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CASE REPORT

Superior Mesenteric Artery Stenosis With Dissection Secondary to Fibromuscular Dysplasia: A Case Report

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Abstract

Superior Mesenteric Artery (SMA) stenosis increases with advancing age and is usually associated with atherosclerosis or thrombosis. Mesenteric ischemia occurs when the blood flow to the visceral organs is inadequate to meet their metabolic needs. While it can be asymptomatic, the patient can experience chronic or acute mesenteric ischemia symptoms. Although not frequently encountered, Superior Mesenteric Artery stenosis can be rarely caused by fibromuscular dysplasia (FMD). Here we report a case of a 66-year-old female with SMA stenosis and subsequent mesenteric ischemia secondary to fibromuscular dysplasia. The main objective of this case report is to raise awareness about the occurrence of mesenteric ischemia from FMD due to a limited understanding of its risk factors and etiology.

Keywords: Mesenteric, Stenosis, Ischemia, Fibromuscular, Celiac

1. Introduction

Mesenteric artery stenosis is the narrowing of the arteries that supply blood to the small intestine, resulting in inadequate blood flow and intestinal ischemia. It is most commonly caused by atherosclerosis, although it can also result from extensive fibromuscular disease or trauma.[1] Fibromuscular dysplasia (FMD) is a non-inflammatory, non-atherosclerotic condition that usually affects the renal and internal carotid arteries but can rarely involve the mesenteric vasculature. To this date, the exact etiology of fibromuscular dysplasia (FMD) remains inconclusive. Genetic and environmental factors have been linked to the development of FMD.[2,3] The three major arteries that can be affected by mesenteric artery stenosis are the celiac trunk, superior mesenteric artery (SMA), and inferior mesenteric artery (IMA). There should be stenosis of at least two of the these three major splanchnic arteries, with the SMA being the most commonly involved. Duplex ultrasound can determine the presence of significant stenosis (≥70%), velocity parameters are utilized in duplex ultrasound. Studies have reported sensitivities and specificities of about 90% when compared to angiography, indicating that duplex ultrasound is reasonably reliable for ruling out the diagnosis of chronic mesenteric ischemia. Gadolinium-enhanced magnetic resonance angiography (MRA) is another reasonable diagnostic tool for mesenteric artery stenosis.[1] Typical symptoms of this condition are post-prandial epigastric pain or abdominal distension, weight loss, abdominal vascular murmur (which is present in only about 60% of patients), and unexplained chronic gastroduodenal ulcer or right colitis, as the SMA supplies blood to these areas.[4] This case study highlights an exceptional instance of a patient with complicated SMA stenosis resulting from FMD.

2. Case report

A 66 year-old African-American female with medical history of hypertension, hyperlipidemia came to the emergency department with chief complaints of chronic intermittent abdominal pain that has been going on intermittently for one year.

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She mentions severe (10/10) burning, post prandial pain in the left lower quadrant. She reported significant weight loss of about 15 lbs over 1 year due to pain and associated non bilious vomiting & watery diarrhea the last one year. Her rest of the review of systems was normal. She had no recent travel history, any change in medications, tobacco use, any excessive alcohol use. She smokes recreational marijuana. Her medications include atorvastatin, Zetia, Aldactone and multivitamin supplements. She is allergic to adhesives and there is a history of pancreatic cancer in maternal grandmother.

Her vitals were stable at rest and physical examination was positive only for tenderness along lower quadrants without any positive signs of epigastric bruit, any changes for intestinal perforation, renal colic or any pelvic inflammatory conditions. Complete blood count revealed hemoglobin of 10.4 g/dl (normal:12–16 g/dl), with hematocrit of 36% (Normal: 36%–48%). Her comprehensive metabolic panel was unremarkable. Lactic acid was 2.8 mmol/dl (normal: 0.5–1 mmol/L). Lipase and amylase levels were in normal range.

