A case of mesenteric ischemia secondary to Fibromuscular Dysplasia (FMD) with a positive outcome after intervention

Neal C. Patel¹, William C. Palmer², Kanwar R. S. Gill³, Michael B. Wallace⁴

¹Division of Gastroenterology, Mayo Clinic, Arizona, Scottsdale, AZ, USA; ²Department of Internal Medicine, Mayo Clinic Florida, Jacksonville, FL, USA;

³Gould Medical Foundation, Modesto, CA, USA; ⁴Division of Gastroenterology & Hepatology, Mayo Clinic Florida, Jacksonville, FL, USA

Key words: fibromuscular dysplasia, mesenteric ischemia, abdominal pain

Abbreviations: FMD, fibromuscular dysplasia

Abstract

Fibromuscular dysplasia (FMD) is a non-inflammatory, non-atherosclerotic angiopathy, which commonly affects the renal and internal carotid arteries. Although rare, FMD has the potential of involving the mesenteric vasculature. Due its low incidence and relatively little knowledge concerning its risk factors and etiology, actual diagnosis of FMD involving the mesenteric vessels requires a very high degree of suspicion. Upon review of the few reported cases of FMD causing mesenteric ischemia, it is clear that therapeutic interventions are rarely discussed and that positive outcomes are even more uncommon. Herein, we present the case of a 47 year-old female with mesenteric ischemia secondary to FMD, which was diagnosed and treated originally with angioplasty, then repeat angioplasty with stent placement, and finally with a bypass graft. Ultimately, the patient had a positive outcome, including eight month follow-up.

Introduction

Fibromuscular dysplasia (FMD) is a rare non-atherosclerotic, non-inflammatory segmental occlusive disorder of medium-sized vessel arteries, and may be intimal, medial, or periarterial depending on the site of fibro-muscular proliferation.¹ It has an incidence of approximately 1% and occurs predominantly in young to middle-aged women.² The most commonly involved sites include renal arteries (60%-75%) and cerebral arteries (25%-30%). The less commonly affected sites include the mesenteric, subclavian, and iliac arteries.² Several cases of mesenteric ischemia from FMD have been documented.³ Herein, we present the case of a female who was diagnosed early enough in her disease process to allow for multiple therapeutic interventions, which eventually resulted in a positive outcome.

Case Report

A 47 year-old female non-smoker presented as a referral for evaluation of ongoing symptoms, which included nausea and abdominal pain. Her past medical history was significant for mitral valve prolapse without regurgitation and irritable bowel syndrome. Home medications included pantoprazole daily

and oral contraceptives. Family history was notable only for hypercholesterolemia.

She was seen approximately one year prior for similar symptoms, at which time an esophagogastroduodenoscopy with endoscopic ultrasound revealed layering gallbladder sludge and no other gastric, pancreatic, or celiac axis abnormalities. The cause of her abdominal pain was thought to be related to a biliary pathology, therefore a cholecystectomy was performed. Following cholecystectomy, the patient had resolution of her symptoms. However, eight months later she began feeling persistent nausea and early satiety, with episodes of burning painful sensations in her upper abdomen. She moderated her dietary intake and continued with very minimal alcohol consumption. However, she continued to experience 1-2 symptomatic episodes per week. Specifically, she described an initial burning sensation followed by a stabbing pain which was followed by a sense of urgency to have a bowel movement.

Physical exam was notable for a two pound weight gain from one year prior with a body mass index of 16.4 kg/m² and a sharp crescendo, decrescendo bruit in the upper mid abdomen, which was also palpable. The results of the following laboratory investigations were within normal limits: blood chemistry profile, liver enzymes, cholesterol, triglycerides, platelets, hemoglobin, and hematocrit. Further lab investigations revealed a normal TSH, amylase, lipase, ANA, ANCAs, ESR, and IgG subclass 4.

An abdominal vascular ultrasound revealed markedly elevated pressures in the superior mesenteric artery suspicious for the presence of a stenosis (velocities of aorta= 96 cm/sec; celiac axis=

*Correspondence to: William C. Palmer; Email: Palmer.william@mayo.edu
Submitted: Sep/04/2012; Revised: Sep/25/2012; Accepted: Oct/31/2012
Previously published online: www.landesbioscience.com/journals/jig
DOI: 10.4161/jig.23747

70 cm/sec; SMA (origin) = 444 cm/sec; SMA (mid) 519 cm/sec; SMA (distal) 367 cm/sec; IMA = 65 cm/sec). Magnetic resonance imaging and angiography of the abdomen confirmed very severe narrowing of the superior mesenteric artery at its origin, with marked hypertrophy of the gastroduodenal artery and pancreaticoduodenal arteries which were supplying collateral flow from the celiac axis to the mid SMA. The celiac axis was not well visualized but also appeared to be narrowed; however, the inferior mesenteric artery was patent.

A mesenteric arteriogram was performed showing that the SMA to the celiac axis was completely occluded, with the stenotic process also involving both the origins of the splenic and the common hepatic arteries. The pressure in the distal SMA was found to be 90/63 mmHg, and abdominal aorta pressure at the same time was 128/67 mmHg. Balloon angioplasty was used to dilate the proximal stenosis. Post-angioplasty, there was no significant residual stenosis in the SMA. Repeat pressures at this time were 112 mmHg systolic in the distal SMA and 123 mmHg systolic in the abdominal aorta. Angioplasty of the celiac axis was attempted; however, the trajectory of this vessel and the severity of the stenosis did not allow passage of a catheter or angioplasty balloon. The patient had a good clinical response with resolution of her symptoms despite the remaining stenosis of the celiac axis, thus no further interventions were pursued at the time.

