A Rare Cause of Mesenteric Ischemia: Celiac Axis Compression Syndrome

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Abstract

Mesenteric ischemia is an uncommon etiology of abdominal pain. Celiac axis compression syndrome is an extremely rare cause of mesenteric ischemia. The primary pathological mechanism is the external compression of the celiac trunk by median arcuate ligament. The clinical manifestation of celiac axis compression syndrome includes postprandial pain, diarrhea, and body weight loss. The diagnosis of this disease is usually difficult and depends on the angiography findings. For treatment, only percutaneous transluminal angioplasty and surgical intervention have been suggested in reviews in the literature. We, herein, report an unusual case of celiac axis compression syndrome and also review the literature pertaining to this disease.

Key words: abdominal pain, angiography, celiac axis compression syndrome, mesenteric ischemia, percutaneous transluminal angioplasty

Introduction

Abdominal pain is a very common and troublesome problem clinically. The diagnosis of abdominal pain is usually difficult because of many etiologies involving its pathophysiology. It may be caused by a functional problem like gastroenteritis, or organic problems such as malignancy, bowel obstruction and mesenteric ischemia (1). Mesenteric ischemia is a clinical severe disorder due to decreased demand of arterial blood supply of the intestine. The clinical characteristic feature of mesenteric ischemia is abdominal pain, especially postprandial, and it may manifest as a acute or chronic episode. The cause of mesenteric ischemia is classified as either mechanical or non-mechanical obstruction. The pathophysiologic factors of mechanical obstruction may result from intraluminal stenosis or extraluminal compression (2).

Celiac axis compression syndrome (CACS) is an extreme condition of mesenteric ischemia due to external compression of the celiac trunk. Reviewing the English literature on Medline, there were only rare cases reported in the past twenty years. Here, we report a case of CACS in a 34-year-old man presenting with abdominal pain, diarrhea and weight loss.

Case Report

A 34-year-old man presented to our emergency department because of 2-month history of abdominal pain. He was a construction laborer and denied systemic disease in the past. He smoked six cigarettes per day for twenty years but denied regular alcohol consumption.

He began to suffer from abdominal pain 2 months prior to this admission. The pain was located at the epigastric area, starting 30 minutes after meals and lasting for 2 hours. Moreover, the pain was moderately relieved by bending forward. He also mentioned diarrhea three to four times per day and loss of body weight of 4 kilograms in the recent 2 months. No vomiting or fever was noted. On physical examination, his abdomen was soft with mild tenderness at the epigastric area; bowel sound was hyperactive without bruit. The laboratory examinations and abdominal radiograph were unremarkable.

During hospitalization, peptic ulcer disease was considered initially and the esophagogastroduodenoscopy showed...

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Received for publication December 11, 2006; Accepted for publication January 9, 2007

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only reflux esophagitis. Subsequently, the colonoscopy and abdominal ultrasound were performed and showed no particular abnormality. However, his cramping abdominal pain persisted and frequently required narcotic analgesics to control pain. Thus, under the suspicion of ischemic bowel disease, we arranged a digital subtraction angiogram (DSA). The anteroposterior (A-P) view of DSA demonstrated decreased vessel branches of the superior mesenteric artery (SMA) accompanying with apparent dilatation of the gastroduodenal artery. Moreover, a short-segment stricture in the proximal portion of common hepatic artery (CHA) was also observed (Fig. 1). However, the root of celiac trunk was not well visualized in this modality. The pictures of DSA indicated that increased hepatic artery flow resulted in a “steal” of blood from the SMA via the gastroduodenal artery. Mesenteric ischemia caused by the SMA insufficiency resulting from the inadequate blood supply of the CHA was considered at that time. We subsequently performed a multidetector-row computed tomographic (CT) angiography for further investigation. The three-dimensional (3D) reconstruction of angiography demonstrated a short-segment stricture with post-stenotic dilatation at the root of celiac trunk (Fig. 2). The diameter of celiac trunk was 2.7 mm in the segment of stricture and 8 mm in the segment of poststenotic dilatation. Celiac trunk stenosis was diagnosed based on the 3D reconstruction of CT angiography. However, we could not clearly distinguish whether it was external compression or intrinsic stenosis. Thus, we performed repeat DSA and found the same stricture at the root of celiac trunk by lateral aortography (Fig. 3). We tried to perform percutaneous transluminal angioplasty (PTA) at the stricture of celiac trunk, but it could not be dilated. Patient’s abdominal pain only improved a little after PTA. Hence, we confirmed the cause of celiac trunk stenosis resulted from external compression. We also reviewed the coronal image of previous CT angiography and found the median arcuate ligament compressing the root of celiac trunk (Fig. 4). Thus, the diagnosis of CACS was finally established based on the above findings.

According to the studies in the literature review, surgical treatment was suggested. However, the patient refused to accept further surgical intervention and decided on regularly follow-up in the out-patient department with symptomatic treatment. Vessel dilator and pain-killer were administrated after discharge. In six months of follow-up in the out-patient department, the symptoms of post-prandial pain improved gradually and the vessel dilator had tapered in the recent three months.

