Celiac artery compression syndrome

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INTRODUCTION — Celiac artery compression syndrome (also referred to as celiac axis syndrome, median arcuate ligament syndrome and Dunbar syndrome) is defined as abdominal pain related to compression of the celiac artery by fibers of the median arcuate ligament. Initially described in the 1960s, it is an uncommon disorder that is characterized by the triad of postprandial abdominal pain, weight loss, and sometimes an abdominal bruit [1]. The etiology is incompletely understood; a report of its occurrence in monozygotic twins suggests a congenital origin [2].

Despite many case reports where the diagnosis appeared to be clear cut, and symptoms were relieved following surgery [3-10], the diagnosis (and existence) of celiac artery compression syndrome has been a source of controversy. In patients with classic chronic mesenteric ischemia, two of the three mesenteric vessels must typically be occluded or severely stenotic for the patient to experience symptoms of abdominal pain because of the extensive collateral network for the bowel. By contrast, in celiac artery compression syndrome, the superior mesenteric artery (SMA) and the inferior mesenteric artery are widely patent thereby, in theory, providing an ample blood supply to the bowel [4,11].

As a result, many have suggested that the symptoms may not be related to blood flow but rather to involvement of the splanchnic nerve plexus, which lies in the same region as the diaphragmatic fibers. Others have demonstrated an association with celiac artery compression and delayed gastric emptying suggesting another possible cause of symptoms [12].

To further complicate matters, celiac artery compression by fibers of the median arcuate ligament has been demonstrated on arteriography in asymptomatic patients [13,14]. Similarly, autopsy studies have shown that the celiac artery is compressed by the median arcuate ligament in up to one-third of persons [15]. Furthermore, surgical correction of the compression does not always relieve symptoms [16].

CLINICAL MANIFESTATIONS — As noted above, celiac artery compression syndrome is characterized by postprandial abdominal pain, weight loss, and sometimes and abdominal bruit. However, a difficulty in interpreting reports of celiac artery compression syndrome is

that not all studies used the same definition for the disorder. Many studies included patients with a range of abdominal complaints and those with a variety of medical (and often psychiatric) problems [8]. Thus, it is not surprising that the results of surgical treatment have also been variable.

In one of the largest series (with 51 patients), the mean age was 47 and 39 patients were women [17]. Predictors of a favorable outcome from surgery included, postprandial abdominal pain, age from 40 to 60 years old, weight loss of greater than 20 pounds, and lack of psychiatric history. Thus, these features might be considered to reflect the true clinical manifestations of the disorder. Similar conclusions have been described by others [8]. An epigastric bruit provided an important clue in many other reports, but is not universally present. The diagnosis has also been described in children [18].

DIAGNOSIS — Diagnosis is challenging and depends upon suspicion based upon the clinical features and judicious testing for other causes of symptoms. Thus, the diagnosis is often one of exclusion. Many patients have undergone evaluation for other causes of abdominal pain including gallbladder disease, peptic ulcer disease, appendicitis, inflammatory bowel disease with several endoscopic and radiologic tests. Some have undergone previous surgery including cholecystectomy, appendectomy, a gynecologic procedure or laparoscopy in an attempt to relieve symptoms [17,19].

Physical examination is usually normal, although it may reveal features of weight loss. The abdomen should be examined for a bruit, although, as noted above, one is not always present.

Duplex ultrasound of the mesenteric vessels with the use of velocity measurements may reveal elevated velocities at the origin of the celiac artery indicative of stenosis [20,21]. MRA or CT angiogram may also document compression of the origin of the celiac axis by fibers of the median arcuate ligament [22,23].

Abdominal and visceral angiography is often helpful. Standard anteroposterior (AP) aortography can identify dilated collateral vessels and evaluate the dynamics of blood flow. There may be retrograde filling of the celiac artery via a dilated gastroduodenal artery from the SMA. Lateral projections clearly define the origins of both the celiac and SMA. During inspiration, the aorta and the celiac artery move in a caudad direction with the abdominal viscera. Conversely, with expiration, the vessels move cephalad thus causing exacerbation of the region of external compression. Thus, selective images of the celiac artery should be obtained in both end-inspiration and end-expiration (picture 1A-B).

