

Endovascular management of an acute type B aortic dissection in a patient with fibromuscular dysplasia

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ABSTRACT

We report the case of a 44-year-old woman who developed an acute type B aortic dissection caused by an entry tear from an aneurysmal left common iliac artery that extended retrograde to the proximal descending thoracic aorta. She experienced refractory chest pain despite optimal medical management, thereby indicating repair. Endovascular aortic repair was subsequently performed. Intraoperatively, fibromuscular dysplasia was diagnosed by the characteristic appearance of her renal arteries. The patient tolerated the procedure and had resolution of her chest pain. In summary, we present a highly unusual case of type B aortic dissection resulting from a retrograde common iliac artery tear in a patient with fibromuscular dysplasia. (*J Vasc Surg Cases and Innovative Techniques* 2018;4:76-9.)

Fibromuscular dysplasia (FMD) is an idiopathic, noninflammatory, and nonatherosclerotic vascular disease commonly found in young women that can lead to aneurysms, stenosis, and dissection. Arterial complications of FMD often involve medium-sized arteries, particularly the carotid and renal arteries.¹⁻⁵ Some have reported FMD in other arterial beds, including the common iliac arteries (CIAs).^{1,6} Aortic involvement, however, is unusual, and aortic dissections are exceptional. A few case reports have documented a type A aortic dissection in patients with FMD.⁶⁻⁸ There has been no published case report of a type B aortic dissection (TBAD) in a patient with FMD to our knowledge. We report a case of a patient with FMD with multivessel involvement who presented with an acute TBAD resulting from an entry tear within a CIA aneurysm.

Consent was obtained from this patient to discuss her medical history. The case study was exempt from Institutional Review Board review.

CASE REPORT

The patient, a 44-year-old woman with history significant for poorly controlled hypertension, presented with acute tearing abdominal and chest pain. Computed tomography (CT)

angiography showed intramural hematoma of the descending thoracic aorta with continuing arterial dissection of the paravisceral and distal segments that terminated in bilateral CIAs (Fig 1). On closer examination, the main entry tear of the dissection was not of thoracic origin but appeared to emanate from an aneurysmal left CIA tear that extended retrograde toward the proximal descending thoracic aorta (Fig 1). Subsequent angiography showed a second entry tear in her aneurysmal right CIA (Fig 2). Despite medical optimization, the patient continued to have refractory abdominal and chest pain. Endovascular aneurysm repair (EVAR) was performed to cover the primary entry tears in the CIA aneurysms.

Procedure. Procedural angiography and intravascular ultrasound during EVAR confirmed the main entry tear of the dissection within the left CIA aneurysm (Fig 2). A bifurcated unibody device (AFX 22 mm × 60 mm × 13 mm × 40 mm; Endologix, Irvine, Calif) was selected for coverage because the primary objective of the repair was to support the distal aorta at the origin of the dissection. Extension iliac endografts were deployed for complete entry tear seal across the CIA. On the left, a Cook Spiral-Z limb (11 mm × 107 mm; Cook Medical, Bloomington, Ind) was delivered and deployed, preserving flow to the left hypogastric artery. On the right, another entry point into the aortic dissection was seen that originated close to the iliac artery bifurcation (Fig 2). A Cook Spiral-Z limb (11 mm × 107 mm) was deployed into the external iliac artery. The hypogastric artery was covered to obtain appropriate seal across the CIA. Angioplasty of the endografts was not performed at the proximal and distal seal zones. A 12 mm × 4 cm balloon was used for angioplasty of the graft-to-graft junctions only. Completion angiography revealed excellent coverage of the entry tears with no evidence of perfusion of the false lumen (Fig 3). Intravascular ultrasound reconfirmed true luminal expansion and ensured no malperfusion of the proximal perivisceral aortic branches.

