

Acute Aortic Dissection and Stroke in Multivessel Fibromuscular Dysplasia

Subrata Kar, DO
Raja R. Gopaldas, MD
Arun Kumar, MD

Fibromuscular dysplasia is a rare, nonatherosclerotic, noninflammatory vascular disease that typically affects women between the ages of 20 and 60 years. Although any artery can be affected, fibromuscular dysplasia most commonly affects the renal and carotid arteries. Fibromuscular dysplasia of the renal arteries usually presents with hypertension, while carotid or vertebral artery disease causes transient ischemic attacks, strokes, or dissection. Fibromuscular dysplasia of the brachial arteries is extremely uncommon. It can induce extremity ischemia, nerve compression, or both—causing coldness, discoloration, pain, ulceration or gangrene of the fingers, paresthesias, or paralysis.

We report a rare case of multivessel fibromuscular dysplasia manifested by acute stroke in association with type I aortic dissection, which progressed rapidly to ascending aortic false aneurysmal development that necessitated arch replacement. Outcomes of aortic arch replacement in this setting are currently unknown. Therefore, our case might well offer some insight. (Tex Heart Inst J 2013;40(1):88-90)

Key words: Aneurysm, dissecting/complications/etiology/pathology; aortic aneurysm, abdominal; aortic aneurysm, thoracic; blood vessel prosthesis implantation; brachial artery/pathology; carotid artery diseases/pathology; differential diagnosis; fibromuscular dysplasia/diagnosis/etiology/therapy; interventional cardiology; radial artery/access; renal artery/pathology

From: Divisions of Cardiovascular Medicine (Drs. Kar and Kumar) and Cardiothoracic Surgery (Dr. Gopaldas), University of Missouri Hospital-Columbia School of Medicine, Columbia, Missouri 65212

Address for reprints: Subrata Kar, DO, Division of Cardiovascular Medicine, University of Missouri Hospital-Columbia School of Medicine, 5 Hospital Dr., Columbia, MO 65212

E-mail: skar762@aim.com

© 2013 by the Texas Heart® Institute, Houston

Fibromuscular dysplasia (FMD) is a rare, nonatherosclerotic, noninflammatory vascular disease that typically affects women (up to 4%) between the ages of 20 and 60 years.¹ Fibromuscular dysplasia can occur in any artery, most commonly affecting the renal and carotid arteries. In the renal arteries, fibromuscular dysplasia most commonly presents with hypertension.¹ In the carotid or vertebral arteries, fibromuscular dysplasia causes transient ischemic attacks, strokes, or dissection.² Fibromuscular dysplasia should be suspected if hypertension occurs in patients younger than 35 years of age or if transient ischemic attacks, stroke, aneurysm, or dissection occurs in young patients.^{1,3} We present an unusual case of a patient with FMD of the brachial, carotid, and renal arteries whose initial manifestation was an acute stroke secondary to type I aortic dissection, which progressed rapidly to an ascending aortic false aneurysm.

Case Report

In October 2010, a 58-year-old white woman with a history of hypertension, chronic kidney disease, anemia, and arthritis presented with acute left-sided hemiplegia. During diagnostic evaluation, carotid angiography revealed a right internal carotid artery occlusion, for which intravenous tissue plasminogen activator was promptly administered. In the course of this same carotid angiogram, FMD of the carotid artery was identified (Fig. 1). An attempt to obtain better imaging of the carotid architecture revealed an aortic dissection that extended into the carotid arteries from the descending aorta. The right renal artery was seen to be marginally perfused due to extension of the dissection flap, and it was also affected by FMD (Fig. 2). Thoracic aortic surgery was deferred indefinitely due to the large area of stroke involvement and the patient's questionable neurologic prognosis. Her initial hospital course was complicated by urosepsis and by respiratory failure that necessitated tracheostomy. After a tenuous hospital course, she was weaned from respiratory support and transferred to a rehabilitation center.

Four months after the initial event, the patient had progressed to a near-complete neurologic recovery and a functionally independent lifestyle. Cardiac magnetic resonance, prompted by suspicious findings of fresh thrombus on follow-up echocardiography, revealed a rapid 2.5-cm increase in aortic diameter caused by a dissecting (false) aneurysm (Fig. 3) that would need surgical intervention.

