology and Prevention, American Heart Association Council on Functional Genomics and Translational Biology, American Heart Association Council for High Blood Pressure Research, American Heart Association Council on the Kidney in Cardiovascular Disease, American Heart Association Stroke Council. Fibromuscular dysplasia: state of the science and critical unanswered questions: a scientific statement from the American Heart Association. *Circulation* **129**: 1048-1078, 2014.
3. Hendricks NJ, Matsumoto AH, Angle JF, et al. Is fibromuscular dysplasia underdiagnosed? A comparison of the prevalence of FMD seen in CORAL trial participants versus a single institution population of renal donor candidates. *Vasc Med* **19**: 363-367, 2014.
 4. Plouin PF, Baguet JP, Thony F, et al.; and the ARCADIA Investigators. High prevalence of multiple arterial bed lesions in patients with fibromuscular dysplasia: the ARCADIA registry. *Hypertension* **70**: 652-658, 2017.
 5. Mishima E, Suzuki T, Seiji K, et al. Selective embolization therapy for intrarenal artery stenosis causing renovascular hypertension: Efficacy and follow-up renal imaging. *J Clin Hypertens* **19**: 1028-1031, 2017.

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