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Case Report

Isolated Symptomatic Celiac Artery Occlusive Disease in a Young Female: A Rare Clinical Presentation. Case Report and Literature Review

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Abstract

Celiac artery (CA) occlusion is a relatively common finding mainly in elderly patient or due to median arcuate ligament syndrome, pancreatitis or malignancy. In the literature, no pathognomonic clinical manifestation has been reported. Most of these occlusions are asymptomatic due to rich collateral circulation from the superior mesenteric artery (SMA). A 49-year-old female patient otherwise healthy, presented with recurrent postprandial epigastric pain and mild weight loss. Initial workup including ultrasound was normal. Gastroscopy revealed the presence of a large deep ulcer, biopsy showed ischemic changes and negative for *H. pylori* and fungal infection. Proton pump inhibitor failed to relief her symptoms. Contrast enhanced abdominal computed tomography scan demonstrated an isolated CA stenosis with absent medial arcuate ligament syndrome. Angiography was performed demonstrating 75% celiac artery stenosis and self-expandable flexible stent was inserted. Patient had an uneventful recovery with no complaint on three years follow up.

Symptomatic CA stenosis is rare medical condition. High index of suspicion and awareness of this entity is essential for earlier diagnosis and better outcome.

Keywords: Celiac Artery (CA); Occlusion; Stenosis; Ischemia; Angiography; Angioplasty

Introduction

Celiac artery (CA) occlusion is a relatively common finding mainly in elderly patient or due to median arcuate ligament syndrome, pancreatitis or malignancy.

Case Presentation

In our manuscript, we report the case of a 49-year-old women, presented to emergency department with repeated bouts of severe colicky postprandial abdominal pain mainly at the epigastric area. Patient noted a recent nausea and occasional nonbilious and nonbloody vomiting. Patient is known to be heavy smoker with irrelevant past medical or surgical history. Upon presentation, she was in pain, thin and pale with tenderness at the epigastric area and no rebound or guarding. X-ray upright detected no air under diaphragm. Laboratory test: White blood count (WBC): 16000, hemoglobin 8.9 g/dl. Amylase, lipase, lactic acid, liver enzymes and blood sugar were all within normal range.

Investigations: Ultrasound was negative. Upper endoscopy showed a large gastric ulcer (Figure 1), biopsy revealed absence of malignancy or *H. pylori* and presence of deep ulceration with evidence of gastric tissue ischemia. Colonoscopy was normal. Patient was initially treated with PPI that failed to relief her symptoms and was readmitted to the hospital with the same clinical picture.

Figure 1

Hence contrast enhanced CT scan abdomen was done and demonstrated CA occlusion small segment at its take off (Figure 2). Median arcuate ligament syndrome was not radiologically noted. Further work up was carried out to exclude connective tissue, autoimmune or hematologic disease and turned out to be negative. Echocardiograph and ECG were unremarkable.

Figure 2

Angiography was performed and confirmed the stenosis that was later managed by deployment of endovascular self-expandable flexible stent (Figure 3).

Patient had an uneventful hospital stay and experienced a prompt improvement of her pain and was discharged in a good condition. Gastroscopy was done after 4 weeks and showed a healed gastric ulcer. No complaint was reported at 38 months follow up.

Figure 3

Epidemiology

In the literature there are few case reports of symptomatic acute celiac artery stenosis, thanks to the rich collateral circulation. It occurs up to 24% for celiac artery occlusion, more common in women (4:1 ratio) and Prevalent among people between 40 - 60 years of age with thinner constitution [1-3].

Despite radiological presence, these occlusions can be asymptomatic due to the collateral circulation supplied by the SMA; gastroduodenal artery, the pancreaticoduodenal arcades and the dorsal pancreatic artery [1]. This picture usually occurs when stenosis develops gradually. However, ischemic symptoms can be significantly manifested in patients with acute insult such as embolism, thrombosis, and tumor compression [3].