Cardiac catheterization was performed as a component of preoperative planning. Because of the extensive nature of the dissection, the radial artery was chosen as the access vessel; this also enabled better evaluation of the right axillary artery in preparation for surgical cannulation. During cardiac catheterization, right brachial artery FMD was discovered (Fig. 4), but there was no upper-extremity ischemia and we encountered no technical difficulties during catheterization. The patient subsequently underwent successful aortic arch reconstruction with the Spielvogel technique. She was dis-



Fig. 1 Carotid angiography performed on admission during diagnostic evaluation for stroke shows fibromuscular dysplasia of the carotid artery.



Fig. 2 This renal angiogram, obtained during the initial angiography, shows fibromuscular dysplasia affecting the right renal artery, complicated by a dissection flap that extends into the ostium.

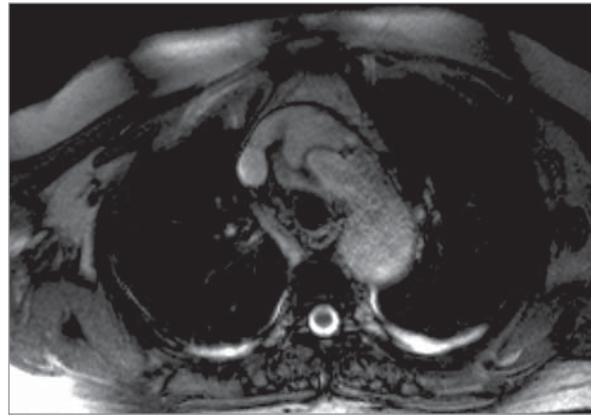


Fig. 3 Cardiac magnetic resonance, performed 4 months after the initial presentation, shows the aortic dissection progressing to a dissecting aneurysm with blood extravasation into the false lumen at the level of the aortic arch.

Real-time motion image is available at www.texasheart.org/journal.

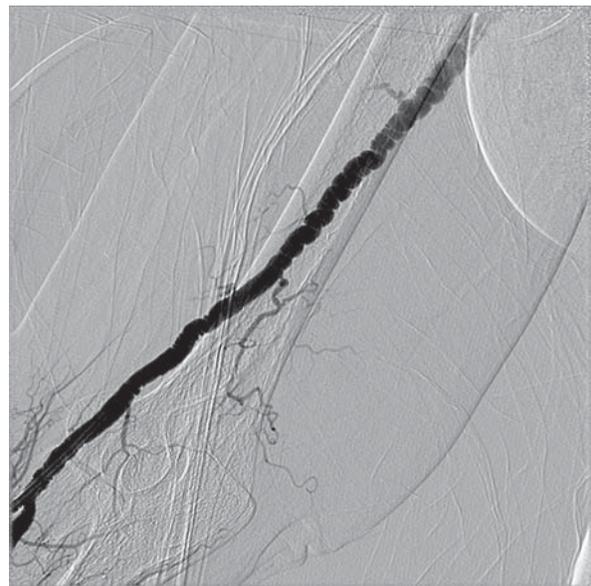


Fig. 4 Cardiac catheterization was performed via the right upper extremity, for purposes of surgical planning. Fibromuscular dysplasia affecting the brachial artery is evident from the "string-of-beads" appearance, which suggests the medial-fibroplasia type. (See Discussion.)

charged after a 2-week hospital course. Postoperative follow-up, one month and 6 months after surgery, revealed an excellent surgical outcome.

Discussion

Fibromuscular dysplasia is a rare disorder of medium- and small-diameter arteries, such as the renal and carotid arteries.⁴ In younger patients, FMD tends to be specifically localized to the mid or distal areas of the

blood vessels.³ Fibromuscular dysplasia of the renal artery occurs in 75% of FMD patients, 35% of whom have bilateral renal involvement.¹

Fibromuscular dysplasia affects the tunica intima, tunica media, and adventitia. Disease of the media is subclassified into medial fibroplasia, perimedial fibroplasia, and medial hyperplasia. Medial fibroplasia is the most common type, occurring in 70% to 95% of cases.⁴ It displays the typical “string-of-beads” appearance, caused by aneurysmal dissection in association with altered blood flow.⁵ Perimedial fibroplasia also has a string-of-beads appearance, but these lesions are infrequent. Adventitial fibroplasia is the least common type (<1%) and is associated with localized, tubular stenosis secondary to collagen replacement.³