Abdominal computed topography (CT) showed severe stenosis of the celiac axis at its origin without poststenotic dilation along with persistent diffusely and severely stenotic appearance of SMA from abdominal aorta with concern for vasculitis. Bilateral renal vessels were patent (Fig. 1). CT angiography (CTA) abdomen showed severe stenosis of SMA and celiac artery with post stenotic dilation changes (Fig. 1). A magnetic resonance angiography (MRA) abdomen showed similar findings of SMA and celiac axis stenosis without any flow limitation. A duplex ultrasound (US) celiac/Mesenteric vessels showed high-grade stenosis and elevated velocities in the celiac artery with diffuse hemodynamically significant disease (Fig. 2- images a, b, c) (Table 1).

A complete autoimmune panel was done which was negative. Viral panel including Hepatitis B virus (HBV), Hepatitis C virus (HCV) and Human immunodeficiency virus (HIV) were non-reactive. Esophagogastro duodenoscopy (EGD) along with push enteroscopy and colonoscopy were done to rule out any further causes of anemia but there was no evidence of significant pathology. Differential diagnosis of fibromuscular dysplasia versus segmental arterial mediolysis (SAM) was considered at this point as the probability of autoimmune vasculitis is low due to negative autoimmune panel. Her symptoms started to improve likely given the fact she has stayed NPO. Diet was gradually advanced and she was discharged with trial of oral prednisone 40 mg with subsequent taper for a period of three weeks without much improvement.

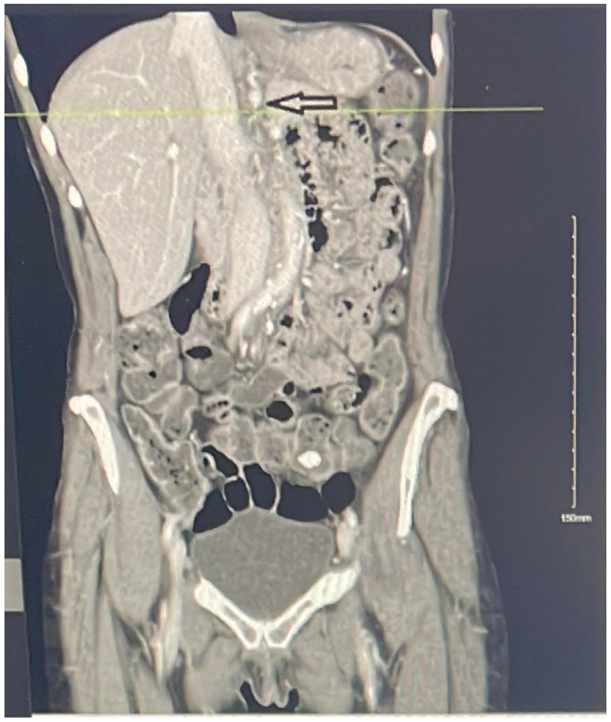

Fig. 1. Shows CT angiogram of abdomen showing persistent diffusely and severely stenotic appearance of the superior mesenteric artery from the abdominal aorta, severe stenotic celiac axis at its origin with now post stenotic dilation.

She presented to the Emergency room again with similar initial presentation, vitals revealed elevated Blood pressure (BP) 177/80 mm hg with rest of vitals stable. Labs showed elevated creatinine of 1.63 mg/dl, Lactic acidosis of 3.2 mmol. At this presentation, she also reported new pulsatile tinnitus and clicking sensation in the right ear. A CT abdomen and pelvis showed celiac dissection with worsening intramural hematoma and stenosis of SMA. CTA head and neck revealed beaded appearance of the distal cervical right internal carotid artery without any tortuosity of the internal carotid arteries. She was managed conservatively with full dose anticoagulation with enoxaparin (1 g/kg twice subcutaneous daily), spiranolactone was switched to carvidelol 12.5 mg twice daily for better blood pressure control. Her symptoms and labs stabilized after conservative managament and she was discharged on oral apixaban with plan for future follow up with for possible mesenteric vasculature intervention. Given female gender, age and involvement of medium sized arteries like carotid vessel in our patient, a diagnosis of multifocal FMD is established.