There also may be post-stenotic dilatation, which is a common physiologic response to stenosis. Systolic and mean blood pressures can be obtained from the celiac artery and the aorta measuring any gradient that may be present across the celiac origin.

One report described a technique involving selective cannulation of the superior mesenteric artery and injection of a vasodilator drug during angiography [24]. Reproduction of symptoms and loss of collateral filling of the celiac territory were considered to represent a positive test. The study involved only eight patients, four of whom had a positive test. Of these four, three had full resolution of their symptoms following surgical correction of the compression. However, because of the small sample size, additional trials are necessary to confirm the validity of this procedure.

TREATMENT — The treatment of celiac artery compression syndrome is also debated. It is generally agreed that a key to successful outcomes is the careful selection of patients. However, experience to guide the identification of such patients is limited [8,17]. As noted above, one of the largest series found the following predictors of a favorable outcome to surgery [17]:

- Postprandial pain pattern (81 percent cured)
- Age between 40 and 60 (77 percent cured)
- Weight loss of 20 pounds or more (67 percent cured)

Symptoms were less likely to be relieved in those with:

- Atypical pain patterns with periods of remission (43 percent cured)
- Age greater than 60 (40 percent cured)
- A history of a psychiatric disorder or alcohol abuse (40 percent cured)
- Weight loss less than 20 pounds (53 percent cured).

Although these features can provide some guidance, the strength of these predictors is uncertain because the study was small, the authors did not perform multivariate analysis, the definition of "cure" was not uniform, and patients had been treated with different surgical approaches.

Operative treatment involves division of the fibers of the median arcuate ligament and other filamentous structures around the origin of the celiac axis. This frees the celiac artery from the diaphragmatic ligament to improve blood flow and divides the fibers of the splanchnic nerve plexus.

Less commonly, celiac artery decompression is combined with either celiac artery dilatation or reconstruction with primary anastomosis or interposition grafting. More recently, release

of the median arcuate ligament has been achieved laparoscopically, with the aid of laparoscopic ultrasound to confirm full patency of the celiac artery intraoperatively [3,25,26].

Unfortunately, percutaneous approaches are not possible. Dilation of the celiac artery is not feasible since the underlying problem is extrinsic compression (not intrinsic disease such as caused by atherosclerosis). Similarly, stenting of the celiac artery is not possible since it is tethered to and restricted by fibers of the diaphragm, which is in constant motion.

Outcome — As noted above, outcomes following surgery are variable. Although long-term relief of symptoms has been described in several reports, others underscore the need for caution since many patients do not achieve a benefit [17].

The largest single-center study (described above) included 51 patients of whom 44 were available for follow-up an average of 9 years after surgery [17]. Surgery consisted of celiac axis decompression only in 16 patients, celiac decompression and dilation in 17, or celiac decompression and reconstruction by primary reanastomosis or interposition grafting in 18. Eight of 15 patients (53 percent) treated by celiac decompression alone remained asymptomatic compared with 22 of 29 (76 percent) of those treated by celiac decompression plus some form of celiac revascularization. Late follow-up arteriograms (in 18 patients) showed a widely patent celiac artery in 70 percent of asymptomatic patients compared with a stenosed or occluded celiac axis in 75 percent of symptomatic patients suggesting that persistent clinical improvement depends upon an operative technique that ensures celiac axis patency.

SUMMARY AND RECOMMENDATIONS — Celiac artery compression syndrome (also referred to as celiac axis syndrome, median arcuate ligament syndrome and Dunbar syndrome) is defined as abdominal pain related to compression of the celiac artery by fibers of the median arcuate ligament. Diagnosis is challenging and depends upon suspicion based upon clinical features and judicious testing for other causes of symptoms. Thus, the diagnosis is often one of exclusion. Celiac artery compression can be suggested on Duplex ultrasonography. Confirmation can be achieved with CT, MR or traditional angiography, although, as noted above, the presence of compression does not necessarily indicate that it is the cause of symptoms.

It is generally agreed that a key to successful outcomes is the careful selection of patients. However, experience to guide the identification of such patients is limited. Clinical features described above may be useful in identifying potential candidates. Nevertheless, outcomes of surgery are variable.

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