PHILIPS

Ultrasound

Case study

Diagnosing fibromuscular dysplasia using EPIQ ultrasound and CT angiography

Ankit R. Shah, MD, FACC, RPVI and Jan M. Sloves, RVT, RCS, FASE, Mount Sinai Heart, New York, NY

Overview

A 55-year-old woman with a past medical history of hypertension and hyperlipidemia was diagnosed with a myocardial infarction due to a spontaneous coronary artery dissection four years prior. She presented to the hospital with acute onset of chest pain. ECG revealed acute ischemic changes in the anterolateral leads. She had elevated cardiac enzymes with a peak troponin-I level of 15.6. Findings were consistent with an acute coronary syndrome. She was taken for an urgent cardiac catheterization. The angiogram revealed a long and narrowed segment of the mid and distal left anterior descending artery. The other coronary arteries were patent with minimal atherosclerosis. She was diagnosed as having a spontaneous coronary artery dissection and was treated medically with a heparin drip. Two days later, she continued to have chest pain and she was taken for a repeat cardiac catheterization. The degree of stenosis in the mid and distal segments of the left anterior descending artery was worse and stents were placed. Her symptoms resolved, and she was discharged on lifelong low-dose aspirin therapy. Her cardiac function was mildly reduced after the myocardial infarction. Since then, she has felt clinically well, and her cardiac function has normalized.

Ultrasound findings

Given that the patient was diagnosed with a spontaneous coronary artery dissection, she underwent screening evaluation for evidence of a connective tissue disorder in the other arterial beds. Physical examination was significant for bilateral carotid bruits. A carotid duplex was performed on a Philips EPIQ 7 with a L12-3 linear transducer. The bilateral cervical internal carotid arteries were imaged from multiple approaches in the longitudinal and transverse image planes, distally under the mandible. Both vessels were interrogated for the presence of atherosclerotic plaque, stenosis, dissection, aneurysm and tortuosity. Her duplex study findings were without evidence of significant atherosclerotic plaque. Grayscale imaging with the L12-3 transducer did not demonstrate any discernable arterial wall defects within the mid or distal ICA segments. However, color flow revealed turbulence and tortuosity. Pulsed wave Doppler displayed increased peak systolic velocity (PSV = 250 cm/s) and end diastolic velocity (EDV = 102 cm/s) in these segments bilaterally **(Figure 1)**.

