



# Fibromuscular Dysplasia: A Rare Case with Multiple Vascular Beds Involvement

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## Abstract

### Keywords

- ▶ fibromuscular dysplasia
- ▶ multiple vascular beds
- ▶ intracranial aneurysm
- ▶ acute coronary syndrome

Fibromuscular dysplasia (FMD) is an idiopathic, noninflammatory, and nonatherosclerotic vascular disease of small to medium-sized arteries. It can occur in almost all arteries and most commonly involving the cervicocranial and renal arteries. FMD is commonly presents as renovascular hypertension and affects mostly young ladies. However, this case demonstrates a casuistically rare form of multiple arterial beds involvement at different sites, that is, vertebral, coronary, hepatic, and lumbar arteries, with the conjunction of both bilateral renal and cervicocranial arteries.

## Key Message

1. FMD is commonly presents as renovascular hypertension.
2. The clinical manifestations are determined primarily by the vessels that are involved. The signs and symptoms are nonspecific. Thus, it may cause a significant delay in diagnosis.
3. The reported incidence of greater than 4 vascular beds involvement is rare.

## Introduction

Fibromuscular dysplasia (FMD) is an idiopathic, segmental, nonatherosclerotic, noninflammatory vascular disease that affects small to medium arteries. In both ARCADIA (Assessment of Renal and Cervical Artery Dysplasia) and U.S. registries,<sup>1,2</sup> the incidence of FMD involvement of greater than 4 vascular beds was reported as 3.2 and 9.1%, respectively. Again, coronary involvement with other vascular beds was extremely rare. For this case, she was not screened for FMD when she first presented with acute coronary syndrome. She

was only investigated after an acute cranial nerve palsy episode, and subsequent angiograms revealed multiple intracranial aneurysms with other vascular beds involvement. From the neurosurgical aspect, there is a controversy regarding managing an unruptured cervicocranial aneurysm or dissection. In addition, FMD is a disease with multiple systems involvements, and management will be more challenging and requires a multidisciplinary team for long-term follow-up.

## Case History

A 30-year-old young lady was first diagnosed with hypertension at the age of 25. Her initial young hypertension workout was unremarkable, which included ultrasound of kidney and Doppler of renal arteries. Four years later, she presented one episode of unstable angina. Coronary angiogram (▶ **Fig. 1**) revealed two-vessel diseases with the presence of markedly diffuse irregularities and stenosis. She was discharged with antiplatelet medication and labeled as a

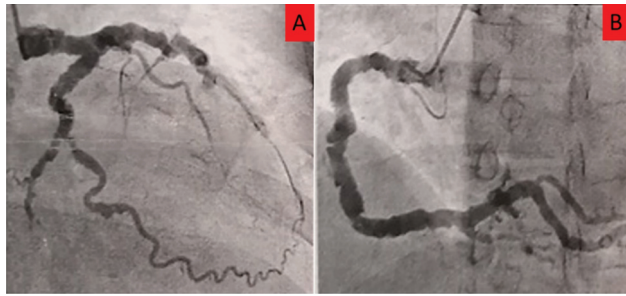
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**Fig. 1** Cardiac angiogram showed diffuse irregularities with “string of beads” appearance on left (A) and right (B) coronary arteries.

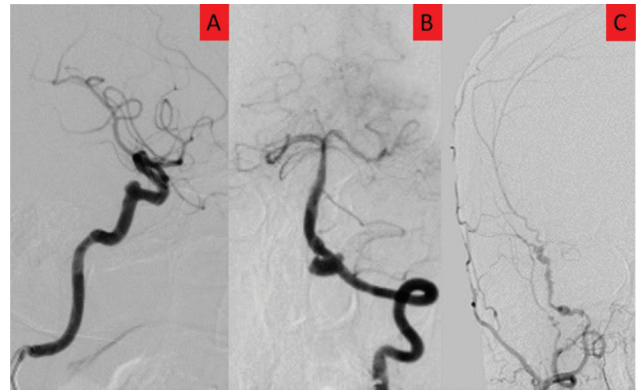
missed Kawasaki disease missed in childhood and presented as an acute coronary syndrome in adulthood.

Unfortunately, she presented with a short duration of right 6th cranial nerve palsy 2 years after the cardiac event. Magnetic resonance imaging brain revealed multifocal infarcts over bilateral corona radiata and centrum semiovale. Magnetic resonance angiography (MRA) also showed a left V4 segment of vertebral artery aneurysm. Her subsequent cerebral digital subtraction angiography (DSA) (→**Fig. 2**) revealed a saccular aneurysm at right internal carotid artery lacerum/cavernous junction and dissecting aneurysm at left V4 segment of vertebral artery. She was further investigated for connective tissue diseases. However, her blood screening result turned out to be unremarkable. Computed tomography aortogram (CTA) (→**Fig. 3**) showed beaded appearance and stenosis of vertebral, lumbar, hepatic, and renal arteries.

She was initially undecided on surgical intervention for her intracranial aneurysm. During her subsequent follow-up (6 months later), her cerebral DSA (→**Fig. 4**) showed a larger aneurysm over left V4 segment of vertebral artery. She was recounseled for endovascular intervention, given the increased risk of spontaneous rupture. She underwent flow diverter-assisted coiling of left vertebral artery aneurysm. She was discharged well without complications.

