CASE REPORT

A patient presenting with stress-induced epigastric pain

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SUMMARY

The median arcuate ligament passes the truncus coeliacus superior to its ostium. If it is thickened and located too low, it can cause external compression and stenosis of the truncus coeliacus, leading to postprandial abdominal pain and vomiting. This combination of symptoms is called median arcuate ligament syndrome. We report the case of a 79-year-old patient who suffered from chronic epigastric pain, which was initially assumed to be caused by either coronary artery disease or atherosclerotic stenosis of the coeliac artery. Angiography excluded coronary artery disease, but showed severe external stenosis of the truncus. The patient underwent laparoscopic release of the median arcuate ligament, which resulted in relief of his symptoms. The median arcuate ligament syndrome should be considered in patients with epigastric stress-induced pain. Further underlying pathologies, especially coronary artery disease, as life-threatening diagnosis have to be initially excluded.

BACKGROUND

Median arcuate ligament syndrome (also called coeliac axis compression syndrome or Dunbar syndrome) is a rare cause of chronic abdominal pain due to external compression of the truncus coeliacus by a fibrous arch that unites the diaphragmatic crura on each side of the aortic hiatus.¹ While 10–24% of the population may suffer from compression of the celiac trunk, it rarely causes patients to develop symptoms. According to earlier studies, the main symptoms are postprandial epigastric pain, vomiting and weight loss.^{2–3} We present a case of a patient with median arcuate ligament syndrome who presented with atypical symptoms. He was admitted to the emergency room because of progredient dyspnoea and chronic epigastric stress-induced pain.

CASE PRESENTATION

A 79-year-old man presented to our clinic with a history of progredient dyspnoea, and chronic thoracic and epigastric pain that was aggravated by physical stress. He was asymptomatic at rest. In his medical history, he had undergone mechanic aortic valve replacement due to an endocarditis lenta in 1985 and had implantation of a two-chamber pacemaker because of atrioventricular block type II; he had also suffered from stable chronic lymphatic leukaemia without current treatment indication. Cardiovascular risk factors were arterial hypertension, diabetes type II and hyperlipoproteinaemia.

Physical examination showed mild abdominal tenderness at the epigastric quadrant; auscultation

of heart and lung revealed discrete basal rales and a systolic murmur.

ECG showed ventricular stimulation by the pacemaker. Electrolytes, liver and pancreas enzymes, C reactive protein, troponin and creatinine were within normal ranges; his leucocytes and international normalised ratio were increased due to the patient's known leukaemia and Warfarin intake. Consequently, an acute myocardial ischaemia was excluded.

Cardiac ultrasound showed normal left and right ventricular function, with the mechanical aortic valve in the right position and functioning regularly. Furthermore, it revealed moderate mitral and tricuspid valve failure and consecutive hypertension of the pulmonary system. X-ray of the chest revealed mild bilateral pleural effusions as well as prominent pulmonary vessels.

At first, an atherosclerotic process with consecutive stenosis of either a coronary artery or the truncus coeliacus was suspected. After medical treatment with diuretics and consecutive weight loss, as well as improvement of the patient's dyspnoea, non-invasive CT angiography of the abdominal vessels was performed. The result did not reveal significant stenosis of the truncus coeliacus. Therefore, we undertook an initial angiography of the coronary arteries, which excluded a coronary artery disease. Next, due to missing pathological findings at

Figure 1 Subtraction angiography of the truncus coeliacus. Injection of contrast agent revealed severe stenosis of the truncus coeliacus.

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coronary angiography and to evaluate a possible vascular cause of the patient's symptoms, an additional angiography of the truncus coeliacus was carried out. An external severe stenosis of the truncus was detected, as shown in [figure 1](#).

After these findings, CT angiography was re-evaluated and reconstructed. A sagittal plain was created, which confirmed the angiographic result.

Doppler sonography of the truncus coeliacus showed flow acceleration, which increased up to 420 cm/s with expiration

([figure 2A](#)). Neither angiography nor CT showed evidence of a relevant atherosclerotic component of the stenosis.

In order to exclude other reasons for epigastric pain, gastroscopy was performed. Apart from mild gastritis, no further correlate for the patient's symptoms was revealed.

After an interdisciplinary discussion regarding the appropriate therapy, a decision was taken for laparoscopic release of the median arcuate ligament. Intraoperative situs showed a fibrotic ligament compressing the truncus coeliacus directly at the

Figure 2 Doppler ultrasound of the truncus coeliacus with (A) remarkable peak flow velocity due to severe stenosis and (B) reduced flow velocity after surgical release.

ostium. The ligament was consequently cut resulting in the truncus coeliacus immediately widening. At postsurgery, the patient showed a slightly prolonged course due to an infected constellation. A cause could not be identified, but following empiric antibiotic therapy, he recovered and was consequently discharged. At follow-up a few weeks later, the patient presented at our outpatient clinic. He reported to be free of his earlier symptoms, even during physical effort. A duplex ultrasound showed reduced blood flow velocity of the truncus coeliacus (figure 2B).

