


A case of type A aortic dissection with underlying fibromuscular dysplasia

Viesha Ciura^{1*}, Amy Bromley², Jason Wong¹

1. Department of Diagnostic Imaging, Foothills Medical Center, Calgary, Alberta, Canada

2. Department of Anatomic Pathology, Foothills Medical Center, Calgary, Alberta, Canada

* **Correspondence:** Viesha Ciura, MD, Department of Diagnostic Imaging, Foothills Medical Center, 1403 - 29th Street NW, Calgary, Alberta, Canada T2N 2T9

 viesha.ciura@albertahealthservices.ca

Radiology Case. 2011 Oct; 5(10):22-28 :: DOI: 10.3941/jrcr.v5i10.457

ABSTRACT

Fibromuscular dysplasia is a rare, non-atherosclerotic non-inflammatory vascular disease that most commonly involves the renal arteries and carotid arteries, but has been described in nearly every vascular bed in the body. Complications of fibromuscular dysplasia include aneurysms and vascular dissection. We present a rare case of fibromuscular dysplasia involving the aorta, complicated by type A aortic dissection.

CASE REPORT

CASE REPORT

A 64 year old male developed sudden onset of throat pain, midline thoracic and abdominal pain, as well as left arm pain, with an associated pulse deficit and coolness of the left arm. He had a complicated past medical history including known aortic root and intraabdominal aortic aneurysms, intracranial anterior communicating artery aneurysm, renal cell carcinoma requiring partial left nephrectomy, hypertension, atrial fibrillation, and anemia. A CT angiogram was performed, which demonstrated an acute type A aortic dissection extending from the aortic root down to the level of both common iliac arteries, and extending cephalad into the left subclavian, left common carotid and right brachiocephalic arteries (Figure 1 A-D). A 7 cm abdominal aortic aneurysm was also noted just above the bifurcation and involving the left common iliac artery, as was aneurismal dilatation of the proximal celiac artery, and the iliac and femoral arteries (Figure 1E-H). The renal arteries appeared normal. He was also known to have an anterior communicating artery aneurysm, seen on previous CT angiogram of the head (Figure 2). CTA of the neck showed marked tortuosity and aneurismal dilatation of the internal carotid and vertebral arteries (Figure 3).

The patient was scheduled for immediate surgery. Intraoperatively, note was made of a pre-existing large aortic root aneurysm, and contained rupture of the ascending aorta with a clear intimal tear seen. Severe aortic insufficiency and dilatation of the right coronary artery ostium were noted. The aortic valve was replaced, and the entire ascending aorta resected and replaced with a 28 mm straight tube Dacron graft.

The surgical specimen consisted of multiple fragments of aorta with macroscopically obvious blood accumulated in the outer portion of the medial layer. Patchy, yellow, atheromatous plaques were present on the intimal surface. Microscopic evaluation of the aortic wall demonstrated diffuse, marked fragmentation, atrophy, and loss of the elastic fibres with smooth muscle cells migrating into the intima (Figure 4). Areas of accumulation of acid mucopolysaccharide material were patchy in the media, but no definitive cystic medial necrosis was observed. Microscopy also confirmed the presence of fresh blood and blood clot in the vascular wall.

DISCUSSION

Fibromuscular dysplasia (FMD) is a rare, non-inflammatory non-atherosclerotic vascular disease, the etiology of which is unclear. The renal arteries, followed by the internal carotid arteries, are most commonly involved, however FMD has been described in nearly every vascular bed in the body [1]. Involvement may be isolated to a single artery or may be generalized involving multiple vascular beds concomitantly. A previous study demonstrated that in approximately one fourth of patients with FMD, two or more arteries are involved [2]. The disease typically manifests as a cause of renovascular hypertension, usually in middle aged women. Several causes have been proposed, including cigarette smoking, hormonal factors, and a history of hypertension [3]. The disease is also more common in first degree relatives of patients with renal FMD [4].

Involvement of the aorta by FMD is extremely rare, and can lead to complications such as aneurysms and aortic dissection. In this report we present a rare case of FMD of the thoracic aorta complicated by type A aortic dissection. The patient also had evidence of systemic disease elsewhere, involving the visceral branches of the aorta, the iliac arteries, the intracranial arteries and carotid and vertebral arteries.

