Received: 1 October 2018 | Revised: 16 November 2018 | Accepted: 5 December 2018

DOI: 10.1002/jgf2.232

IMAGES IN CLINICAL MEDICINE

Journal of General and Family Medicine Open Access WILEY

Isolated superior mesenteric artery dissection and median arcuate ligament syndrome

Tomoya Tsuchida MD[1,2] | Atsushi Komiyama MD, PhD[1,2] | Shinji Saruya MD[3] | Takahide Matsuda MD, PhD[1]

[1]Division of General Internal Medicine, Department of Internal Medicine, St. Marianna University School of Medicine, Kawasaki, Kanagawa, Japan

[2]General Medicine Center, Kawasaki Municipal Tama Hospital, Kawasaki, Kanagawa, Japan

[3]Department of Radiology, Kawasaki Municipal Tama Hospital, Kawasaki, Kanagawa, Japan

Correspondence
Tomoya Tsuchida, Department of Internal Medicine, Division of General Internal Medicine, St. Marianna University School of Medicine, Kawasaki, Kanagawa, Japan.
Email: tsuchidatomoya@gmail.com

KEYWORDS: celiac artery, celiac artery compression syndrome, epigastric pain, hooked appearance, median arcuate ligament syndrome, superior mesenteric artery dissection

A 55-year-old woman was seen with severe epigastric pain three hours after acute onset. Her medical history included longtime hypertension and postprandial epigastralgia over the last 5 years. The patient had often developed epigastralgia for about 2 hours after taking in a large quantity of meal. She reported the present pain was much more severe than those she had ever experienced. She denied any loss of her body weight. She had neither diabetes nor dyslipidemia. She had been placed on antihypertensive medication (20 mg olmesartan and 5 mg amlodipine) and proton pump inhibitor (15 mg lansoprazole). She appeared ill, and her blood pressure

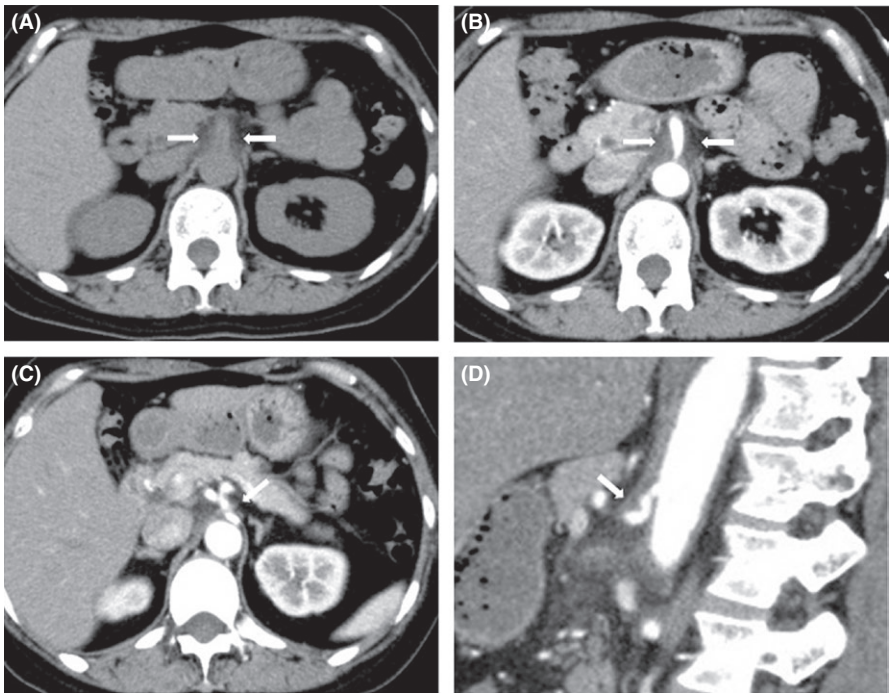

FIGURE 1 Superior mesenteric artery dissection with the thrombosed false lumen is shown by arrows on a plain (A) and a contrast-enhanced (B) CT images. The celiac artery was compressed by the median arcuate ligament as depicted by arrows on an axial view (C) and a sagittal view (D) of contrast-enhanced CT images

