Videolaparoscopic management of arcuate ligament syndrome in pediatric patients

C. Patiño González, M.E. Balacco, S. Contreras, M.F. Toledo, S. Boetsch, H. Arrieta, G. Crespo, E. Romero Manteola

Allende Sanatorium. Córdoba (Argentina).

Abstract

Median arcuate ligament syndrome, which is characterized by postprandial pain, occurs as a result of the compression of the celiac trunk by the ligament. It is a rare pathology in pediatric patients.

We present the case of a 14-year-old girl with recurrent abdominal pain. Ultrasound examination showed an increase in celiac trunk flow rate with flow reversal, while CT angiography demonstrated compression. It was surgically managed by dividing the arcuate ligament through videolaparoscopy. Symptoms disappeared right after surgery and did not reappear in the 24-month follow-up.

The arcuate ligament is a fibrous band located at the level of the diaphragmatic crus. The fact that the celiac trunk originates at the supradiaphragmatic aorta makes the ligament exert compression during expiration, with transitory distal ischemia. Diagnosis is achieved through Doppler ultrasonography of the celiac trunk or CT angiography, among others. Surgical management involves dividing the arcuate ligament.

This syndrome should be considered in the presence of recurrent abdominal pain. The laparoscopic route is the treatment approach suggested.

Key Words: Abdominal pain; Celiac artery; Median arcuate ligament syndrome; Pediatrics; Laparoscopy.

Resolución por videolaparoscopia del síndrome de ligamento arcuato en pediatría

Resumen

El síndrome de ligamento arcuato medio caracterizado por dolor posprandial se debe a la compresión del tronco celíaco por dicho ligamento. En pediatría su presentación es infrecuente.

Niña de 14 años con dolor abdominal recurrente. Se diagnosticó por ecografía un aumento de la velocidad del flujo del tronco celíaco con inversión de flujo. La angiotomografía evidenció la compresión. Su resolución fue quirúrgica mediante la sección del ligamento arcuato por videolaparoscopia. Los síntomas desaparecieron inmediatamente luego de la cirugía y no recurrieron en 24 meses de seguimiento.

El ligamento arcuato es una banda fibrosa en la crura diafragmática. El nacimiento del tronco celíaco en la aorta supradiafragmática conlleva que este ligamento comprima durante la espiración con isquemia distal transitoria. El diagnóstico se realiza con ecografía Doppler del tronco celíaco o angiotomografía, entre otros. La resolución quirúrgica consiste en la sección del ligamento arcuato.

Este síndrome debe tenerse en cuenta ante un caso de dolor abdominal recurrente. La vía laparoscópica es sugerida para el tratamiento.

Palabras Clave: Dolor abdominal; Arteria celíaca; Síndrome de ligamento arcuato medio; Pediatría; Laparoscopia.

INTRODUCTION

Median arcuate ligament syndrome (MALS) is characterized by postprandial pain, nausea, vomit, and weight loss. It is etiologically described as a high origin of the celiac trunk at the aorta, superior to or at the level of the arcuate ligament, and also as an anomaly of ligament fibers. This causes transitory ischemia at the level of the artery as a result of compression during diaphragmatic excursion. It is a rare pathology in pediatric patients[1-5].

CLINICAL CASE

14-year-old female patient with postprandial epigastric recurrent abdominal pain (RAP). She had no pathological history and said she had been suffering from unspecific pain for two or three months. Physical examination demonstrated an epigastric murmur which varied with respiration. Ultrasound examination revealed an increase in full expiration flow rate at the level of the celiac trunk (CT), with rates close to 448 cm/s (mean normal flow rate in the CT:

Corresponding author: Dra. Celeste Patiño González. Pedro Simón Laplace 5749. Villa Belgrano. CP: 5000. Córdoba. Argentina.
E-mail address: celestecarolina@hotmail.com

The presentation of this case won the first prize in the module "Tricks in surgery and novel procedures" at the 52nd Argentine Congress of Pediatric Surgery (ACACIP) in October 2018.

Date of submission: July 2019 *Date of acceptance:* August 2020

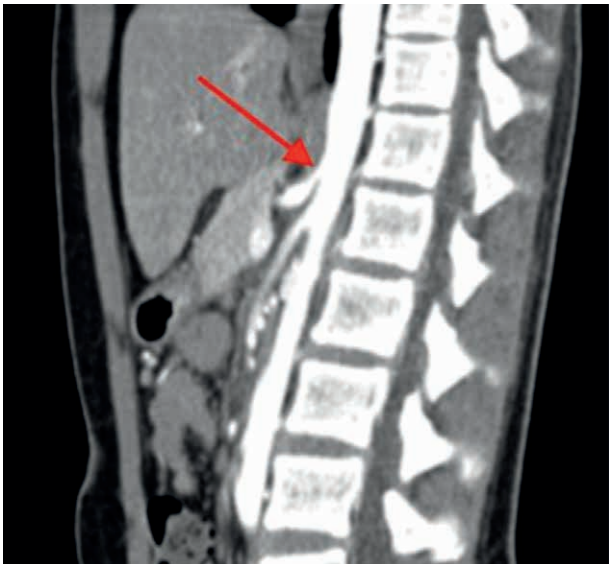

Figure 1. CT angiography during inspiration, sagittal slice.