Hypertension is the most important risk factor for patients presenting with aortic dissection. Atherosclerosis appears to have an important role in the etiology of aortic dissection, as in some cases, there appears to be a close relationship between the site of the intimal tear and atherosclerotic plaque [5]. Other underlying vascular pathology may predispose to aortic dissection, including Marfan's syndrome, and cystic medial necrosis. The most important predisposing factor is advancing age. Previous aortic surgery, including for aortic dissection, also predisposes to dissection [5]. Other diseases, such as Ehlers-Danlos syndrome, are associated with increased risk of aortic aneurysms [6], and there has also been a case of aortic dissection reported [7].

There have been less than 30 previously reported cases of FMD involving the aorta, manifesting as thoracic aortic aneurysms, abdominal aortic aneurysms, aortic hypoplasia and aortic coarctation [8-21]. Rarer yet, with only one case reported in the literature [22], is aortic dissection as a complication of fibromuscular dysplasia.

Although this patient had normal appearing renal arteries, with no CT angiographic evidence for fibromuscular dysplasia, several of the visceral branches of the aorta, the iliac vessels and the carotid and vertebral arteries showed radiologic evidence for involvement. The patient also had an aneurysm of the anterior communicating artery, which has previously been described as a complication of generalized FMD [23,24].

FMD is classified based on the arterial wall layer in which the lesion predominates: the intima, media, or adventitia [25]. The most common dysplastic lesion is medial fibroplasia, which is characterized by a homogenous collar of elastic tissue that presents as multiple stenoses interspersed with aneurismal outpouchings. Perimedial fibroplasia is second most

common, and involves excessive tissue deposition at the junction of the media and adventitia. A less common form of FMD is intimal fibroplasia, which is characterized by irregularly arranged mesenchymal cells within a loose matrix of subendothelial connective tissue and a fragmented internal elastic lamina. Medial hyperplasia and adventitial (or periarterial) hyperplasia are the rarest types of FMD. These subtypes are not mutually exclusive, and characteristics of each can be seen in the same arterial segment [25]. The angiographic appearance as well as the relative frequency of these different subtypes of FMD are summarized in Table 1 [adapted from references 1 and 26].

FMD is a rare disease, which is often diagnosed solely based on angiographic and clinical features. In this case we were fortunate to also have the pathologic correlation, which confirmed the diagnosis based on the findings of excess smooth muscle component and frayed elastic fibers which are characteristic of fibromuscular dysplasia. The pathology and angiographic appearance must both be taken into consideration, as well as the clinical features, and the disseminated nature of vascular disease in this patient.

TEACHING POINT

Involvement of the aorta by FMD is rare, and is mostly detected as a result of complications such as dissection or aneurysm. Hypertension is the most important risk factor for patients presenting with aortic dissection, however, in a patient with diffuse systemic abnormal appearing vessels, other entities including FMD should be considered.

REFERENCES

1. Slovut DP, Olin JW. Fibromuscular Dysplasia. *N Engl J Med* 2004;350:1862-1871. PMID: 15115832
2. Luscher TF, Keller HM, Imhof HG, Greminger P, Kuhlmann U, Largiader et al. Fibromuscular hyperplasia: extension of disease and therapeutic outcome. *Nephron* 1986;44:109-114 (suppl 1). PMID: 2944009
3. Sang CN, Whelton PK, Hamper UM, Connolly M, Kadir S, White RI et al. Etiologic factors in renovascular fibromuscular dysplasia: a case-control study. *Hypertension* 1989;14:472-479. PMID: 2680961
4. Pannier-Moreau I, Grimbert P, Fiquet-Kempf B, Vuagnat A, Jeunemaitre X, Corvol P et al. Possible familial origin of multifocal renal artery fibromuscular dysplasia. *J Hypertens* 1997;15:1797-1801. PMID: 9488242
5. Meszaros I, Morocz J, Szilavi J et al. Epidemiology and clinicopathology of aortic dissection: a population-based longitudinal study over 27 years. *Chest* 2000;117:1271-1278. PMID: 10807810