Chang Min Park., *et al*. investigated the hemodynamically marked celiac axis occlusion in the asymptomatic Korean population [4]. This prospective study involved 400 patients referred for celiac arteriography between April and July 1999. Twenty-nine patients had celiac axis stenosis (7.3%). 55% of patient with celiac axis stenosis was caused by extrinsic compression by the median arcuate ligament, 10% were caused by atherosclerosis, while in 35% of cases there was no determined cause. Nevertheless, sex, age and atherosclerosis were irrelevant to the incidence to celiac artery stenosis [4].

Age /Decade	Normal CA	Stenotic CA %	Total
< 30	9	0 (0)	9
31 - 40	15	1 (6.3)	16
41 - 50	71	4 (5.3)	75
51 - 60	123	15 (10.9)	138
61 - 70	111	6 (5.1)	117
71 - 80	42	3 (6.7)	45
Total number	371	29 (7.3)	400

Table 1: Incidence of CA stenosis according to age.

Cause of CA stenosis	Gender		Age		Calcified Aortic plaque		
	Male	Female	Young	Old	Present	Absent	Total
Number	319	81	200	200	176	224	400
Compression of MAL	14 (4.4%)	2 (2.5%)	10 (5%)	6 (3%)	6 (3.4%)	10 (4.5%)	16
Atherosclerosis	2 (0.6%)	1 (1.2%)	0	3 (1.5%)	3 (1.7%)	0 (0)	3
Inconclusive	7 (2.2%)	3 (3.7%)	7 (3.5%)	3 (1.5%)	3 (1.7%)	7 (3.1%)	10
Total	23(7.2%)	6 (7.4%)	17 (8.5%)	12 (6%)	12 (6.8%)	17 (7.6%)	29

Table 2: Incidence and etiology of CA stenosis according to age, sex and presence of calcified aortic plaque.

Following a review 1330 CT/MRA reports, a total of 109 patients were identified to radiographically apparent celiac artery compression (CAC). 48 (44%) patients were only symptomatic, majority of them were younger than 30 years old with a history of prior abdominal surgery and had high-grade stenosis [5].

Pathophysiology

Arterial occlusion occurs commonly at the area of vessel branching or bifurcation. It can be acute like thrombosis due to embolus (Atrial fibrillation, hypercoagulable state) or iatrogenic (Post TACE, Whipple's procedure) and progressive due to connective tissue disease or aneurysm [6-8]. Lumen narrowing of at least 50% can reduce the blood volume causing inadequate perfusion to the affected area. At cellular level, ischemia leads to mitochondrial dysfunction, loss of ion transfers regulation and intracellular acidosis. Altered membrane permeability and release of free radicals and degradative enzymes triggers apoptosis and tissues death. Ischemic necrosis develops more frequently in organs supplied by end arteries. Gastric ischemia is less seen due to a rich blood supply unless there is a severe fungal infection or immunity disorder or significantly compromised perfusion [9]. Due to severe CA stenosis, our gastric patient had a fundus ulcer in spite of SMA patency.

Anatomy

Splanchnic blood supply

An extensive collateral circulation that provide adequate perfusion to the intestine. However, prolonged decrease in splanchnic blood flow leads to vasoconstriction in the affected vascular bed, which can eventually reduce collateral blood volume. Intestinal circulation accounts for 10% of cardiac output and can increases up to 25% after eating a meal. It is regulated by the sympathetic nervous system and a variety of systemic (angiotensin II, vasopressin) and local (prostaglandins, leukotrienes) factors [10].

Mesenteric ischemia can be occlusive (arterial embolus, arterial thrombus, venous thrombosis and vasculitis) or nonocclusive (Hypoperfusion conditions). Bowel can usually adapt up to 75% reduction in normal blood flow for 12 hours [11].

Collateral pathways

Most important collateral vessels from the SMA in patients with celiac axis stenosis are the pancreaticoduodenal arcades and the dorsal pancreatic artery.

Figure 4: Blood supply to the pancreas, duodenum, spleen, anterior views.