3. Discussion

Superior mesenteric artery stenosis (MAS) is a common arterial disease, primarily caused by

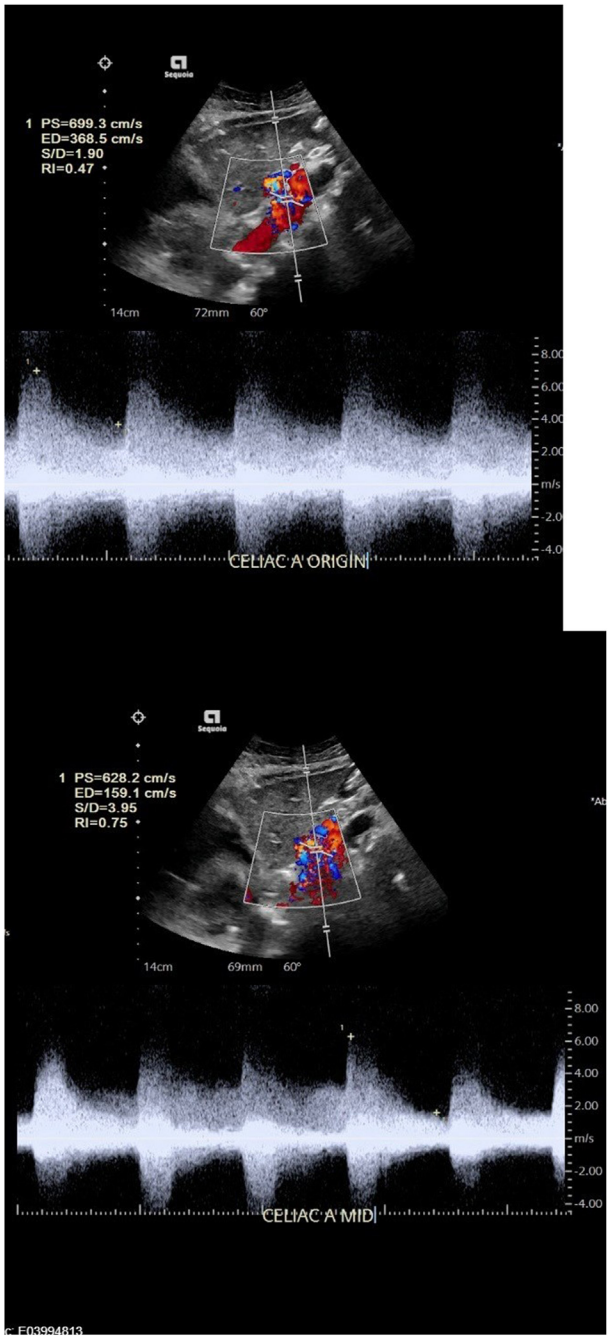

Fig. 2. Image a severe grade stenosis along proximal celiac artery with increased velocity 757 cm/s & image b of the severely stenotic middle celiac artery with elevated velocity of 742 cm/s..

atherosclerosis or thrombosis and tends to increase gradually with age. While many patients are asymptomatic, progressive SMA stenosis can lead to chronic or acute mesenteric ischemia, resulting in various symptoms especially the classic triad of post-prandial epigastric pain or abdominal distention, weight loss, and abdominal vascular murmur (present in about 60% of SMA stenosis patients).

Table 1. Showing velocities in mesenteric vessels from doppler US.

Abdomen	End diastolic velocity (EDV)	Peak Systolic Velocity (PSV)
Proximal celiac artery, stenosis	460 cm/s	768 cm/s
Mid celiac artery, stenosis	278 cm/s	487 cm/s
Distal celiac artery, normal	49 cm/s	91 cm/s
Hepatic artery, normal	93 cm/s	199 cm/s
Splenic artery, normal	43 cm/s	91 cm/s
SMA proximal, normal	40 cm/s	74 cm/s
Mid SMA, stenosis	300 cm/s	605 cm/s
Distal SMA, stenosis	113 cm/s	227 cm/s
Inferior Mesenteric artery, normal	154 cm/s	123 cm/s
Infrarenal abdominal aorta (AA), normal	44 cm/s	8 cm/s
Juxtarenal AA	48 cm/s	11 cm/s
Mid infrarenal AA	53 cm/s	7 cm/s

There is extensive collateral network in the mesenteric circulation which acts as a protective mechanism against most cases of ischemia.[5,6] Hence there is a high incidence of MAS but relatively low occurrence of Chronic Mesenteric Ischemia (CMI) cases. Most individuals do not experience symptoms due to involvement of a single mesenteric vessel. However, review of literature suggests that patients with both celiac artery (CA) and SMA stenoses like our patient are more likely to have CMI.[7]