6. de Paiva Magalhaes E, Fernandes SRM, Zanardi VA et al. Ehlers-Danlos syndrome type IV and multiple aortic aneurysms: a case report. *Angiology* 2001;52:223-228. PMID: 11269788
7. Einstein AJ, Dellegrottaglie S, Rajagopalan S. Triple-barrelled aortic dissection in Ehlers-Danlos syndrome. *Heart* 2006;92:1166. PMID: 16844880
8. Radhi JM, McKay R, Tyrrell MJ. Fibromuscular dysplasia of the aorta presenting as multiple recurrent thoracic aneurysms. *Int J Angiol* 1998;7:215-218. PMID: 9585453
9. Odero A, Bozzani A, Arici V, Agozzino M. Hypoplasia and fibromuscular dysplasia of infrarenal abdominal aorta with downstream aneurysm: case report and review of the literature. *J Vasc Surg* 2008;48:1589-1592. PMID: 19118740
10. Ohteki H, Itoh T, Natsuaki M, Sakurai J, Tazaki H, Watanabe T et al. Atypical coarctation of the thoracic aorta with fibromuscular dysplasia: report of a successful surgical repair and review of the literature. *Nippon Geka Gakkai Zasshi* 1989;90:130-133. PMID: 2649786
11. Vuong PN, Trinh AC, Lagneau P, Camilleri JP. Coarctation of the abdominal aorta and stenosis of the left renal artery with hypertension caused by fibrodysplasia. *Arch Pathol Lab Med* 1989;113:809-811. PMID: 2742463
12. Sousa Uva M, Bical O, Vuong PN, Gigou F, Foiret JC et al. Atypical coarctation of the thoracic aorta caused by fibromuscular dysplasia. *Arch Mal Coeur Vaiss* 1994;9:1233-1236. PMID: 7646238
13. Matsushita M, Yano T, Ikezawa T, Sakurai T, Nimura Y, Shionoya S. Fibromuscular dysplasia as a cause of abdominal aortic aneurysm. *Cardiovasc Surg* 1994;2:615-618. PMID: 7820524
14. Vuong PN, Janzen J, Bical O, Sousa Uva M. Fibromuscular dysplasia causing atypical coarctation of the thoracic aorta: histological presentation of a case. *Vasa* 1995;24:194-198. PMID: 7793154
15. Miyake T, Sakuma M, Shiiya N, Matsui Y, Yasuda K, Hosoda L. A case of fibromuscular dysplasia with abdominal aortic dissecting aneurysm and tubular stenosis of the aorta. *Jpn J Vasc Surg* 1995;4:109-112.
16. Golosovkaia MA, Serov RA, Lebedeva TM. Two cases of fibromuscular dysplasia of the aorta (intimal hyperplasia). *Arkh Patol* 1996;58:62-66. PMID: 8929145
17. Paraf F, Bruneval P. Arterial fibromuscular dysplasia and Bourneville's tuberous sclerosis. *Ann Pathol* 1996;16:203-206. PMID: 8766180
18. D'Souza SJ, Tsai WS, Silver MM, Chait P, Benson LN, Silverman E et al. Diagnosis and management of stenotic aorto-arteriopathy in childhood. *J Pediatr* 1998;132:1016-1022. PMID: 9627596
19. Suarez WA, Kurczynski TW, Bove EL. An unusual type of combined aortic coarctation due to fibromuscular dysplasia. *Cardiol Young* 1999;9:323-326. PMID: 10386705
20. Janzen J, Vuong PN, Rothenberg-Janzen K. Takayasu's arteritis and fibromuscular dysplasia as causes of acquired atypical coarctation of the aorta: retrospective analysis of seven cases. *Heart Vessels* 1999;14:277-282. PMID: 10901482
21. Dejardin A, Goffette P, Moulin P, Verhelst R, Cornu G, De Plaen JF et al. Severe hypoplasia of the abdominal aorta and its branches in a patient and his daughter. *J Intern Med* 2004;255:130-136. PMID: 14687249
22. Gatalica Z, Gibas Z, Martinez-Hernandez A. Dissecting aortic aneurysm as a complication of generalized fibromuscular dysplasia. *Hum Pathol* 1992;23:586-588. PMID: 1568754
23. Brodsky SV, Ramaswamy G, Chander P, Braun A. Ruptured cerebral aneurysm and acute coronary artery dissection in the setting of multivascular fibromuscular dysplasia. *Angiology* 2007;58:764-767. PMID: 18216385
24. Mettinger KL, Ericson K. Fibromuscular dysplasia and the brain. I. Observations on angiographic, clinical and genetic characteristics. *Stroke* 1982;13:46-52. PMID: 7064180
25. Harrison EG Jr, McCormack LJ. Pathologic classification of renal arterial disease in renovascular hypertension. *Mayo Clin Proc* 1971;46:161-167. PMID: 5553126
26. Begelman SM, Olin JW. Fibromuscular dysplasia. *Curr Opin Rheumatol* 2000; 12:41-47. PMID: 10647953