Twenty months later, the patient returned to the clinic with similar complaints of post-prandial abdominal pain and nausea, thus a repeat arteriogram was performed. The SMA was once again stenotic at the previous angioplasty site. Repeat angioplasty was performed with two stents also placed in the SMA. The celiac axis had continued stenosis but was once again inaccessible. The patient's symptoms initially improved, however she had a reoccurrence of severe abdominal pain 3 days later. Repeat arteriogram showed in stent thrombosis while on anti-platelet medication, and thrombolytic therapy with thrombectomy

was performed. Post-therapy the patient's chronic nausea had resolved, however she continued to feel some abdominal pain with large meals. Five months later, the patient underwent an aorto-superior mesenteric artery and an aorto-hepatic artery bypass. Subsequently, the patient had complete resolution of all previous symptoms and has remained asymptomatic at the four, six, and eight month follow up.

Discussion

Fibromuscular dysplasia is a non-inflammatory, non-atherosclerotic angiopathy which is not well understood, due to its low incidence of approximately 1%. The typical patient with FMD is a white woman in her 30-40's.³ Although many theories exist on the pathogenesis, risk factors, and etiology of FMD exist, no definitive associations have been made. Furthermore, FMD can closely mimic much more common diseases such as atherosclerosis and vasculitis.⁴ Although there has been a widely described classic "string of beads" appearance of involved vessels on angiogram, this is typically only seen in medial fibroplasias type of FMD, which is a small subset of an already rare condition.⁵ It is unclear why the celiac artery appeared normal on the initial endoscopic ultrasound, as abdominal ultrasound has been used to identify mesenteric FMD.^{3,6} On literature review, the authors did not find a documented case of mesenteric FMD identified on endoscopic ultrasound. For all of these reasons, the diagnosis of FMD presents a difficult clinical challenge for physicians.

Fibromuscular dysplasia occurs most commonly in middle-aged women of about 39 years for renovascular disease, and 50 years for aortocranial disease.⁷ However, the disease can occur in males, and does not have an identified association with sex hormones. Mechanical factors have been considered, such as cyclic stretching of arterial smooth muscle cells and trauma to the blood vessel wall.³ Cigarette smoking has also been investigated as

Figure 1. T1 postcontrast sagittal which demonstrates smooth segment narrowing of the proximal SMA and celiac

Figure 2. Contrasted enhanced CT with a sagittal reformatted image that demonstrates a potent proximal SMA stent

a risk factor for the development of FMD.⁸ A previous study of 20 families by Rushton, et al. found that 60% of cases demonstrated an autosomal dominant pattern with variable penetrance, with the remaining 40% developing presumed new mutations.⁹ This would explain the lack of family history in our patient.

In the case above, our patient presented with diffuse abdominal pain, nausea and anorexia. However, on physical examination, an abdominal bruit and a high degree of suspicion for FMD pointed to a need for further evaluation of the patient's abdominal vasculature. Although a definitive diagnosis can only be made with tissue biopsy, without any signs or risk factors for atherosclerotic disease and no lab findings consistent with an inflammatory vasculitis, FMD is the likely diagnosis. Symptomatic FMD of the mesenteric arteries is rare, and there are only a few case reports describing this phenomenon.³ When reviewing the literature, it is clear that therapeutic options are rarely discussed and, unfortunately, most cases resulted in death due to the delay in diagnosis.³ Due to a high degree of suspicion, our patient underwent aggressive therapeutic interventions without delay including multiple arteriograms with dilations and stent placements. Eventually, due to reoccurrence of symptoms with an in-stent thrombosis, our patient benefited from vascular bypass surgery. At her eight month follow-up, our patient had complete resolution of her symptoms and leading a normal lifestyle.

Ultimately, our case presents a few important points. First, the diagnosis of FMD requires a high degree of suspicion.

Second, although rare, FMD can severely affect the mesenteric vasculature without involvement of more common sites such as renal or cerebral vessels. Finally, after multiple attempts to provide revascularization with percutaneous transluminal angioplasty and stent placements, the patient did not have complete resolution of her symptoms until bypass surgery was conducted.

Conflicts of Interest

All authors have nothing to disclose and have no conflicts of interest.

References

1. Harrison EG, Jr., McCormack LJ. Pathologic classification of renal arterial disease in renovascular hypertension. *Mayo Clin Proc* 1971; 46: 161-7.
2. Slovut DP, Olin JW. Fibromuscular dysplasia. *N Engl J Med* 2004; 350:1862-71.
3. Guill CK, Benavides DC, Rees C, Fenves AZ, Burton EC. Fatal mesenteric fibromuscular dysplasia: a case report and review of the literature. *Arch Intern Med* 2004; 164:1148-53.
4. Jones HJ, Staud R, Williams RC, Jr.. Rupture of a hepatic artery aneurysm and renal infarction: 2 complications of fibromuscular dysplasia that mimic vasculitis. *J Rheumatol* 1998; 25:2015-8.
5. Luscher TF, Lie JT, Stanson AW, Houser OW, Hollier LH, Sheps SG. Arterial fibromuscular dysplasia. *Mayo Clin Proc* 1987; 62:931-52.
6. Begelman SM, Olin JW. Fibromuscular dysplasia. *Curr Opin Rheumatol* 2000; 12: 41-7.
7. Mettinger KL, Ericson K. Fibromuscular dysplasia and the brain. I. Observations on angiographic, clinical and genetic characteristics. *Stroke* 1982; 13:46-52.
8. 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; 14:472-9.
9. Rushton AR: The genetics of fibromuscular dysplasia. *Arch Intern Med* 1980; 140: 233-6.