Mesenteric vascular diseases that result in ischemic changes can be caused by as noninflammatory, non-atherosclerotic arterial conditions like fibromuscular dysplasia and segmental arterial mediolysis (SAM). FMD and SAM share some similarities in radiologic and histologic diagnosis, but they have distinct clinical profiles in terms of age of onset, gender, affected artery distribution, imaging characteristics, symptoms, and treatment. FMD affects middle-aged women, while SAM lacks age or gender predilection. FMD commonly manifests as stenosis and aneurysms in medium-sized arteries like the renal, extracranial carotid, and vertebral arteries, whereas SAM affects larger arteries such as the celiac and mesenteric arteries, posing a higher risk of arterial rupture and hemorrhage.[8] One other differential diagnosis for mesenteric ischemia is Superior mesenteric artery syndrome, which is a rare condition characterized by compression of the duodenum between the abdominal aorta and the superior mesenteric artery. Ct scan is the standard for diagnosis due to its ability to measure the aorto-mesenteric angle (cut off of 22°) which is markedly decreased.[9]

The prevalence of renal FMD is estimated to occur in 0.4% of the population, while craniocervical FMD is much rarer occurring in 0.1% population.[3] Of all the cases reported with mesenteric FMD, only 1.3% reported mesenteric ischemia with 6.8% and 22.3% reporting aneurysm & dissection respectively.[11] FMD mainly affects females below the age of 50 (mean age 47.2 years), with a female predomination.[3,11] There is no evidence suggesting a racial or ethnic predisposition, although Caucasians are more commonly affected than Blacks and 15% population have bilateral involvement. Although the etiology for FMD is unclear, both environmental and genetic factors have been associated with it. Associations with the polymorphisms of the renin-angiotensin gene & Human Leucocyte Antigen- DR isotype (HLA-DRw6) histocompatibility antigen were described in Bowfinger et al. and Pollen Et al. respectively.[3] Spontaneous isolated superior mesenteric artery dissection (ISMAD) is a rare cause of abdominal pain that sometimes leads to intestinal ischemia or fatal SMA aneurysm rupture.[12]

Doppler ultrasound (US) is the primary screening test for FMD. The presence of turbulence, tortuosity, increased velocities, and elevated resistive indices in the middle and lower artery sections on duplex US may indicate FMD.[3] Diagnosis of FMD is confirmed using catheter-based angiography but with the advancements in radiology, FMD changes can be determined on non-invasive imaging techniques such as computed tomography (CT) with high resolution. The classical finding on CT is "the string of beads" appearance but may not be seen in all FMD cases.[3] Histopathological examination shows thinned media and thickened fibromuscular ridges. Conservative therapy with anticoagulation and anti-hypertensive medications is the main line of management. When conservation approach fails, revascularization procedures like percutaneous transluminal angioplasty by interventional radiology are considered. Prognosis is usually favorable, but when cerebrovascular system is involved, there is a risk of cerebral infarction and rupture.[3,11]

Differential Diagnosis of vasculitis should always be considered in patients presenting with abdominal symptoms, and screening for underlying viral infections such as hepatitis B, C & HIV is needed. Polyarteritis nodosa (PAN) which commonly affects SMA can result in intestinal ischemia and infarction. ANCA-associated vasculitis like granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA), involves necrotizing vasculitis in small vessels, with Anti-neutrophilic cytoplasmic antibodies (ANCA) antibodies commonly detected in these diseases. Lupus mesenteric vasculitis (LMV) can occur in up to 10% of systemic lupus erythematosus (SLE) patients and is commonly accompanied by active vasculitis in other organ systems. Treatment of systemic vasculitis mainly involves glucocorticoids and immunosuppressive therapies, with the addition of biological agents depending on the underlying cause and addition of steroids often leads to resolution symptom improvement in patients.[9,10]

Disclaimer

The following manuscript is not submitted to any other journal, has not been presented at any conferences/meetings.

Consent

Appropriate consent was acquired before writing the manuscript.

Declaration of funding source

Nill.

Conflict of interest

There is no conflict of interest.

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