FIGURES

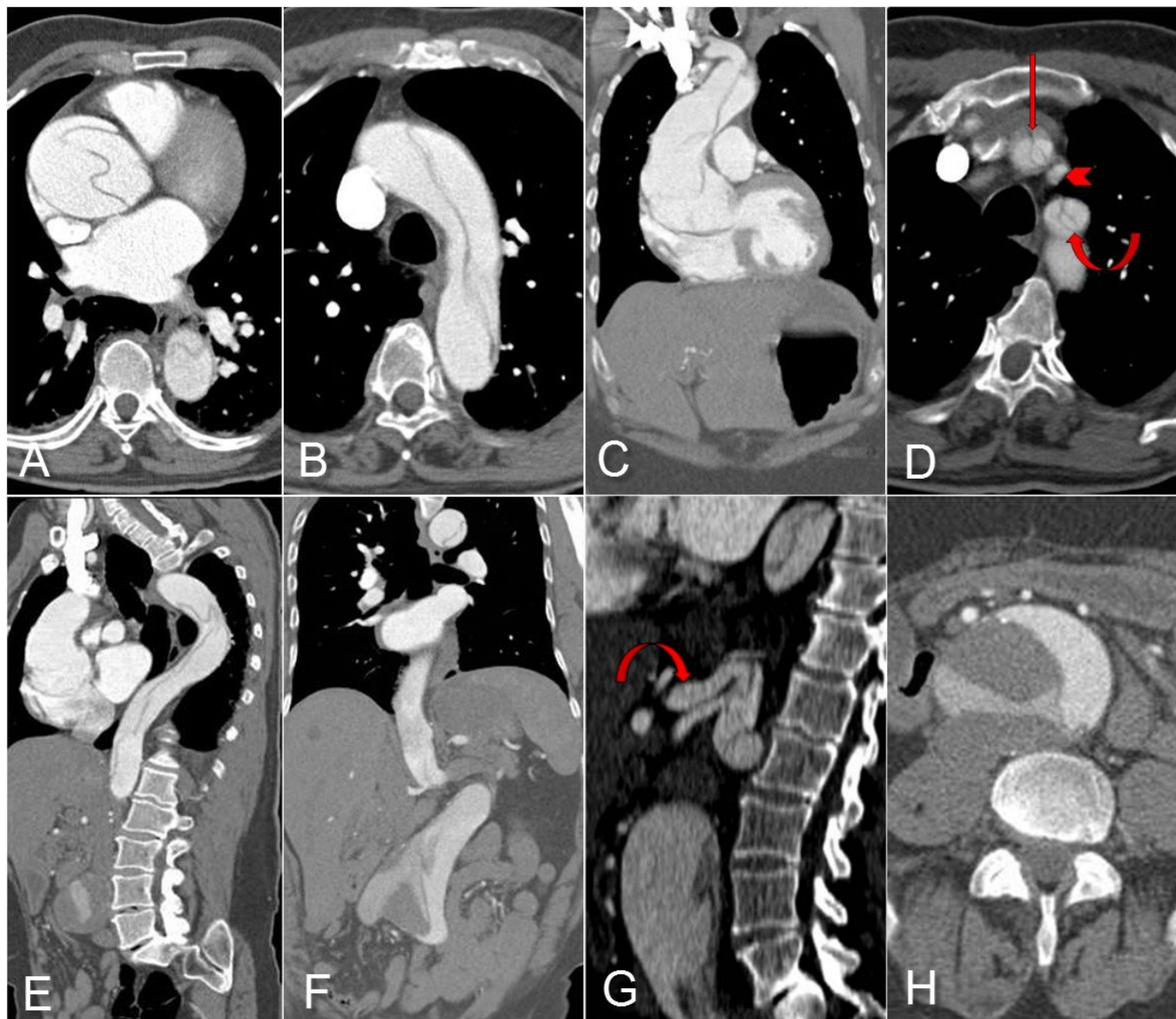


Figure 1: 64 year old male with fibromuscular dysplasia. Axial (A, B, and D) and coronal (C) images from a CT angiogram of the chest, abdomen and pelvis demonstrate an intimal dissection flap extending from the aortic root and across the aortic arch, in keeping with a type A aortic dissection. The aortic root is also noted to be aneurismal. The intimal flap is seen extending cephalad into the left subclavian (curved arrow), left common carotid (arrowhead) and right brachiocephalic arteries (straight arrow) (D). Sagittal (E and G), coronal (F) and axial (H) images from the same series demonstrate the dissection flap extending inferiorly into the abdominal aorta, as well as a large, partially thrombosed abdominal aortic aneurysm extending to the aortic bifurcation. Aneurysmal dilatation of the proximal celiac artery is shown in H (curved arrow). (CTA protocol: 725 Eff mAS, 120 kV, 0.6 mm slice thickness, pitch/rotation time 0.2/0.33s, 116 mL of Optiray 320 contrast (28 mL injected at 5.5 mL/second followed immediately by 88 mL injected at 4.4 ml/second)).