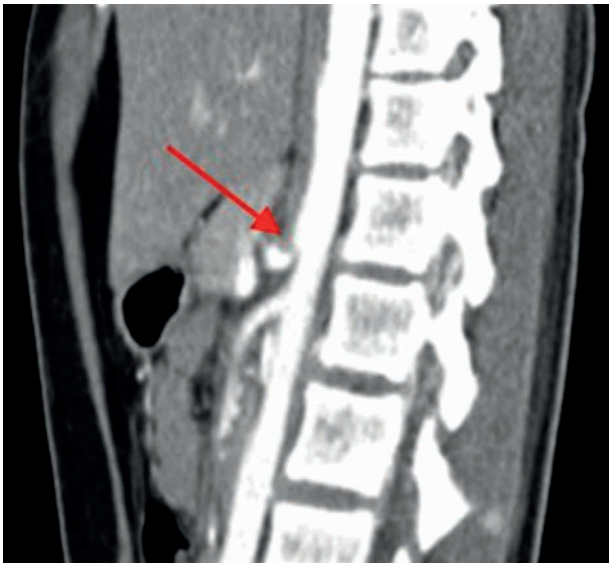

Figure 2. CT angiography during expiration, sagittal slice (red arrow: celiac trunk).

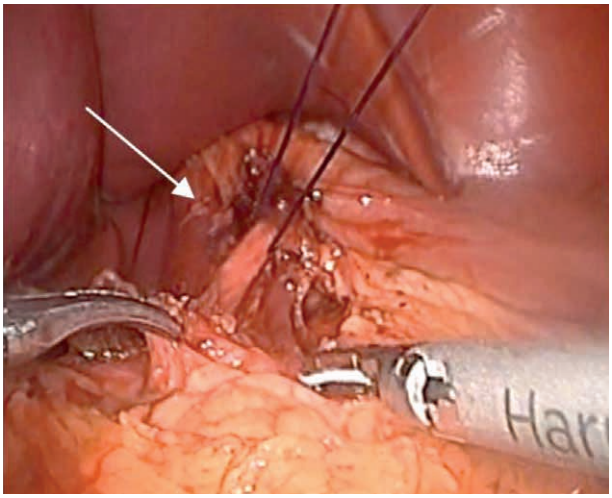

Figure 3. Traction of the left gastric artery (white arrow).

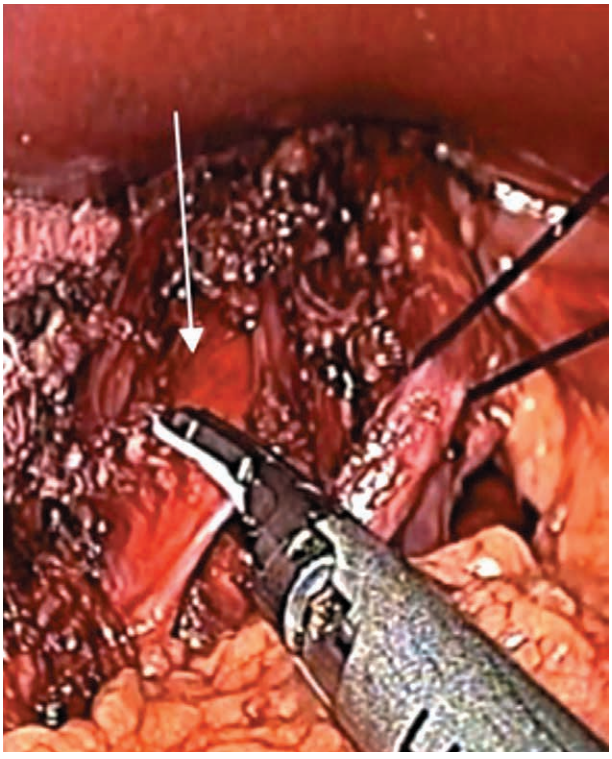

Figure 4. Median arcuate ligament division using an ultrasonic scalpel (white arrow).

103 ± 18 cm/s) and flow reversal at the level of the hepatic and splenic artery[6]. A CAT-scan with endovenous contrast was carried out. In the slices achieved during inspiration, the CT had a 3 mm diameter in the sagittal plane (Fig. 1), whereas in the slices achieved during expiration, it had a 1.3 mm filiform aspect (Fig. 2). Once CT compression by the ligament had been confirmed, surgical planning was defined. Videolaparoscopy was decided upon for CT freeing purposes. An umbilical port was used to introduce the 10 mm 30-degree scope, and three further 5 mm ports were placed: one on the right flank, one on the left flank, and one at the epigastrium. Diaphragmatic pillars were identified through the gastrohepatic ligament. Traction was exerted on the left gastric artery (Fig. 3), and the confluence with the splenic artery was dissected to facilitate CT identification up to its origin at the aorta. At this level, the arcuate ligament, which was compressing the anterior side of the CT, was visualized. The ligament was divided using an ultrasound harmonic scalpel (Fig. 4). Total operating

time was 80 minutes. The patient had a good postoperative evolution. At the control visit on day 30, the patient had no symptom recurrence. CT flow rate was still altered at ultrasound examination –350 cm/s–, without repercussion at the level of the hepatic and splenic artery flow. 24 months following surgery, the patient has no symptoms, and flow is normal at ultrasound examination.