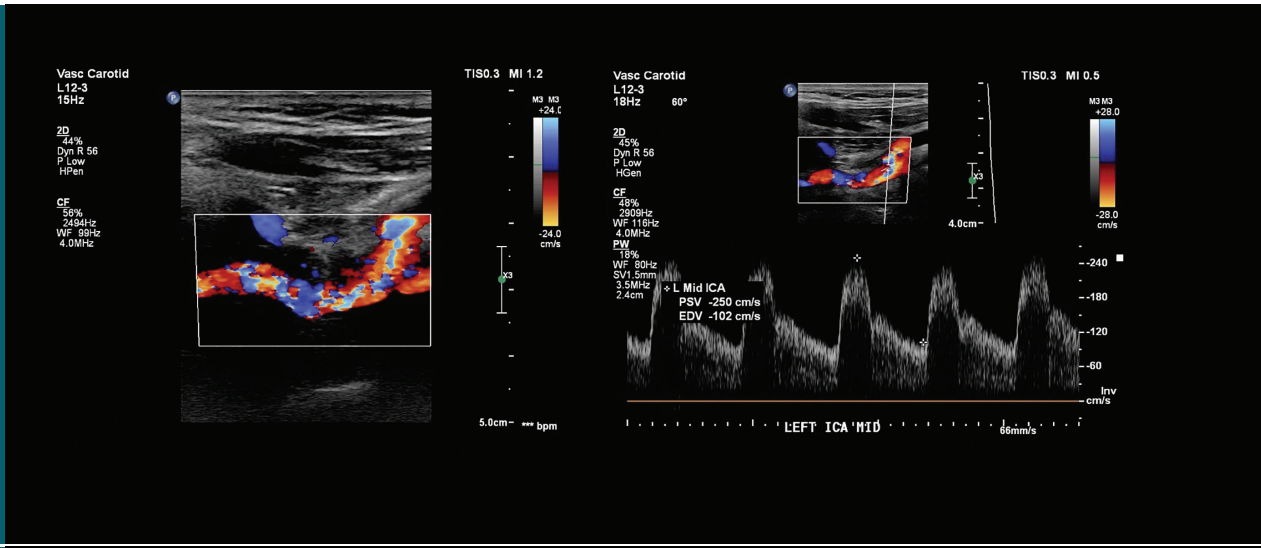


Figure 1
Turbulence and tortuosity revealed by color flow imaging with the L12-3 transducer.

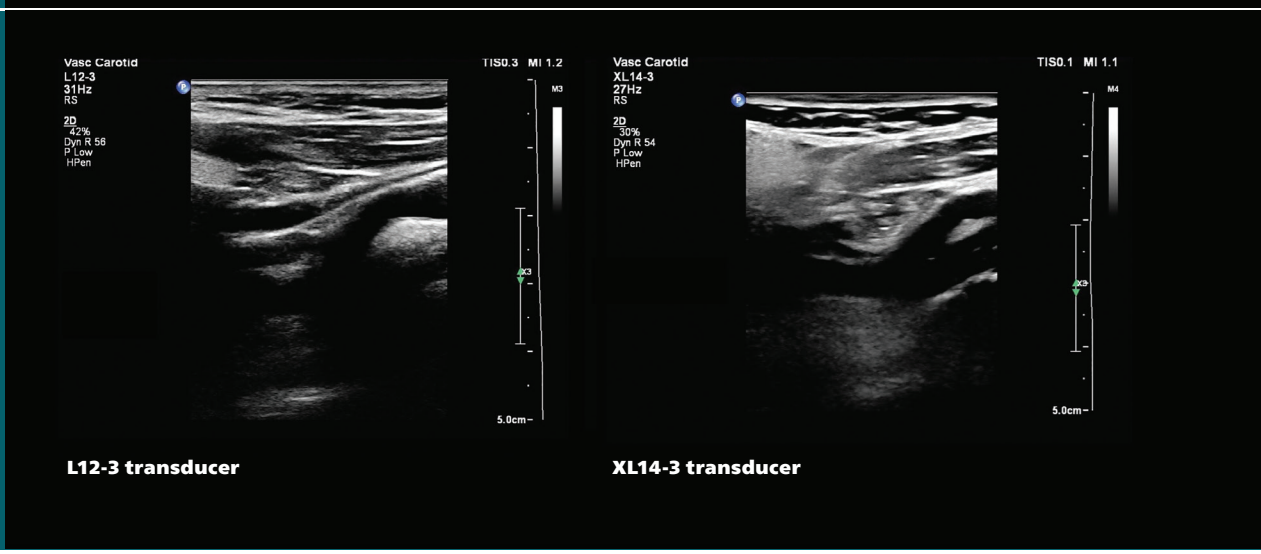


Figure 2
Improved grayscale resolution provided by the XL14-3 transducer illustrated irregularities not shown by the L12-3 transducer.

In light of these findings, the XL14-3 xMATRIX transducer was utilized to optimize the imaging of the mid-distal ICA segments. The XL14-3 transducer provided improved grayscale resolution in comparison to the L12-3 transducer. Grayscale imaging illustrated morphological arterial luminal irregularities within the area of turbulence and increased velocity **(Figure 2)**.

Next, a 3D rendering was used to further interrogate this segment with color power angio (CPA), which created a vessel cast of the lumen. The vessel cast clearly defined the classic beading formation consistent with fibromuscular dysplasia (FMD) **(Figure 3)**.

