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Case Report

Multiple Abdominal Vascular Compression Syndromes in A Patient with Hypermobile Ehlers-Danlos Syndrome

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Abstract

We present a case of a 24-year-old female with hypermobile Ehlers-Danlos syndrome who presented with a diagnosis of symptomatic median arcuate ligament syndrome, superior mesenteric artery syndrome, and May-Thurner syndrome, along with asymptomatic nutcracker phenomenon. Based on her presentation, she elected to undergo surgical correction by decompression of all symptomatic visceral and vascular compression syndromes. She reports improved quality of life, though she still suffers from impaired small bowel motility, which could be due to delayed correction of the vascular compressions or other comorbidities of Ehlers-Danlos syndrome.

Keywords: Hypermobile ehlers-danlos syndrome; Abdominal vascular compression; Duodenojejunostomy; Superior mesenteric artery syndrome; Median arcuate ligament syndrome

Introduction

Hypermobile Ehlers-Danlos syndrome (hEDS) has autosomal dominant inheritance and variable expressivity with no current identified gene [1]. Abdominal Visceral and Pelvic Vein Compression Syndromes (AVPCS)-such as Median Arcuate Ligament Syndrome (MALS), Superior Mesenteric Artery Syndrome (SMAS), May-Thurner Syndrome (MTS), and nutcracker syndrome-cause a variety of symptoms, including abdominal pain, nausea, vomiting, pelvic pain and rarely hematuria, depending on the location and degree of the compressed structure [2-5]. These syndromes are frequently overlooked and, thus, seem to be rare. However, recently there have been an increasing number of reports of patients developing more than one vascular compression-a typical constellation of the lordogenetic midline syndrome [6,7] often in conjunction with hEDS [2-5].

Herein, we present a case of a 24-year-old female with hEDS who underwent surgical correction of MALS, SMAS, and MTS after years of severe gastrointestinal symptoms, which led to reliance on jejunal feeds and intravenous hydration. The patient provided written informed consent for the report of her case and imaging.

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Case Presentation

A 24-year-old female with a history of hEDS and dysautonomia presented with a one-month diagnosis of SMAS, MALS, and MTS, discovered during a workup for postprandial nausea, vomiting, abdominal distention, and chronic diarrhea. Gastrointestinal symptoms began at the age of 18, with abdominal distention, early satiety, and epigastric pain. Initial workup included a gastric emptying scan, gallbladder ultrasound, and upper and lower endoscopy with biopsy, which were all normal. Color doppler ultrasound of the celiac artery demonstrated elevated peak systolic velocities compared to the aortic ones (97 cm/s) in the respiratory position of the diaphragm 341 cm/s, celiac in deep inspiration 230 cm/s, and celiac extensive expiration 375 cm/s. These findings, in combination with typical clinical symptoms, were consistent with MALS and consistent across multiple measurements. Additionally, findings were supported with an abdominal CT with contrast and improvement of early satiety and pain after a celiac plexus block but the improvement remained for no longer than 48 hours. Breath testing indicated Small-Intestinal Bacterial Overgrowth (SIBO) and fructose malabsorption. Treatment with Rifaximin for SIBO and a low fructose diet did not improve symptoms. She attempted to manage symptoms with small, frequent meals.

By 22 years of age, she lost 20% of her body weight, with a body mass index of 16.6 kg/m^2, due to inadequate oral intake. A port-a-cath was placed for daily intravenous fluids, due to worsening orthostatic intolerance, followed by a gastrojejunostomy feeding tube for nutritional support. Following that tube placement, she developed gastric perforation and peritonitis, which required surgical repair. It was explained that SMAS and/or hEDS could have played a role in the development of the gastric perforation but no further workup was performed since it was felt that weight gain would alleviate the SMAS, if present. Despite full nutritional support and a 15-pound weight gain, her symptoms continued to progress to include bilious vomiting and intractable nausea.

At age 24, further testing was performed. Color doppler ultrasound indicated elevated systolic celiac artery velocities, consistent with previous studies, and respiratory changes of the angle of take-off and systolic flow velocities as well. (Figure 1) Functional duodenal sonography, pre- and post-meal, demonstrated duodenal compression, along with dilation and reverse peristalsis of the descending duodenum, confirming a diagnosis of SMAS. Additional doppler findings included MTS, which could account for her left leg heaviness and pain, severe Left Renal Vein (LRV) compression and severe pelvic congestion-both of which were clinically inapparent, and ptosis of the right kidney without hemodynamic consequence. Abdominal and pelvic CT with intravenous contrast was performed, which showed an increased lumbar lordosis lifting the aorta against the superior mesenteric artery, thus, narrowing the passage of the duodenum, with an angle of 18 degrees, from which the SMA was arising from the aorta, along with compression of the origin of the celiac trunk, further corroborating the diagnosis of SMAS and MALS. (Figure 2A) The additional finding of asymptomatic LRV compression noted on color doppler ultrasound was again noted on the CT (Figure 2B).