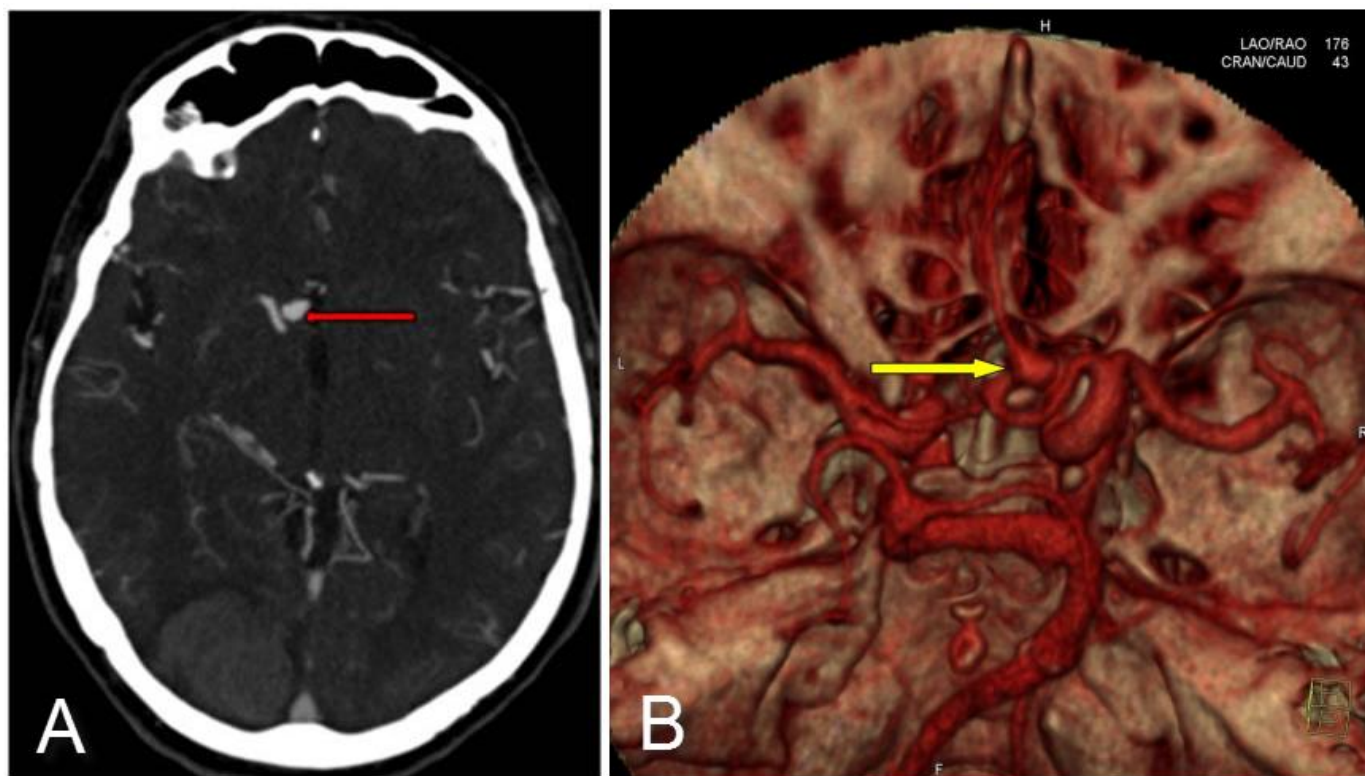


Figure 2: 64 year old male with fibromuscular dysplasia. Axial image from a CT angiogram of the circle of Willis demonstrates a wide-necked lobulated aneurysm arising from the proximal A2 segment of the right anterior cerebral artery (red arrow) (A). Volume-rendered 3D reformat from the same image series also shows the aneurysm, denoted by a yellow arrow (B). (CTA protocol: 300 mAs, 120 kV, 1 mm slice thickness, pitch/rotation time 0.9/0.5s, 100 mL of Optiray 320 contrast injected at 5 mL/second).