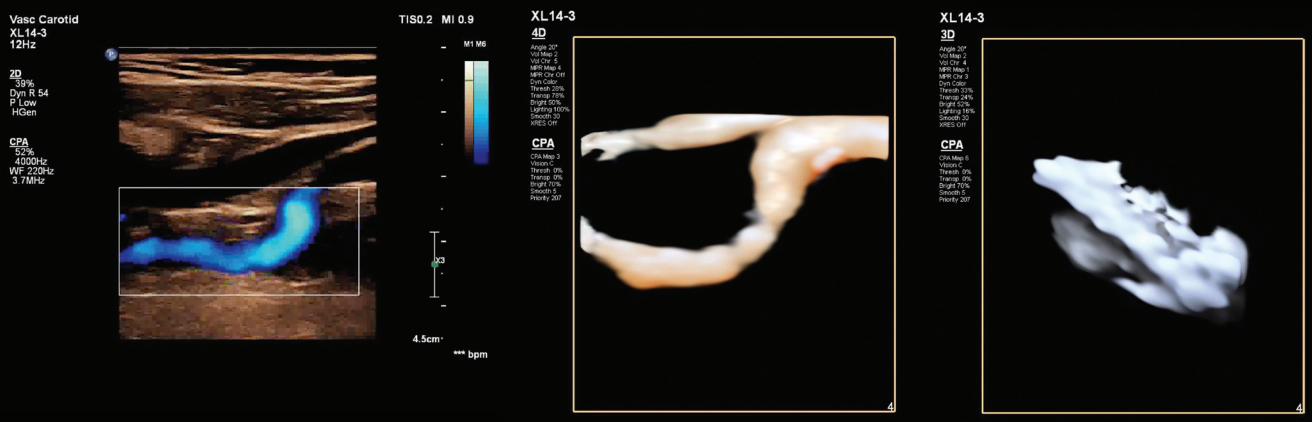
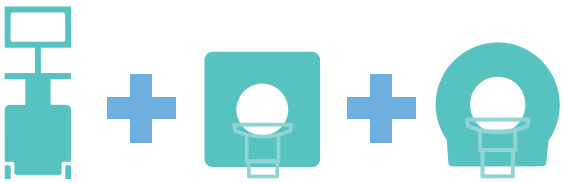


Figure 3
Using the XL14-3 transducer, color power angio (CPA) was used to create a vessel cast that helped identify FMD.



Evaluation by CT and MR

Computed tomography angiography of the neck confirmed the diagnosis **(Figure 4)**. The patient underwent a renal artery duplex, which revealed bilateral elevated velocities, turbulence, and tortuosity in the mid and distal segments, also consistent with FMD. Further evaluation with magnetic resonance angiography revealed no other associated abnormalities within the abdomen and pelvis.

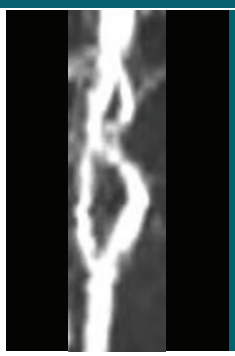


Figure 4
FMD diagnosis confirmed with CT angiography of the neck.

Diagnostic considerations for FMD

FMD is a nonatherosclerotic, noninflammatory cause for arterial stenosis, occlusion, aneurysm, dissection, and arterial tortuosity.[1] The most affected arteries are the internal carotid and renal arteries, followed by the vertebral, visceral and external iliac arteries. Nearly two-thirds of patients have multiple affected arteries.[2] Among adults, approximately 90% of cases are in women.[3] According to the United States FMD Registry, the mean age at diagnosis was 52 years.[2] FMD is commonly classified by angiographic appearance. The two subtypes are multifocal FMD, which is the most common, and focal FMD. Multifocal FMD has an appearance of a "string of beads", and corresponds pathologically to medial fibroplasia, the most common histologic type. Focal FMD has the appearance of a "circumferential or tubular stenosis" and corresponds pathologically to intimal fibroplasia.[4] Pathological specimens are rarely obtained, therefore classification based on histology is not done.