Pancreaticoduodenal arcades

The pancreaticoduodenal arcades supply the head of the pancreas and the C loop of the duodenum with at least one anterior and one posterior networks [11]. The anterior pancreaticoduodenal arcade is formed by the anterior superior pancreaticoduodenal artery. The two arcades either unite with the SMA via separate inferior pancreaticoduodenal arteries (Figure 5a) or end in a common inferior pancreaticoduodenal artery (Figure 5b).

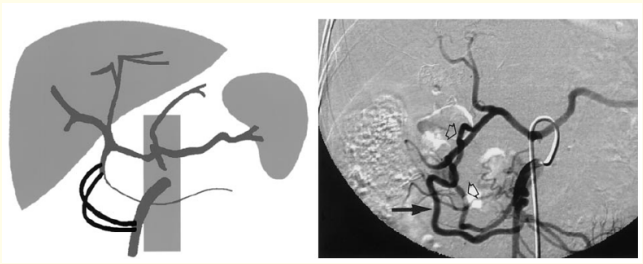

Figure 5a

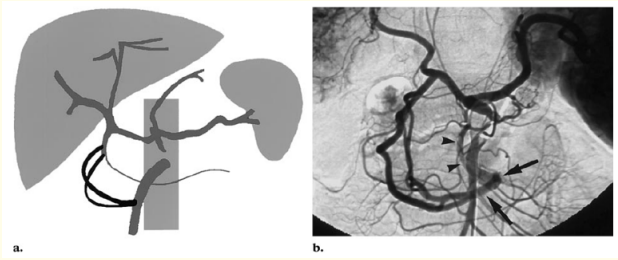

Figure 5b

Dorsal pancreatic artery

Dorsal pancreatic artery of SMA origin has numerous anastomoses and it typically divides into two right branches and one left branch that unites with the pancreaticoduodenal arcade constituting an important longitudinal collateral pathway between the celiac artery and the SMA [11].

SY Song., *et al*. in his retrospective analysis by angiographic findings noted that the most common collateral vessels were the pancreaticoduodenal arcades and the dorsal pancreatic artery [11].

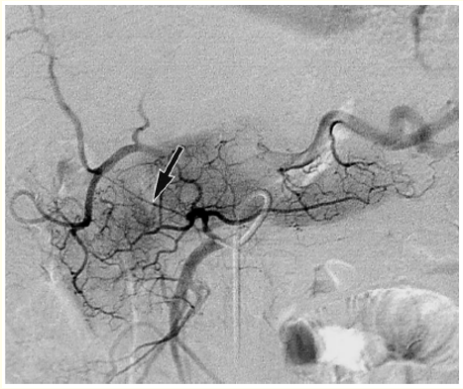

Figure 6: Dorsal pancreatic artery showing a numerous anastomosis between SMA and CA.

Etiology

Acute celiac artery occlusion is a critical medical condition that leads to hypoperfusion and eventually ischemia to many significant organs (liver, stomach, pancreas and spleen). It can be caused by extrinsic by compression from the median arcuate ligament, mass or lymph nodes [12] or intrinsic due to intimal hyperplasia, trauma aneurysm or atherosclerosis (as in our case) and embolic thrombi (atrial fibrillation, coagulation disorders and direct tumor infiltration). Moreover, stenosis can also occur during Whipple's procedure and endovascular management of artery dissection [6,13]. In western countries, arthrosclerosis is more frequent however in Japan the arcuate ligament syndrome is the primary etiology. Studies that utilized Computed Tomography (CT) or angiography revealed a 7% incidence of asymptomatic celiac artery stenosis where by 50% of them are attributable to median arcuate ligament compression [14].