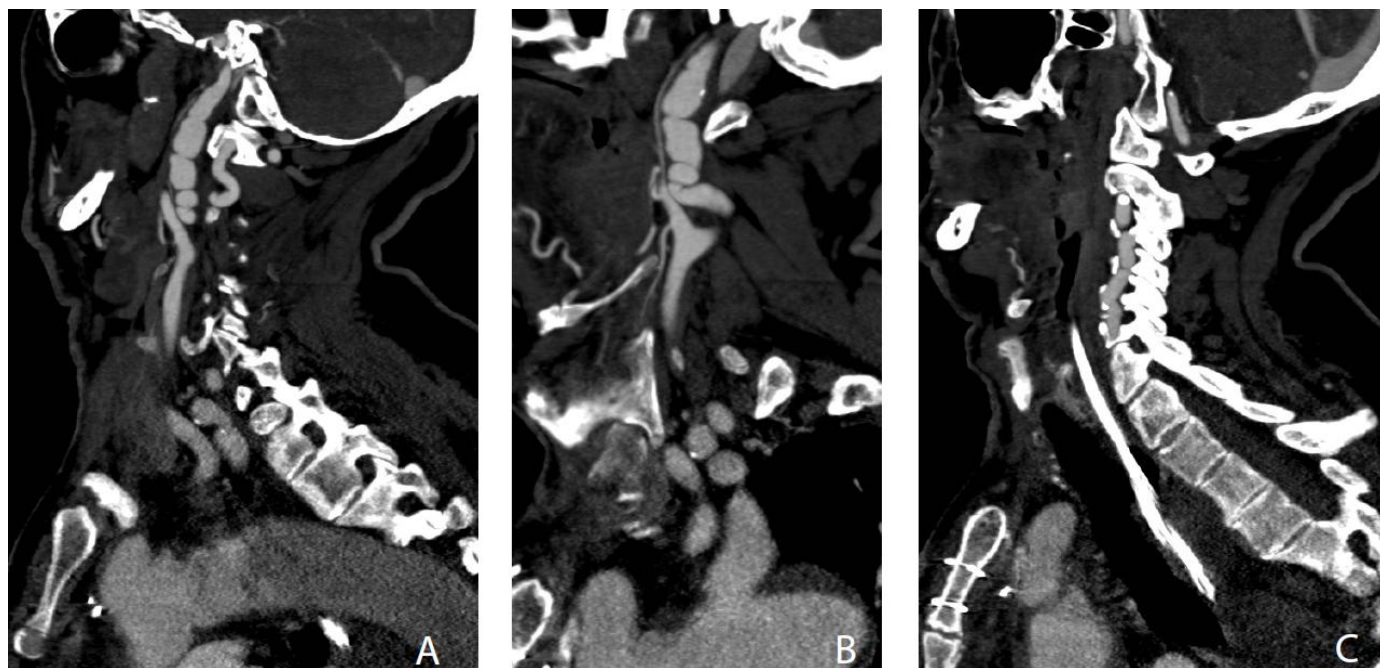


Figure 3: 64 year old male with fibromuscular dysplasia. A and B: Sagittal and sagittal oblique images through the left internal carotid artery show a markedly tortuous appearing vessel, with a beaded appearance and focal segments of aneurysmal dilatation. Sagittal image through a segment of the left vertebral artery (C), which has a similar appearance, with the vessel appearing tortuous with an irregular beaded contour. (CTA protocol: 300 mAs, 120 kV, 1 mm slice thickness, pitch/rotation time 0.9/0.5s, 100 mL of Optiray 320 contrast injected at 5 mL/second).