DISCUSSION

Median arcuate ligament syndrome was first described by Harjola.³ Dunbar *et al*⁴ defined it as a syndrome that causes postprandial pain, nausea and vomiting as its most common symptoms. Affected patients are normally young, slim women between the ages of 30 and 50 who have undergone extensive examinations to find the source of their symptoms.⁵ We report an atypical case of median arcuate ligament syndrome. The patient's leading symptoms were epigastric stress-induced pain and, second, progredient dyspnoea, which could be explained by mitral valve leakage, as seen at cardiac ultrasound. The reason for epigastric pain was revealed by CT and angiography, which showed a stenosis of the truncus coeliacus. This was caused by a fibrotic median arcuate ligament that resulted in external compression and led to ischaemia of the stomach during physical effort. Diagnostic findings (CT, Doppler ultrasound, angiography), as well as a high percentage of symptom release after surgical incision of the median arcuate ligament as described in earlier reports supported this hypothesis.^{5 6} Another pathogenetic theory is that epigastric pain is related to neurogenic activation from compression, irritation and intermittent ischaemia of the plexus coeliacus that surrounds the truncus and might also be affected by a low fibrotic median arcuate ligament.⁷

To diagnose median arcuate ligament syndrome, other underlying pathologies should initially be excluded. The gold standard is aortic angiography.² Nowadays, CT and MR angiography have been introduced, which are increasingly replacing conventional angiography.⁵

There have been several approaches to establish an adequate therapy. In our patient, an endovascular intervention to widen the stenosis was discussed. However, due to the assumption that the pathogenesis was an extrinsic compression, a prime endovascular intervention was not deemed effective. In 2007, Delis reported on a case of a young woman who underwent three consecutive attempts to treat stenosis of the truncus coeliacus primarily by balloon angioplasty and stenting. Despite the initial response of this approach, extrinsic pressure resulted in slippage of the stents or fragmenting of their material.⁸ The most common treatment is surgical release of the median arcuate ligament.⁵ Since the introduction of the laparoscopic approach in 2000, a shortened hospital stay as well as less perioperative complications such as blood loss and postoperative pain have been reported.^{9 10} For that reason, our patient was primarily treated with laparoscopic release of the median arcuate ligament

and was free of symptoms at his follow-up examination. However, in approximately 14% of cases, laparoscopic release is not effective.¹⁰ This might be due to transformation of the arterial wall by continuous long-lasting external compression. Therefore, in these cases, a secondary endovascular angioplasty and stenting after surgical release might be helpful.^{10 11}

In conclusion, this case report shows median arcuate ligament syndrome to be a rare cause for thoracic and epigastric stress-induced pain. If other more common diagnoses have been excluded, this syndrome should be considered in patients with these symptoms.

Learning points

- ▶ Median arcuate ligament syndrome is a rare cause for thoracic and epigastric stress-induced pain.
- ▶ The gold standard to diagnose median arcuate ligament syndrome is an aortic angiography, but nowadays this is increasingly being replaced by non-invasive diagnostic techniques such as CT and MR angiography.
- ▶ The most common treatment for this syndrome is surgical release of the median arcuate ligament.

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Competing interests None.

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REFERENCES

- 1 Lainez RA, Richardson WS. Median arcuate ligament syndrome: a case report. *Ochsner J* 2013;13:561–4.
- 2 Duffy AJ, Panait L, Eisenberg D, *et al*. Management of median arcuate ligament syndrome: a new paradigm. *Ann Vasc Surg* 2009;23:778–84.
- 3 Harjola PT. A rare obstruction of the coeliac artery. Report of a case. *Ann Chir Gynaecol Fenn* 1963;52:547–50.
- 4 Dunbar JD, Molnar W, Beman FF, *et al*. Compression of the celiac trunk and abdominal angina. *Am J Roentgenol Radium Ther Nucl Med* 1965;95:731–44.
- 5 A-Cienfuegos J, Rotellar F, Valenti V, *et al*. The celiac axis compression syndrome (CACCS): critical review in the laparoscopic era. *Rev Esp Enferm Dig* 2010;102:193–201.
- 6 Duncan AA. Median arcuate ligament syndrome. *Curr Treat Options Cardiovasc Med* 2008;10:112–16.
- 7 Skeik N, Cooper LT, Duncan AA, *et al*. Median arcuate ligament syndrome: a nonvascular, vascular diagnosis. *Vasc Endovascular Surg* 2011;45:433–7.
- 8 Delis KT, Gloviczki P, Altuwaijri M, *et al*. Median arcuate ligament syndrome: open celiac artery reconstruction and ligament division after endovascular failure. *J Vasc Surg* 2007;46:799–802.
- 9 Jaik NP, Stawicki SP, Weger NS, *et al*. Celiac artery compression syndrome: successful utilization of robotic-assisted laparoscopic approach. *J Gastrointest Liver Dis* 2007;16:93–6.
- 10 Jimenez JC, Harlander-Locke M, Dutson EP. Open and laparoscopic treatment of median arcuate ligament syndrome. *J Vasc Surg* 2012;56:869–73.
- 11 Cina CS, Safar H. Successful treatment of recurrent celiac axis compression syndrome. A case report. *Panminerva Med* 2002;44:69–72.

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