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Case Report

Fibromuscular Dysplasia: A Rare Case with Multiple Vascular Beds Involvement

Abstract

Fibromuscular dysplasia (FMD) is an idiopathic, non-inflammatory, and non-atherosclerotic vascular disease of small- to medium-sized arteries. It can be occurred in almost all arteries and most commonly involving cervicocranial and renal arteries. FMD is commonly present as renovascular hypertension and affecting most young ladies. However, this case demonstrates a casuistically rare form of multiple arterial beds involvement at different sites, i.e. vertebral, coronary, hepatic, and lumbar arteries, with the conjunction of both bilateral renal and cervicocranial arteries.

Keywords: *Acute coronary syndrome, fibromuscular dysplasia, intracranial aneurysm, multiple vascular beds*

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Introduction

Fibromuscular dysplasia (FMD) is an idiopathic, segmental, non-atherosclerotic, and non-inflammatory vascular disease that affects small- to medium-sized arteries. In both ARCADIA and US registries,[1,2] the incidence of FMD involvement of >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 Report

A 30-year-old young female was first diagnosed with hypertension at the age of 25. Her initial young hypertension workout was unremarkable, which included an ultrasound of the kidney and Doppler of the renal arteries. Four years later, she presented 1st episode of unstable angina. Coronary angiogram [Figure 1] revealed 2-vessels diseases with the presence of markedly diffuse irregularities and stenosis. She was discharged with antiplatelet medication, and labeled as a missed Kawasaki disease missed in childhood and presented as an acute coronary syndrome in adulthood.

Unfortunately, she presented with a short duration of the 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 the vertebral artery aneurysm. Her subsequent cerebral digital subtraction angiography (DSA) [Figure 2] revealed a saccular aneurysm at the right internal carotid artery lacerum/cavernous junction and dissecting aneurysm at the left V4 segment of the vertebral artery. She was further investigated for connective tissue diseases. However, her blood screening result turned out to be unremarkable. Computed tomography aortogram (CTA) [Figure 3] showed beaded appearance and stenosis of the vertebral, lumbar arteries, hepatic, and renal arteries.

She was initially undecided on surgical intervention for her intracranial aneurysms. During her subsequent follow-up (6 months

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later), her cerebral DSA [Figure 4] showed a larger aneurysm over the left V4 segment of the vertebral artery. She was counseled for endovascular intervention, given the increased risk of spontaneous rupture. She underwent flow diverter-assisted coiling of the 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 4–9 years.[1,3] 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%).[1]

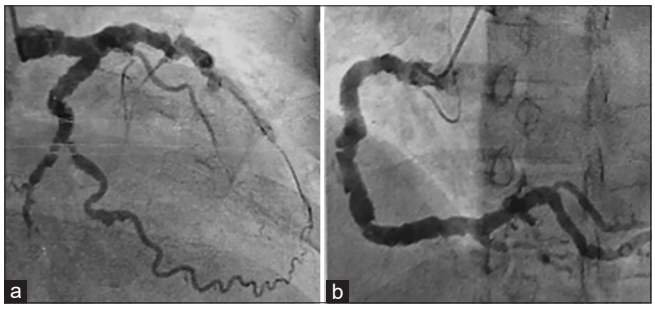

Figure 1: Cardiac angiogram showed diffuse irregularities with "string of beads" appearance on the left (a) and right (b) coronary arteries