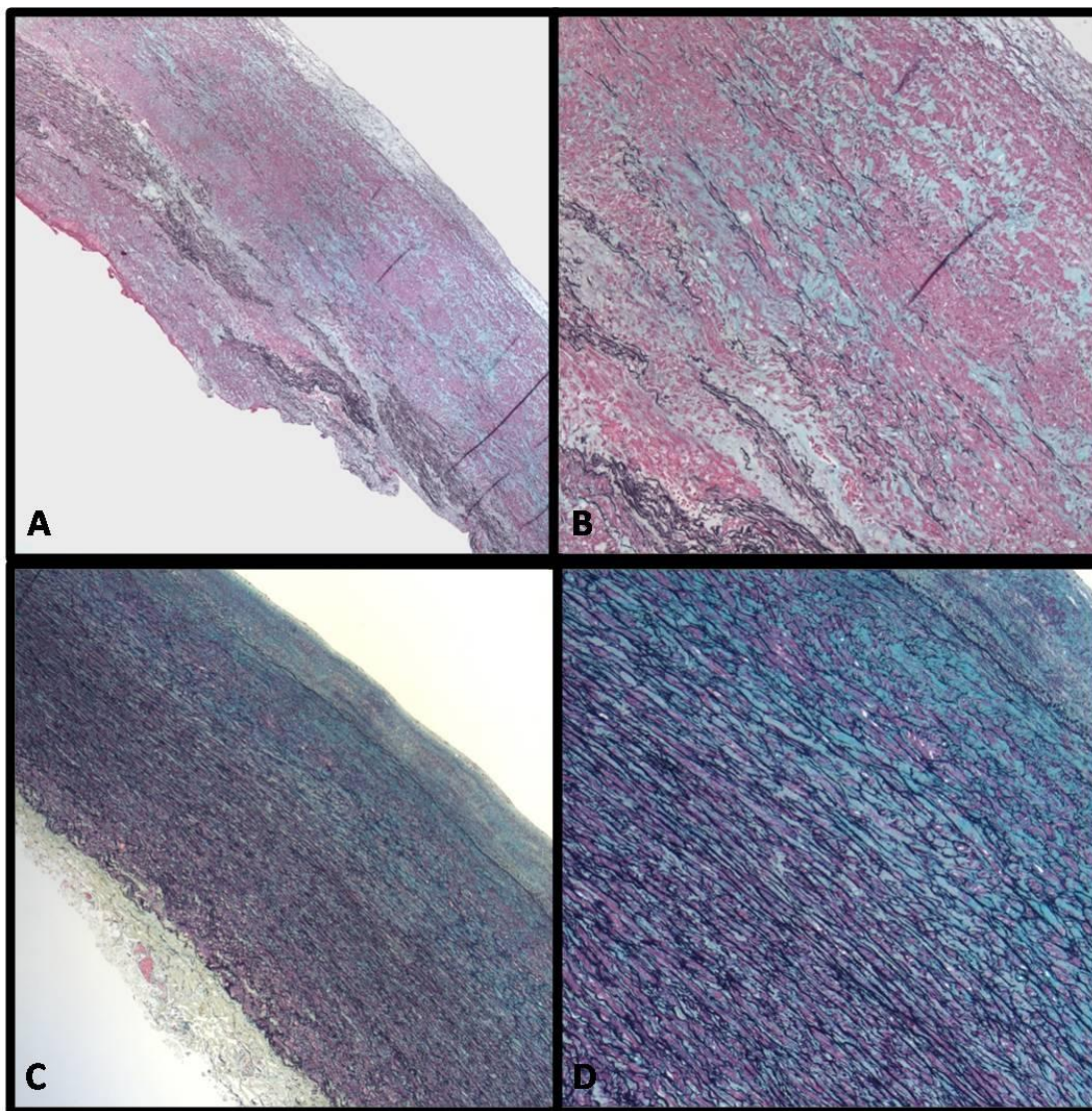


Figure 4: 64 year old male with type A aortic dissection with underlying fibromuscular dysplasia. Movat's Pentachrome stained sections from the operative specimen of the thoracic aorta showing the excess smooth muscle component and frayed elastic fibers characteristic of fibromuscular dysplasia at 4x (A) and 10x (B). These are compared to the appearance of a normal aorta from a different patient at 4x (C) and 10x (D).

| Classification | Estimated frequency | Angiographic appearance |
|---|---------------------|---|
| Medial fibroplasia | 75-80% | -“string of beads” appearance, where the diameter of beading is larger than the normal caliber of the artery -mid-to distal portion of artery affected |
| Perimedial fibroplasia | 10-15% | -focal stenoses and multiple constrictions -“beading” in which the “beads” are smaller than the caliber of the artery |
| Medial hyperplasia | 1-2% | -concentric smooth stenosis (may be indistinguishable from intimal disease) |
| Intimal fibroplasia | <10% | -focal concentric stenosis, or -long, smooth narrowing, or -redundancy of the artery |
| Adventitial (periarterial) fibroplasia | <1% | -sharply localized tubular areas of stenosis |

Table 1: Imaging findings for histologic subtypes of FMD. *References: [1] and [26]*

| Differential diagnosis | Chest X-ray | CT angiography | Other clues on imaging (not including findings of aortic dissection) |
|--|--|--|--|