## Discussion

The clinical manifestations of FMD are determined primarily by the vessels that are involved. Many of the signs and symptoms of FMD are nonspecific, thus causing a significant delay in diagnosis with an average delay of 4 to 9 years.<sup>1,3</sup>



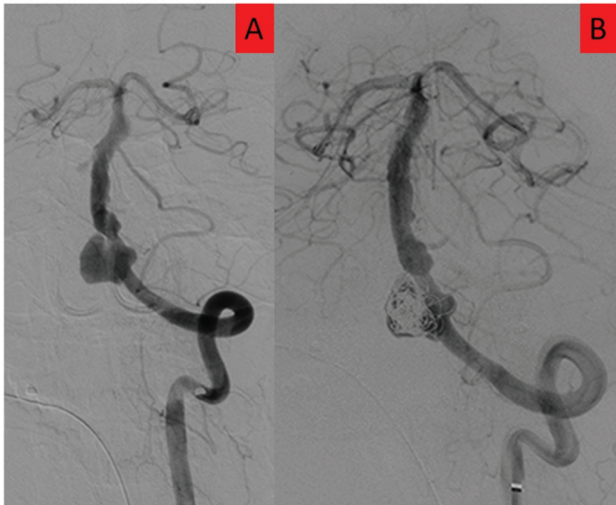
**Fig. 2** (A) Saccular dilation at the junction of right internal carotid artery (ICA) lacerum/cavernous segment, measuring 6.3 × 6.1 mm (AP × W) and 5.1 mm at the neck, and pointing posterior medially. (B) Bilobed fusiform dilatation at V4 segment of left vertebral artery with larger lobe oriented anteromedially measuring 12 × 10 mm (AP × W) and 5.9 mm at the neck, while smaller lobe measures 3.3 mm in width and 3.8 mm in the neck. (C) Right middle meningeal artery showed a short segment with a beaded appearance.

Currently, there are not many registries to show a true prevalence of this disease. The prevalence of FMD in multiple vascular beds could not be determined accurately as all vascular territories were not imaged for the same patients. Much of the previous literature reported that FMD most commonly affected the renal arteries (70%) and less frequently the carotid and vertebral arteries (25–30% involvement).<sup>1</sup>

Nowadays, diagnosing FMD is almost exclusively radiographically. It further limits the utility of histopathological classifications developed by Harrison and McCormack<sup>4</sup> in 1971. With introducing percutaneous revascularization, obtaining diagnostic histological specimens have become quite rare. Therefore, imaging has become the primary method for diagnosing FMD. The American Heart Association scientific statement<sup>3</sup> and European consensus statement<sup>5</sup> on FMD have described a classification based on angiographic appearance to replace the histopathologic classification. Noninvasive imaging studies include duplex ultrasonography, CTA, and MRA, but the gold standard remains catheter-based angiography. The pathognomonic of FMD is a string of beads’ appearance on diagnostic angiography. The classic appearance is characterized by long-segment tubular



**Fig. 3** (A) A short segment stenosis with a beaded appearance at the common hepatic artery. (B) Right kidney: Wedge-shaped hypodensity at the upper pole with renal scarring. Left kidney: Calcified plaque at left middle interlobar artery with > 50% stenosis with the beaded appearance of interlobular arteries at the mid pole. Minimal wedge-shaped hypodensity at the mid pole in keeping with renal scarring. (C) Calcified plaques with a beaded appearance (multiple short segment stenoses) of lumbar arteries at L3 level.



**Fig. 4** (A) Bilobed fusiform dilatation at V4 segment of left vertebral artery with larger size measured  $9.8 \times 12\text{mm}$  (AP  $\times$  W) while smaller lobe measured  $7 \times 5.4$  (AP  $\times$  W). (B) Post-coiling and stent insertion.

stenosis or ovoid-shaped outpouchings. Unfortunately, there are no specific guidelines for the diagnosis of FMD. Imaging all vascular beds from head to pelvis for screening may cause unnecessary radiation exposure and may not be cost effective. On the other hand, a delay or missed diagnosis may lead to poor outcomes and its sequelae.

Primary therapeutic goals of FMD include controlling risk factors, controlling blood pressure, and preventing ischemic events.<sup>6</sup> In patients with ischemic strokes, antithrombotic medications such as antiplatelet agent or anticoagulants are prescribed to prevent further vascular events. However, there is no protocol devoted explicitly to the management of FMD-related aneurysms. The treatment of intracranial aneurysms is similar to other types of aneurysms.<sup>6</sup> Therapeutic options for securing intracerebral aneurysms are microvascular neurosurgical clipping and endovascular coiling. With an uncertain and inadequate understanding of the natural history of FMD, we could not predict disease progression. In the general population, the mean annual risk of rupture of unruptured intracranial aneurysms is  $< 1\%$ . Touzé et al<sup>7</sup> summarized data from 14 studies of cervical and intracranial FMD, the incidence of subarachnoid hemorrhage was varied between 3 and 49%. To bear in mind, patients with nonruptured cerebral aneurysm pose a more challenging clinical dilemma as we do not know whether FMD is associated with an increased risk of spontaneous rupture. In this case, we offered intervention as her aneurysm was increasing in size with greater than 10 mm dimension. This was supported by a recent study by the Unruptured Cerebral Aneurysm Study Japan Investigators.<sup>8</sup> It mentioned a hazard ratio of 10 to 24 mm and posterior circulation aneurysms

were 9.09 (95% confidence interval [CI], 5.25 to 15.74) and 1.90 (95% CI, 1.12 to 3.21), respectively.

In conclusion, the management of FMD-related aneurysms is influenced by the life expectancy of the patient, the estimated risk of rupture, and the risk of complications of preventive treatment. Besides, the risk of rupture depends on many factors, including aneurysm-related factors (number, size, shape, and location of aneurysms). Nevertheless, an enlarging aneurysm is warranted for intervention to prevent a debilitating neurological consequence during a spontaneous rupture event. Also, there is a great need for further study or research on FMD in terms of natural history and outcome to determine the best treatment options.

#### Conflict of Interest

None declared.

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