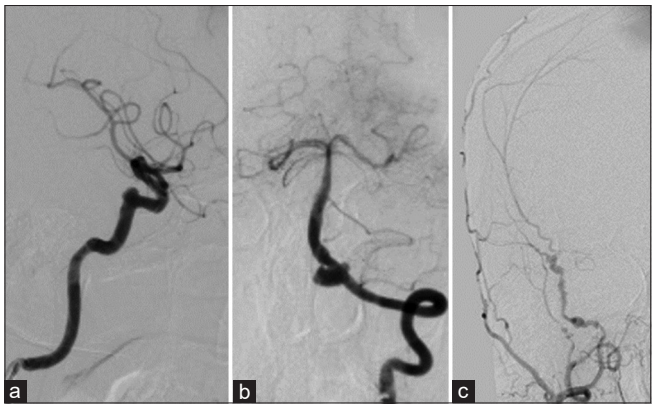

Figure 2: (a) Saccular dilation at the junction of the right internal carotid artery-lacerum/cavernous segment, measured 6.3 mm × 6.1 mm (AP(Anterio-posterior) × W(Width)) and 5.1 mm at the neck, and pointing posterior-medially, (b) bilobed fusiform dilatation at the V4 segment of the left vertebral artery with larger lobe oriented anteromedially measured 12 mm × 10 mm (AP × W) and 5.9 mm at the neck, while smaller lobe measured 3.3 mm in width and 3.8 mm at the neck, (c) the right middle meningeal artery showed a short segment with beaded appearance

Nowadays, diagnosing FMD is almost exclusively radiographically. It further limits the utility of histopathological classifications developed by Harrison and McCormack[4] 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[3] and European consensus statement[5] 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 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, blood pressure, and preventing ischemic events.[6] In patients with ischemic strokes, antithrombotic medications such as antiplatelet agents 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.[6] 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.*[7] summarized data from 14 studies of cervical and intracranial

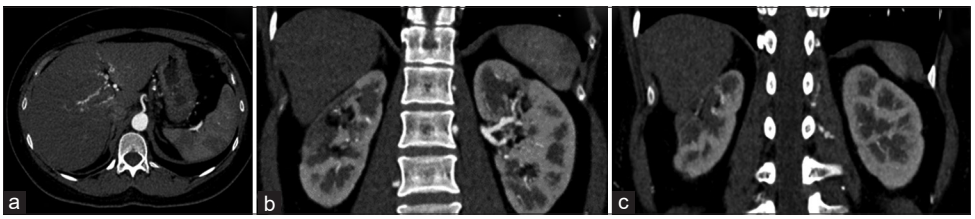

Figure 3: (a) A short segment stenosis with beaded appearance at the common hepatic artery, (b) the right kidney: Wedge-shaped hypodensity at the upper pole in keeping with renal scaring. Left kidney: Calcified plaque at the left middle interlobar artery with >50% stenosis with beaded appearance of the interlobular arteries at the midpole. Minimal wedge-shaped hypodensity at the midpole in keeping with renal scarring, (c) multiple short segment stenosis with calcified plaques and beaded appearance at the left L3 lumbar artery

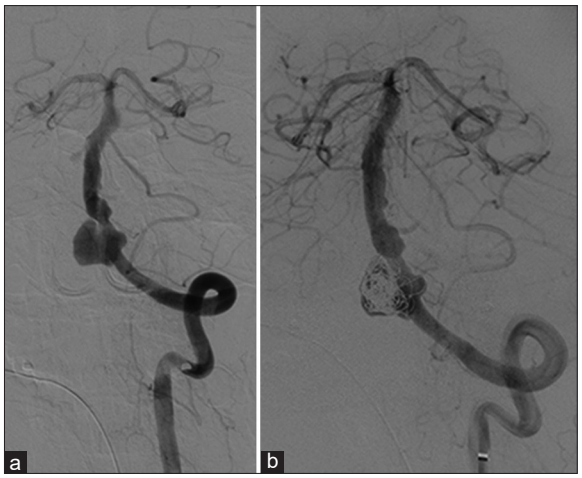

Figure 4: (a) Bilobed fusiform dilatation at the v4 segment of the left vertebral artery with larger size measured 9.8 mm × 12 mm (AP × W) while smaller lobe measured 7 × 5.4 (AP × W), (b) post-coiling and stent insertion

FMD, the incidence of subarachnoid hemorrhage was varied between 3% and 49%. To bear in mind, patients with non-ruptured cerebral aneurysms 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 >10 mm dimension. This was supported by a recent study by the Unruptured Cerebral Aneurysm Study Japan investigators.[8] It mentioned hazard ratio of 10–24 mm and posterior circulation aneurysms were 9.09 (95% confidence interval [CI], 5.25–15.74) and 1.90 (95% CI, 1.12–3.21), 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 is multifactorial, which include 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. Furthermore, 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.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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