| General imaging features in aortic dissection | -widened mediastinum -left apical cap -tracheal deviation -depression of left mainstem bronchus -esophageal deviation -loss of paratracheal stripe -pericardial effusion | -intimal flap between true and false lumens -delayed flow in false lumen (less dense) -compression of true lumen by expanding false lumen -pericardial effusion | |
| Hypertension/ atherosclerosis | -may see displaced intimal calcification | -atherosclerotic calcification along aortic arch -aortic aneurysm | -left ventricular hypertrophy |
| Ehlers-Danlos syndrome | | -aortic aneurysm | -other aneurysms (medium and large vessels) -acrogeria -ecchymoses and hematoma |
| Marfan syndrome | | -aortic annulus ectasia -aortic aneurysm | -arachnodactyly -dural ectasia -bullae and pneumothorax |
| Fibromuscular dysplasia | | -aortic aneurysm | -renovascular FMD: classic "string of beads" appearance -intracranial aneurysms -other aneurysms |

Table 2: Differential diagnoses for aortic dissection.

ABBREVIATIONS

CT: Computed tomography
CTA: Computed tomography angiography
FMD: Fibromuscular dysplasia

KEYWORDS

Fibromuscular dysplasia; Aortic dissection

Online access

This publication is online available at:
www.radiologycases.com/index.php/radiologycases/article/view/457

Peer discussion

Discuss this manuscript in our protected discussion forum at:
www.radiopolis.com/forums/JRCR

Interactivity

This publication is available as an interactive article with scroll, window/level, magnify and more features.
Available online at www.RadiologyCases.com

Published by EduRad



www.EduRad.org