Median Arcuate Ligament Syndrome in a Pediatric Patient: A Rare Case Report

Nandini K Bedi, H S Bedi, Alka G Grewal

1Associate Professor, Department of Pediatric Surgery, Christian Medical College and Hospital, Ludhiana, Punjab, India, 2Associate Professor and Head, Department of Cardio-Thoracic and Vascular Surgery, Christian Medical College and Hospital, Ludhiana, Punjab, India, 3Assistant Professor, Department of Pediatric Surgery, Christian Medical College and Hospital, Ludhiana, Punjab, India

INTRODUCTION

Median arcuate ligament syndrome (MALS) was first reported by Harjola in 1963, and was first described with an abdominal angiography by Dunbar in 1965, who is credited by naming the condition Dunbar syndrome. It is a rarely reported condition caused by compression of the celiac artery and possibly also the celiac ganglion by the MAL. It is a condition known by various names including celiac artery compression syndrome. The MAL is the fibrous arch across the two crura of the diaphragm anterior to the aorta as it enters the abdomen. In an estimated 10-24% of individuals, the MAL extends to cover the origin of the celiac trunk but only 2-3% of these are symptomatic. The classic triad of symptoms is post-prandial abdominal pain, nausea/vomiting and weight loss; often associated with an abdominal bruit in mid-epigastrium. It is seen more often in young female adults and older males. There are no reported estimates of incidence in pediatric patients. Division of the MAL with or without excision of the celiac ganglion is the treatment of choice.

CASE REPORT

A 15-year-old girl presented to us with a 10 months history of post-prandial, upper abdominal pain, vomiting, off and on after meals, and a documented weight loss of 9 kg over the last 3 months. Her dietary intake was significantly reduced and she had anemia (hemoglobin 9 g/dl). She had undergone an ultrasonography of the abdomen, upper and lower gastrointestinal endoscopy, and barring superficial gastric erosions; the tests were normal. She then underwent abdominal computed tomography (CT) angiography with a multi-slice CT scanner having three-dimensional (3D) reconstruction imaging. This reported a significant stenosis of the celiac artery with a post-stenotic dilatation, best seen in a lateral view of a 3D reconstruction image (Figure 1). We decided to operate and release the compression on the celiac trunk.

An upper midline incision, extending from xiphoid to just below umbilicus was used with the point in mind that if the celiac artery has to be grafted the incision can be extended into a left thoracoabdominal incision, into the left 7th or 8th intercostal space. The upper abdomen was raised on a roll sheet and a self-retaining retractor was used to lift up the ribs superiorly. The hepatogastric ligament was incised to approach the lesser sac and the esophagus was lifted in a sling and pulled aside to visualize the aortic hiatus. While doing the dissection, a bruit could be palpated over the pancreas in the midline. There was dense fibrous tissue between the crura extending like a sheet, right up to the...
origin of the celiac artery which took place just above the upper margin of the pancreas. The pancreas was reflected downward (Figure 2) and the fibrous sheet dissected and divided with blunt and sharp dissection until the celiac artery was seen arising from the aorta. By the end of the dissection, the bruit had disappeared.

Recovery was smooth and when the child started to feed there was an amazing reversal of pre-operative symptoms. In 6 months time, she had recovered her weight, and at 1 year follow-up she came happy but complaining of weight gain and had started dieting. Currently, at 2 years follow-up she is well and symptom-free.

**DISCUSSION**

MALS is a rare condition and more so in the pediatric patient. It is thought that as the organs supplied by the celiac artery have an excellent collateral supply from the pancreaticoduodenal branches of superior mesenteric artery (SMA) and that the symptom of pain is due to compression of the celiac ganglion which is also compressed by the MAL and not ischemia. Hence, some advocate removal of the celiac ganglion, while some do not comment on it. We did not encounter the celiac ganglia during dissection, but nerve fibers in the path of dissection were sacrificed. We did not hunt for the celiac ganglia which would have been a challenging task at that point. After the compression over the celiac artery was released, the bruit vanished. If compression of the celiac ganglia causes pain then release of the MAL should relieve it. It should not be necessary to excise it. A fine network of nerves was also excised with the MAL very likely coming from the celiac ganglia.

If ischemia is not the cause of pain because of a good collateral blood supply from the SMA, then probably the majority of people who have compression of the celiac artery by MAL, and are asymptomatic, have a good collateral blood supply and hence do not suffer ischemia after meals. It also stands to reason that the symptomatic patients probably do not have good collaterals.

We approached with an upper midline incision through the hepatogastric ligament, keeping in mind that if the exposure is not sufficient, or there are complications like, a very short celiac artery which does not straighten out after release of compression, injury to the celiac artery requiring a graft, injury to the aorta requiring repair or other intervention then we would have to extend the incision into a left thoracoabdominal extension toward the left 7th or 8th intercostal space. The spleen, tail of the pancreas, and splenic flexure of colon would have to be lifted off the posterior abdominal wall to visualize the entire upper abdominal aorta to clamp above and below for the repair.

Endovascular dilatation and stenting have not proved to be useful as the problem is not addressed and symptoms tend to recur. The laparoscopic approach is reported to be successful and is being practiced by many via abdominal approach as well as retroperitoneal approach. The recurrence of symptoms is reported requiring a long term follow-up.

**CONCLUSION**

MALS is seen most often in the young adult female, but, though rare, is also reported in the pediatric age group. It is important to know that surgical release of the MAL, whether open or laparoscopic, gives lasting results. Before undertaking the procedure the pediatric surgeon should be aware that if a vascular graft is required he/she should be ready to expose the aorta as described above. Endovascular...
procedures have not shown lasting results. Long term follow up is required as symptoms may recur.

REFERENCES


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