Discussion

The CACS was first described in the 1960s but was not...
Figure 4. The coronal image of CT revealed external compression of the root of celiac artery by the median arcuate ligament (arrow).

well understood until recently (3). In a Western country, the incidence of CACS was reported to be as high as 24% of asymptomatic patients (4). Female is in the majority (5). At present, it is widely accepted that CACS is a disease of mesenteric ischemia. In most cases, the external compression of celiac axis is by median arcuate ligament or celiac ganglion (6), but a rare case of CACS caused by acute type B aortic dissection was reported (7). Some people consider that it arises from the neurogenic stimulation of celiac ganglion caused by the throbbing of the compressed artery (6). In the present patient, the celiac axis was compressed by the median arcuate ligament.

The median arcuate ligament is a part of diaphragmatic crura around the aortic hiatus. The celiac axis usually originates inferior to the median arcuate ligament. Only in a few people (about 15%), however, the celiac axis may originate posterior to the median arcuate ligament (8). The median arcuate ligament may compromise the blood flow of celiac axis in some of these people and produce symptoms such as in our case.

The characteristic clinical triad of CACS includes postprandial pain, diarrhea, and weight loss. In addition, an abdominal bruit that intensifies with expiration may be heard in the epigastric region (9). Traditionally, DSA with lateral aortography is a golden standard modality in diagnosing of patients with suspected mesenteric ischemia. However, this modality is invasive, expensive and time-consuming. Additionally, some vascular diseases, such as SMA syndrome or CACS, maybe be misdiagnosed when an A-P view is only done in performing the procedure of angiography. Color Doppler sonography is also a choice in diagnosing of CACS. In fact, some criteria had been advanced in previous studies (10-12). However, because of the influences of iatrogenic factors and the high dependence on the performer’s technique, the role of Color Doppler sonography for diagnosis of CACS is still controversial. Recently, a study reported by Scholbach revealed that color duplex sonography might be a powerful tool for CACS screening (13). In the present patient, for example, the CACS was misdiagnosed as the stricture of CHA because of poor visualization of the root of celiac trunk in an A-P view of the initial DSA. However, the second DSA with lateral aortography confirmed the external compression of the proximal celiac trunk. The newly developed multi-detector CT angiography has become a more favorable modality in diagnosing mesenteric ischemia because it is rapid, non-invasive and non-expensive. It is useful in evaluating the presence and degree of stenosis of the celiac trunk and SMA, demonstrating the collateral circulation, and also helpful in excluding other causes of vascular obstruction. It also allows visualization of small vessels and vessel wall abnormalities in the absence of significant stenosis. Vessels with a complex anatomic configuration can easily be visualized with proper postprocessing techniques. However, the limitations of this modality include the lack of dynamic representation of flow abnormalities and difficulty in evaluating heavily calcified vessels (14, 15). Hence, multi-detector row CT angiography with appropriate postprocessing techniques is highly effective for the diagnosis, evaluation, and treatment of suspected CACS.
In the treatment of CACS, only PTA and surgical approaches were suggested in the documents reviewed (9, 16-18). Although in a few reported cases the symptoms were completely relieved after performing PTA; the symptoms of most patients receiving PTA are only relieved for a short time (18, 19). In the present patient, we also performed a PTA to differentiate the true cause of celiac trunk stenosis, and then the external compression was proved. Moreover, the patient’s symptoms improved a little after PTA and returned again several days later. It may be due to the effect of vasodilator injection while performing angiography. Hence, the vasodilator is possibly another choice of treatment for patients without severe symptoms. Thus, PTA plays the role of “diagnosis” much more than of “treatment” (19). In addition, stent placement via angiography is not suggested because the continuously repeated compression of median arcuate ligament from the respiratory cycle will damage the stent (19).

The surgical approaches for the CACS include division of the median arcuate ligament or arterial reconstruction or bypass (20). Sometimes periarterial neurectomy is also performed at the same time to enhance the effect of surgery (21, 22). Division of the median arcuate ligament by conventional laparotomy has been reported in many studies (20, 23). Recently, laparoscopy is considered a novel approach for the management of CACS (7). Division of the ligament alone is usually adequate in some patients, but revascularization of the entrapped artery after its release is indicated when flow studies or assessments of pressure gradients indicate persistent compromise (24).

In the prognosis of CACS, a large study reported the long-term evaluation of CACS patients treated by surgery, 83% of patients were asymptomatic in the first 6 months after decompression, but only 41% of patients remained asymptomatic 3 to 11 years later (17).

In conclusion, CACS is a rare cause of mesenteric ischemia and is difficult to diagnose clinically. But it must be considered in patients presenting postprandial pain without other apparent presence of peptic ulcer disease, biliary tract disease or pancreatic disease. At present, multi-detector-row CT angiography is the preferred modality for diagnosing this disease. Surgical intervention is the only mainstay of treatment.

References