During visceral aortography, a classic "string of beads" sign was noted in bilateral renal arteries, thus confirming FMD (Fig 3). Her renal FMD was not treated at the time because of the hyperacuity of her acute aortic dissection. There were no postoperative

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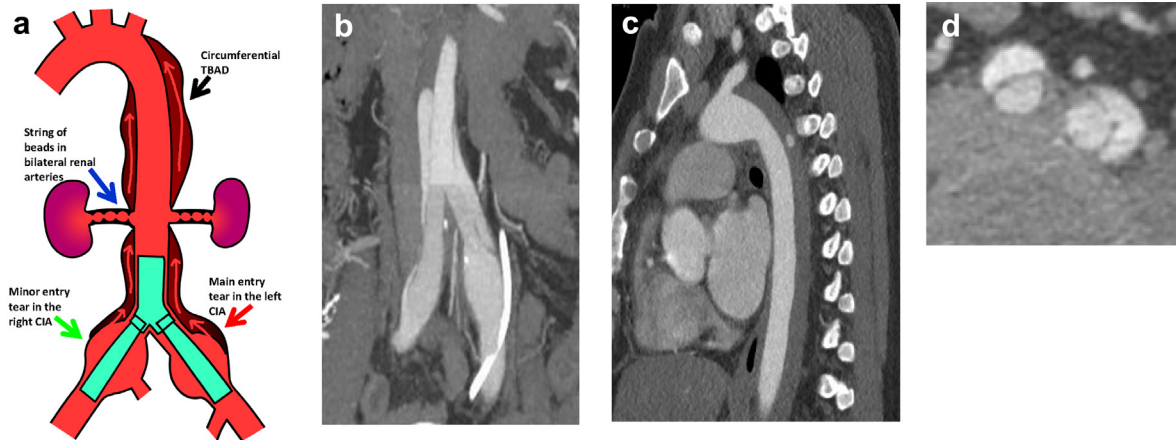


Fig 1. Overview and radiographic findings of the aortic dissection in our patient. **a**, Overview illustration demonstrating the aortic disease process in our patient with fibromuscular dysplasia (FMD). **b**, Coronal view computed tomography (CT) angiogram demonstrating extension of dissection to the distal aorta involving bilateral common iliac artery (CIA) aneurysms. **c**, Sagittal view CT angiogram demonstrating an intramural hematoma and dissection of the descending thoracic aorta with areas of false lumen perfusion. **d**, Axial view CT angiogram of the CIA aneurysms with apparent entry tear of the left CIA. TBAD, Type B aortic dissection.

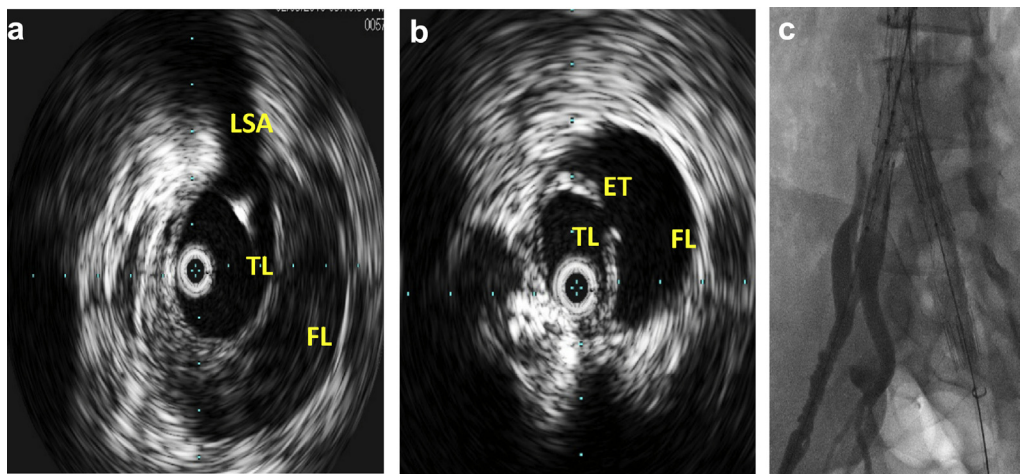


Fig 2. Endovascular aneurysm repair (EVAR) procedural images. **a**, Intravascular ultrasound image of the descending thoracic aorta at the level of the left subclavian artery (LSA). A large false lumen (FL) and comparatively smaller compressed true lumen (TL) were noted. **b**, Intravascular ultrasound image of the left common iliac artery (CIA) aneurysm and dissection. Entry tear (ET) and separation of the TL and FL are clearly seen. **c**, Intraoperative angiography during EVAR demonstrating separate entry tear of the dissection from the right CIA.