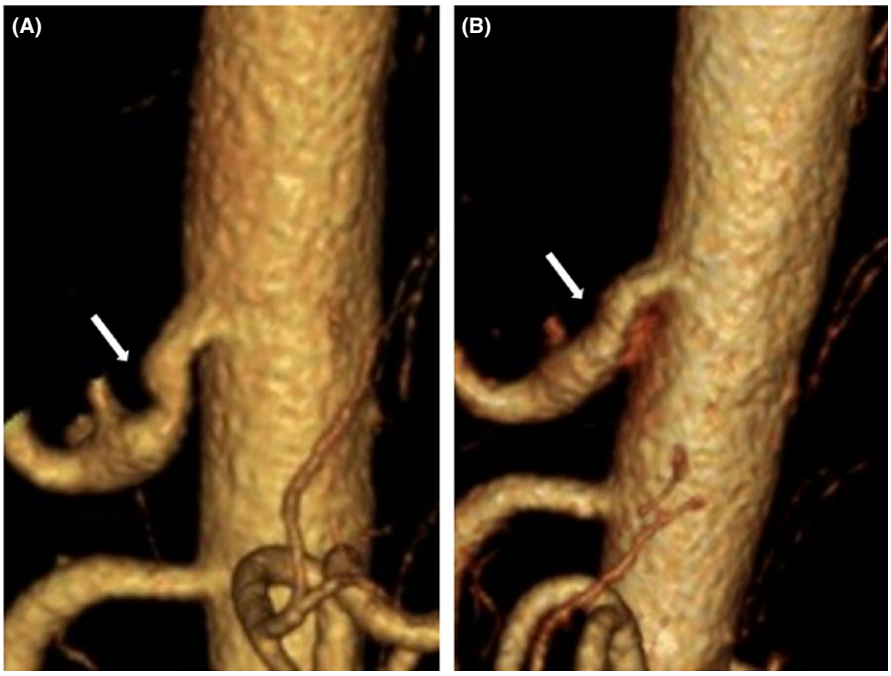

FIGURE 2 Three-dimensional reconstruction of the enhanced CT images (arrows) showing indentation in the celiac axis, "hooked appearance," during expiration (A) and its release during inspiration (B)

was 154/94 mmHg. Her body mass index was calculated as 23 kg/m^2. Abdominal examination revealed tenderness over the epigastric area. There were no other abnormal findings like rebound tenderness, muscular defense, or Carnett sign; any abnormal bruit was not heard over her abdomen. A plain (Figure 1A) and a contrast-enhanced (Figure 1B) abdominal CT scans demonstrated isolated superior mesenteric artery (SMA) dissection with the thrombosed false lumen; stenosis caused by arterial dissection was found about 2 cm from the base of SMA. Signs of bowel ischemia were not present. In addition, contrast-enhanced CT scans depicted celiac axis stenosis by the median arcuate ligament of the diaphragm (Figure 1C,D). Three-dimensional reconstruction of the enhanced CT images explicitly revealed indentation in the celiac axis, "hooked appearance" pathognomonic of median arcuate ligament syndrome (MALS), during expiration (Figure 2A) and its release during inspiration (Figure 2B).[1] The results of the abdominal sonographic examination were consistent with the diagnosis of MALS in that the peak systolic velocity of the celiac artery was faster during expiration (233 cm/s) than during inspiration (175 cm/s). Upper gastrointestinal endoscopy revealed no abnormal findings. Following admission, the patient was successfully treated with fasting and parenteral administration of nitroglycerin with the resultant blood pressure less than 120/60 mmHg. Her abdominal pain subsided by the next day. Subsequently, nitroglycerin treatment could be replaced with oral antihypertensive medication with 40 mg azilsartan, 40 mg nifedipine CR, and 2.5 mg carvedilol. She remains fine over 6 months.

MALS or celiac artery compression syndrome is generally regarded as a rare disease occurring in 0.4% of the population.[2] However, asymptomatic MALS happens to be found incidentally in 2.4%-8% of the people on examination.[2] This syndrome is induced by the overgrowth of the inferiorly placed median arcuate ligament of the diaphragm, leading to celiac artery compression during the expiration phase. Common symptoms include postprandial epigastric pain, nausea/vomiting, and mild weight loss.[2] The following three requirements need to be met to diagnose MALS: (a) occurrence of characteristic symptoms; (b) exclusion of other causes of the symptoms; and (c) confirmation of celiac axis stenosis without atherosclerosis through additional examination.[3] Our case fulfilled all the above criteria.

In MALS, aneurysms are reportedly formed in the pancreaticoduodenal artery region.[4] Celiac axis stenosis represents an attributable etiology for those aneurysms; the increased blood flow through the SMA to the pancreaticoduodenal arcade can lead to the aneurysmal formation.[4] Likewise, in this hypertensive patient possibly with underlying arteriosclerosis, increased blood flow into the SMA as a collateral pathway may have led to the dissection.[4] Because of the absence of surgical indications including ischemic intestinal symptoms, aneurysm rupture, and true luminal narrowing, we employed conservative treatment with success over half a year.

In conclusion, we reported on a case of a 55-year-old woman who was diagnosed as isolated SMA dissection associated with MALS. This combination of illnesses was documented only in two patients.[4,5] Our case underscores the importance of recognizing MALS not only as a cause of postprandial epigastric pain but a rare facilitating factor in the development of isolated SMA dissection.

CONFLICT OF INTEREST

The authors have stated explicitly that there are no conflicts of interest in connection with this article

REFERENCES

1. Horton KM, Talamini MA, Fishman EK. Median arcuate ligament syndrome: evaluation with CT angiography. Radiographics. 2005;25:1177–82.
2. Rongies-Kosmol M, Jakimowicz T. Celiac artery compression syndrome. Mini-review. Acta Angiol. 2015;21:21–4.
3. Scholbach T. Celiac artery compression syndrome in children, adolescents, and young adults. Clinical and color duplex sonographic features in a series of 59 cases. J Ultrasound Med 2006;25:299–305.
4. Takahashi T, Ito T, Endo H, Kudo S, Takei T, Yagi K. A case of isolated superior mesenteric artery dissection accompanied by stenosis at the base of celiac artery relieved by conservative therapy. Nihon Kyukyu Igakukai Zasshi. 2013;24:200–6.
5. Solis M, Ranaval TJ, McFarland DR, Eidt JF. Surgical treatment of superior mesenteric artery dissecting aneurysm and simultaneous artery compression. Ann Vasc Surg 1993;7:457–62.

How to cite this article: Tsuchida T, Komiyama A, Saruya S, Matsuda T. Isolated superior mesenteric artery dissection and median arcuate ligament syndrome. *J Gen Fam Med*. 2019;20:74–76. https://doi.org/10.1002/jgf2.232