DISCUSSION

The arcuate ligament is a fibrous band located at the level of the diaphragmatic crus. In most human beings, the CT originates at the level of the abdominal aorta, underneath the ligament. However, in 33% of cases, it does originate above it, which predisposes to significant compression during expiration[1,3,7]. Etiologically speaking, this syndrome may occur as a result of the CT originating at a higher place, or because the crus has a more caudal position. The connective tissue also becomes fibrous owing to chronic compression, which favors gradual CT stenosis[3,8]. This is associated with ischemia as a result of deteriorated flow or vascular steal at the level of splanchnic circulation. Compression also damages the celiac lymph node and the celiac plexus, which may account for symptom persistence and in some cases chronic diarrhea[8,9].

MALS is also known as Dunbar syndrome[10,11]. Dunbar was the first to report a series of cases demonstrating CT compression in patients with abdominal symptoms through aortography[11]. It is characterized by epigastric RAP with postprandial frequency and associated with nausea and vomit. Weight loss and occasionally diarrhea may also occur[3,8]. Physical examination is typically normal, and an epigastric murmur varying with respiration may be noted. It tends to be more prevalent in women aged 20-40 years old[1]. Our patient had all typical symptoms, including murmur.

RAP was defined as abdominal pain lasting for more than two months. Differential diagnoses of children with RAP are multiple and have been widely described in the literature. Anatomical causes include intestinal malrotation, biliary lithiasis, duodenal membrane, and inflammatory or infectious factors causing epigastric abdominal pain, such as gastritis, gastric ulcer, duodenal ulcer, or *Helicobacter pylori* infection, among others[12]. Fortunately, in our case, selecting a complementary method allowed us to rapidly achieve diagnosis.

Complementary methods include Doppler ultrasonography of the celiac trunk, angiography, magnetic resonance angiography, and CT angiography with endovenous contrast. Lateral aortic arteriography with pressure gradient measurement at the level of the artery is the gold standard diagnostic technique. However, combining Doppler ultrasonography and CT angiography with inspiration and expiration is less invasive and allows for a relatively accurate diagnosis[4]. Doppler ultrasonography should be carried out with deep expiration and deep inspiration, and flow rates at the level of the celiac artery should be measured. Significant stenosis is suspected when rates exceed 200 cm/s[3]. Our patient had ultrasound alteration, with rates exceeding 440 cm/s at the level of the celiac artery, as well as flow reversal at the level of the hepatic and splenic artery. Sagittal slices from the CT angiography demonstrated focal narrowing of the CT and subsequent dilation. The differences between images during deep inspiration and deep expiration are characteristic[13]. This study was highly valuable for us, since it allowed us to visualize the compression exerted by the arcuate ligament on the artery.

Angiography is a good diagnostic method and allows for endovascular treatments, but it has poor results as a single therapy given that this syndrome is caused by the extrinsic compression of the diaphragmatic fibers[14].

Other diagnostic methods described include magnetic resonance angiography and gastric tonometry, which are little used in children[4,14,15].

Treatment objective lies in achieving decompression by dividing the ligament, and in some cases, it can also be accompanied by vascular bypass. Surgical approach can be either open or videolaparoscopic[16,17]. Videolaparoscopy allows for a better visualization of the celiac plexus, a quick recovery, and a short hospital stay[3,16,17]. Even though conversion rate has been reported to be less than 15%, potential vessel wall damage should be considered. Dissection should be carefully performed, especially when surrounding the celiac trunk in full to free it from fibrous bands accessory to the arcuate ligament[1,18]. In our case, the laparoscopic approach allowed for an excellent visualization. Symptom improvement has been described in 90% of cases in the first postoperative year. Lack of surgical treatment response could be explained by the fact chronic extrinsic compression alters the vessel wall, with intimal hyperplasia, proliferation of elastic fibers, and de-organization of the adventitia. In these cases, endovascular management with angioplasty with or without stent, and open vascular reconstruction with aortic-celiac bypass are suggested[19,20]. The pediatric cases reported did not require vascular bypass. This could be explained by the fact action is taken before the vessel wall is obstructed[2,4,21]. Long-term follow-up should include Doppler ultrasonography. Our patient has no symptoms 24 months following surgery, and CT is normal at Doppler ultrasonography.

CONCLUSION

Given how little prevalent it is in pediatric patients, MALS should be considered as a differential diagnosis before a case of RAP. Laparoscopy is the technique suggested for treatment, since it allows for an adequate visualization of the anatomical structures and quick patient recovery.

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