complications. As she was recovering, further evaluation was completed, and the patient was found to have bilateral internal carotid artery ectasia near the carotid siphon. She did not have intracranial artery aneurysms.

Follow-up. The patient had resolution of her abdominal and chest pain postoperatively. She was discharged but required five antihypertensive medications for blood pressure control. Subsequent CT scans evaluating her repair 1 month and 6 months postoperatively demonstrated progressive false lumen thrombosis and nonprogression of her CIA aneurysms. There was no evidence of endoleak. Angioplasty of the bilateral renal arteries

was eventually performed at 14 months postoperatively for refractory hypertension. The patient tolerated the procedure without complications, and her hypertension subsequently improved. Adequate blood pressure control was achieved with two antihypertensive medications. Her 18-month follow-up CT study showed excellent overall aortic remodeling and no EVAR device complications (Fig 3).

DISCUSSION

The clinical presentation of FMD can vary from asymptomatic to multisystem disease that simulates vasculitis.⁹ Leadbetter and Burkland² first described FMD as the

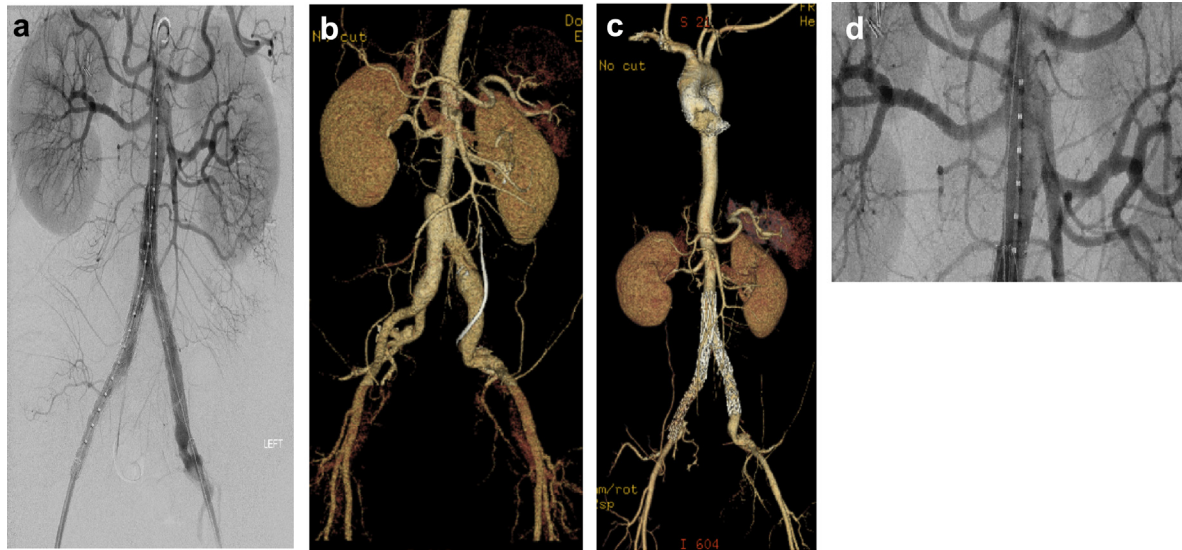


Fig 3. Postprocedural imaging and follow-up imaging. **a**, Completion angiogram after endovascular aneurysm repair (EVAR). **b**, Three-dimensional volumetric image of aorta at presentation. **c**, Remodeling of aorta after EVAR. **d**, Bilateral renal arteries demonstrating classic “string of beads” appearance signifying fibromuscular dysplasia (FMD).