Fibromuscular dysplasia has a familial link, with autosomal dominant inheritance and reduced penetrance in males.⁶ Suzuki and colleagues⁶ reported a case of bilateral brachial artery FMD in a mother and daughter, which suggests a possible hereditary relationship. Fibromuscular dysplasia has been shown to coexist with cystic medial necrosis in patients with Marfan syndrome.⁷ Fibromuscular dysplasia can also be caused by trauma, ischemia, or infection, by hormonal, metabolic, or immunologic factors, or by a deficiency of elastic fibers.^{4,6}

Fibromuscular dysplasia of the brachial arteries is extremely uncommon.⁴ Brachial artery FMD can induce extremity ischemia, nerve compression, or both—causing a myriad of symptoms.⁴ However, it most commonly causes distal embolization, which leads to ischemia of the digits and other extremities.⁵ The differential diagnosis should include neurofibromatosis-related vascular lesions, Takayasu arteritis, arteriosclerosis, Ehlers-Danlos syndrome, and catecholamine-induced functional stenosis, as in pheochromocytoma.⁴ Because of the rarity of brachial artery FMD, the course of the disease remains unknown, as does the optimal treatment.

For brachial artery FMD, the use of angioplasty, surgical resection, stenting (in cases of dissection), or arterial bypass might be preferred over long-term antiplatelet or anticoagulative therapy, the benefits of which are unknown.^{1,3} Cutts and colleagues⁵ reported a case of brachial artery FMD that was treated with bilateral sequential saphenous vein bypass.

The treatment of cerebrovascular FMD includes antiplatelet therapy with 81 mg of aspirin daily.³ Angioplasty is the first-line therapy in symptomatic patients, whereas stenting should be considered in patients with vascular dissections who fail anticoagulation therapy.³

Our patient displayed unusual involvement of multiple arterial beds—including the carotid, renal, and brachial—which might have contributed to the eventual type I aortic dissection. However, we were unable to produce definitive pathologic evidence. The final pathology report on the ascending aorta revealed the pres-

ence of an organizing hematoma within the sub-tunica media of the artery, which was consistent with chronic aortic dissection. Also noted, in the adjacent soft tissue, was chronic inflammation with fibrosis. There was no obvious microscopic change or evidence of cystic medial necrosis that might suggest a predisposition to aortic dissection. Nevertheless, the clinical course of events is certainly compelling. Multivessel FMD with aortic or major branch-vessel dissection has been reported, but it is extremely rare.^{8,9} Outcomes of aortic arch replacement in the setting of FMD are currently unknown. Therefore, our case might well offer some insight into this rare problem.

References

1. Olin JW, Sealove BA. Diagnosis, management, and future developments of fibromuscular dysplasia. *J Vasc Surg* 2011;53(3):826-36.e1.
2. Yoshimuta T, Akutsu K, Okajima T, Tamori Y, Kubota Y, Takeshita S. Images in cardiovascular medicine. “String of beads” appearance of bilateral brachial artery in fibromuscular dysplasia. *Circulation* 2008;117(19):2542-3.
3. Olin JW, Pierce M. Contemporary management of fibromuscular dysplasia. *Curr Opin Cardiol* 2008;23(6):527-36.
4. Kolluri R, Ansel G. Fibromuscular dysplasia of bilateral brachial arteries--a case report and literature review. *Angiology* 2004;55(6):685-9.
5. Cutts S, Grewal RS, Downing R. Bilateral brachial artery fibromuscular dysplasia. *Eur J Vasc Endovasc Surg* 2000;19(6):667-8.
6. Suzuki H, Daida H, Sakurai H, Yamaguchi H. Familial fibromuscular dysplasia of bilateral brachial arteries. *Heart* 1999;82(2):251-2.
7. Schievink WI, Bjornsson J, Piepgras DG. Coexistence of fibromuscular dysplasia and cystic medial necrosis in a patient with Marfan's syndrome and bilateral carotid artery dissections. *Stroke* 1994;25(12):2492-6.
8. Akashi H, Nata S, Kanaya K, Shintani Y, Onitsuka S, Aoyagi S. Spontaneous dissection of the iliac artery in a patient with fibromuscular dysplasia. *Ann Vasc Surg* 2010;24(7):952.e13-6.
9. Gatalica Z, Gibas Z, Martinez-Hernandez A. Dissecting aortic aneurysm as a complication of generalized fibromuscular dysplasia. *Hum Pathol* 1992;23(5):586-8.