The pathogenesis of FMD remains unknown. Genetics is thought to play an important role in FMD,[1] with some studies reporting an autosomal inheritance pattern with variable penetrance.[5] Hormonal effects are thought to also play an important role in the pathogenesis of FMD, given the predominance of affected women of childbearing age, although this has not been studied.

The common clinical manifestations of FMD are dissection of a major artery leading to ischemia due to stenosis or occlusion of an artery, distal embolization of intravascular thrombi, and rupture of an aneurysm.[6] In men, clinical manifestations related to renal artery FMD, such as hypertension or flank pain, is seen more often. In contrast, women tend to have more cerebrovascular FMD clinical manifestations, such as headaches, neck pain, or carotid bruits.[7] FMD should be considered in the following scenarios: resistant hypertension; early onset hypertension; sudden increase in serum creatinine with the initiation of angiotensin-converting enzyme (ACE) inhibitor or angiotensin receptor blocker (ARB); spontaneous dissection of a peripheral (carotid, vertebral, renal) or coronary artery; aneurysm in a visceral, carotid, vertebral or intracranial vessel; epigastric or carotid bruits, especially in younger patients.

The diagnosis of FMD is most commonly made by noninvasive testing. The most common modalities are computed tomography angiography (CTA), magnetic resonance angiography (MRA) and duplex ultrasonography. The need for catheter-based digital subtraction angiography is less frequent. Duplex ultrasound is a very good, noninvasive mode of imaging the carotid, vertebral and renal arteries. However, the quality is highly dependent on operator expertise. When this is not available, CTA or MRA are better choices, with CTA having better spatial resolution.[1]

FMD management and treatment

The management and treatment of FMD depends on the clinical manifestation. In general, medical treatment focuses on antihypertensive therapies and monitoring of renal function. Revascularization should be considered if there is evidence of ischemia or infarction, resistant hypertension or worsening renal function.

Balloon angioplasty is the preferred mode of revascularization over stenting. If an aneurysm is detected, the use of ultrasound or MRA to follow the size of the aneurysm is recommended over CTA to limit radiation exposure. Most clinicians believe that multifocal FMD is not a progressive disease.[8]

Conclusion

Multimodality imaging involving ultrasound performed by experienced operators is useful in the diagnosis of FMD. Duplex ultrasound offers quality noninvasive imaging of the carotid, vertebral and renal arteries, helping clinicians arrive at a definitive diagnosis.

References

1 Gornik HL, Persu A, Adlam D, et al. First International Consensus on the diagnosis and management of fibromuscular dysplasia. Vasc Med. 2019;24:164.

2 Olin JW, Froehlich J, Gu X, et al. The United States Registry for Fibromuscular Dysplasia: results in the first 447 patients. Circulation. 2012;125:3182.

3 Green R, Gu X, Kline-Rogers E, et al. Differences between the pediatric and adult presentation of fibromuscular dysplasia: results from the US Registry. Pediatr Nephrol. 2016;31:641.

4 Olin JW, Gornik HL, Bacharach JM, et al. Fibromuscular dysplasia: state of the science and critical unanswered questions: a scientific statement from the American Heart Association. Circulation. 2014;129:1048.

5 Perdu J, Boutouyrie P, Bourgain C, et al. Inheritance of arterial lesions in renal fibromuscular dysplasia. J Hum Hypertens. 2007;21:393.

6 Olin JW, Sealove BA. Diagnosis, management, and future developments of fibromuscular dysplasia. J Vasc Surg. 2011;53:826.

7 Kim ES, Olin JW, Froehlich JB, et al. Clinical manifestations of fibromuscular dysplasia vary by patient sex: a report of the United States registry for fibromuscular dysplasia. J Am Coll Cardiol. 2013;62:2026.

8 Kadian-Dodov D, Goldfinger JZ, Gustavson S, Olin JW. Natural History of Cervical Artery Fibromuscular Dysplasia and Associated Neurovascular Events. Cerebrovasc Dis. 2018;46:33.

www.philips.com

Printed in the Netherlands.
4522 991 78101 * NOV 2022