pathologic culprit responsible for hypertension observed in a patient with unilateral renal disease in 1938. FMD was primarily considered a renal artery phenomenon until the 1960s, when it was observed to be responsible for extrarenal arterial dysplasias.³ Later, FMD was confirmed and implicated in stroke caused by carotid disease in young women.⁴ As more research was gathered on FMD, the dominant incidence of the disease in women was also established.¹⁰

Despite much progress, the pathogenesis and prevalence of FMD remain unclear. Multiple studies have suggested a genetic predisposition, possibly an autosomal dominant with variable penetrance inheritance pattern.^{11,12} Some have theorized hormonal influence because FMD occurs in women more frequently than in men, but there is little evidence to support the theory.¹⁰ Smoking is another possible risk factor. Although some reports have shown a dose-dependent relationship between cigarette smoking and risk of FMD and that the proportion of current smokers is higher among FMD patients, these data still need to be verified with larger studies.^{13,14}

Among extrarenal manifestations of FMD, aortic involvement is rare.¹⁶⁻⁸ This is the first occurrence of TBAD in a patient with FMD to our knowledge. The pathogenesis of TBAD in this patient is likely secondary to her uncontrolled hypertension from primary renal FMD involvement. Although tissue diagnosis is lacking, the development of bilateral CIA aneurysms in this patient is possibly a consequence of a systemic FMD phenotype. The primary entry tear in this patient occurred within her bilateral CIA aneurysms, which is also unusual. On the initial CT scan, a penetrating ulcer appeared to be

present in the descending thoracic aorta and was believed to be the entry point for this aortic syndrome. On a subsequent CT scan before her operation, however, the area of pooling of the contrast material in the hematoma could be tracked back to an intercostal artery. Her presentation was also unusual for an acute antegrade aortic process originating from the chest in that she presented with tearing abdominal pain followed by chest pain. The distribution of the intramural hematoma isolated to the thoracic aorta followed by a distal aortic dissection near the aortic bifurcation also implicates a retrograde propagation in the pathophysiologic mechanism of this aortic syndrome. Retrograde rather than antegrade progression of the dissection is perplexing but may be related to the suspected entry tear's being situated closer to the aortic bifurcation as opposed to the iliac bifurcation. Perhaps the distal aorta itself was abnormal in structural makeup and affected by FMD, allowing an easier path for the dissection to extend retrograde.

Optimal treatment of FMD remains uncertain for clinically active disease. Options range from conservative to invasive.¹⁵ For benign conditions, FMD can be surveyed with noninvasive imaging and conservative measures. Modification of the patient's lifestyle is recommended, such as cessation of smoking, reduction of cardiovascular risk factors, and medical therapy with antiplatelet, antithrombotic, and antihypertensive agents. Surgery and, in the current era, endovascular therapy may be required on the basis of disease type and extent. The natural history of FMD is poorly understood, and consideration for invasive treatment must also take

into account the disease potential, such as aneurysmal degeneration and flow-limiting stenosis. Because of the low disease prevalence, however, treatment is typically guided by single case reports or small retrospective case series.

In our patient, successful treatment outcome was achieved with endovascular means by addressing both the renal and extrarenal manifestations of FMD. Historically, endoluminal techniques have proven efficacy in renal and extracranial carotid involvement.¹³ Aortic complications have traditionally required open surgery for management.^{1,6-8} In this case, EVAR, although applied in an off-label situation, was an ideal modality for managing both the TBAD and iliac aneurysmal disease.

CONCLUSIONS

We present an exceptionally unusual case of a TBAD in a patient with FMD that was successfully treated with EVAR. Endovascular techniques play an integral role in the management of FMD complications and appear to be safe and effective. More studies are needed to better elucidate the pathophysiologic mechanism and natural history of FMD, and more research is needed on differing endovascular repair techniques for the management of disease complications.

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