Diagnostic modalities

Doppler ultrasound: Splanchnic blood flow is a complex process influenced by mechanical, metabolic, and neurologic factors as well as numerous vasoactive substances. There is no universal criterion for grading of celiac and mesenteric stenosis. Doppler US is a noninvasive tool with a sensitivity, specificity and accuracy of 100,87 and 89% in 70% stenosis and respectively 100%, 98%, and 99% in greater than 70% stenosis [15]. Normal values for peak systolic velocity are 60 - 180 cm/sec, a rise more than 180 cm/sec in a fasting patients suggests a hemodynamically sufficient stenosis. A velocity higher than 280 cm/sec and/or turbulence indicates a stenosis of more than 75% in the celiac trunk or superior mesenteric artery [16]. Doppler US can be used as a screening method to help detect CA or SMA stenosis or occlusion and helps to avoid the unnecessary angiography with some limitation due to deeply located splanchnic arteries orificies [15].

Figure 7: Transverse view of the CA and its branches, Doppler study [17].

Computed tomography: Contrast enhanced computed tomography (CECT) may accurately depict acute

celiac artery occlusion and its sequelae. Accompanying gastric ischemia is very uncommon, radiologists should, however, be aware of its possibility and imaging findings, and evaluate carefully stomach wall enhancement in patients scanned with suspicion of acute mesenteric ischemia [6]. The role of CT is to identify vascular calcification, hyperattenuating intraluminal clotting, and intramural hemorrhage. Nevertheless, CECT can detect thrombi in the mesenteric arteries and veins, abnormal enhancement of the bowel walls, and the presence of embolism or infarction of other organs [17].

Recently CECT abdomen and CT angiography started to challenge conventional angiography as a diagnostic tool for acute mesenteric ischemia. These modalities are affordable and should be performed early to prevent progression of mesenteric ischemia to significant infarction. Magnetic resonance imaging showed no superior diagnostic value. A CT angiogram can diagnose embolisms but with lower sensitivity than angiography. A study of 26 patients

underwent mesenteric CT angiography demonstrated 100% specificity and only 73% sensitivity in the diagnosis of splanchnic arteries occlusion or embolism [18].

Management

Therapeutic options for celiac artery stenosis depend on the nature, acuity and severity of the occlusion as well as the collateral circulation and the patient's clinical condition. Early intervention involves debridement and resection of nonviable tissue, restoration of blood flow to the ischemic tissue and later supportive care. In the acute thromboembolic event, direct catheter thrombolysis or surgical embolectomy has been the traditional procedure with reasonable short and long term outcome. For chronic occlusive disease two methods are used, surgical (endartectomy or bypass procedure like aorto-mesenteric or celiac bypass using variable grafts) and endovascular (Balloon and stenting) [19,20].

At the mayo clinic, 5 years' graft patency rates have been 90%, 54%, 0% with one vessel, two vessel and three vessel bypasses respectively. Anterograde aortoceliac bypass and transaortic endartectomy have been successful for high risk patient and are usually adequate for multiple outflows with low chance of kinking and thrombosis but it is technically challenging and has been associated with renal ischemia. Retrograde bypass from the infrarenal aorta or iliac artery is technically easier and avoids renal ischemia but has been related to lower inflow and graft kinking [21].

Percutaneous transluminal angioplasty is a minimally invasive modality and avoids the necessity for general anesthesia however, long term results however are less satisfactory and recurrence of stenosis is significantly greater and up to 50% of patient develop restenosis within 1 year. When the cause is an intrinsic one minimally invasive intraoperative angioplasty with or without stenting can be performed [22]. A retrospective study done by Ahanachi., *et al*. published recently in the society of vascular surgery 2013, to compare the outcome of angioplasty and stent in CA and SMA. One hundred twenty-one patients underwent visceral stenting for visceral stenosis. CA stenting demonstrated a significantly poorer patency and hence requiring higher reintervention rate [23].

Conclusion

Isolated symptomatic CA stenosis is rare and a critical medical condition that has no pathognomonic features. A long lasting gastric ulcer in the absence of malignancy and *H. pylori* can be a leading sign. Knowledge of this entity and familiarity with its radiological and clinical presentations is fundamental for accurate diagnosis and proper management.

Conflict of Interest

None.

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