Due to failure of conservative treatment to address the SMAS and reduced oral intake being the primary clinical problem, decompression of both the duodenum and celiac trunk was recommended, since both likely contributed to the abdominal pain, and it was felt that disturbed duodenal peristalsis, due to the SMAS, was unlikely to resolve without surgery. The patient decided to address the MTS simultaneously to help with left leg pain and heaviness. The LRV compression was asymptomatic, though the proposed surgical technique described below addressed the SMAS and LRV compression simultaneously.

Surgery included (from head to toe) MAL release, resecting the MAL by 1 cm, not just dividing the ligament, to avoid restructuration by a scar; placement of a 16 mm, ring-enforced Polytetrafluoroethylene (PTFE) graft around the LRV, which enlarged the aortomesenteric angle, alleviating the duodenal and LRV compression, (Figure 3A)-a technique first described Barnes et al. for treatment of the so-called "nutcracker syndrome;"[8] and elimination of the MTS (Figure 3B, 3C) by placement of a 20 mm, ring-enforced PTFE graft around the left common iliac vein.

The grafts were fixed by two PTFE sutures at each ring on both sites to the midline. Between the sutures, the graft was bisected along the long axis and then wrapped around each vein, LRV and Left Iliac Vein (LIV). Next, the graft was reconstructed by a 4 × 0000 PTFE overlock running suture technique. The LRV graft was then fixed with single-stay sutures to the vein itself at 3, 6, 9, and 12 o'clock, 360 degrees around, along with the retroperitoneum before and after crossing the aorta and, finally, to the SMA. The LIV graft was fixed by single stay sutures at 3, 6, 9, and 12 o'clock to the LIV, the retroperitoneum, before and after the course of the right common iliac artery to the fascia of the retroperitoneum, and to the adventitia of the right common iliac artery, which was positioned on the "roof of the graft."

The patient was discharged on postoperative day 13 with no major complications on a combination of oral and jejunal feeds. Abdominal and pelvic CT with contrast post-operation showed decompression of the celiac artery, normalization of the duodenal passage between the aorta and SMA with an angle of 35 degrees, and presence of the PTFE graft around the LRV and LIV (Figure 2C, 2D).

At two and a half years follow up, she continues to experience postprandial nausea and occasional vomiting and has been diagnosed with small bowel dysmotility. However, she is no longer on intravenous fluids or jejunal feeding, using gastric-tube feeding as a supplement to oral intake as her oral tolerance continues to improve. She reports her quality of life and functionality have increased exponentially since surgery.

Discussion

Esophagogastroduodenoscopy (EGD) are frequently used to visualize the gastrointestinal system and take biopsies in patients with upper gastrointestinal symptoms. In patients with SMAS, EGD does not always show external compression in the third portion of the duodenum since SMAS is a dynamic stenosis of the duodenum, and when air is blown into the duodenum, the findings may be negative. Unfortunately, this often leads to patients being labeled with symptoms that are functional rather than structural in nature, delaying proper diagnosis and treatment. This highlights the importance of using additional imaging modalities in patients with symptoms of SMAS, such as ultrasonography, CT abdomen and pelvis, and/or upper gastrointestinal series with barium contrast.

Duodenojejunostomy is the frequently favored approach to surgical management of SMAS, as it is a technic to bypass the clamp between the aorta and SMA [9]. Although it has been shown to have long-term benefits, poor outcomes are seen in individuals with motility disorders [9]. The Strong procedure, by which the duodenal C-loop is moved out of the aorto-mesenteric clamp, is another surgical approach to SMAS, though outcomes of duodenojejunostomy was

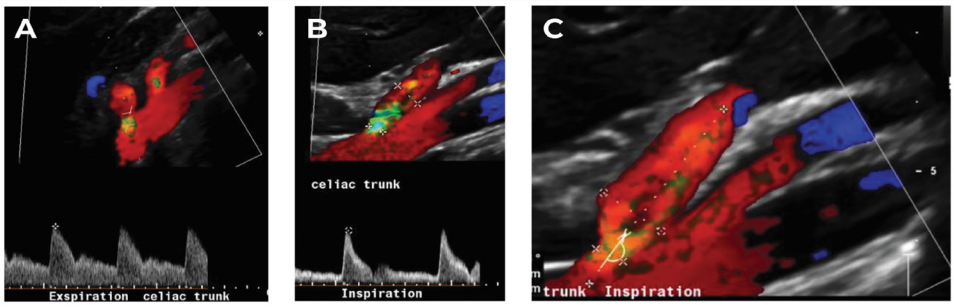

Figure 1: Ultrasound of celiac artery. (A) Expiratory phase US celiac artery-angulation at the celiac trunk. Turbulence of flow is visible with elevated flow velocity. (B, C) Inspiratory phase US celiac artery, even in deep inspiration, with retraction of the median arcuate ligament, there is compression of the origin of the celiac trunk, with a sagittal diameter of 4 mm, and 10 mm more distally. Flow turbulence is visible at the compressed origin, though angulation of the celiac trunk disappears.

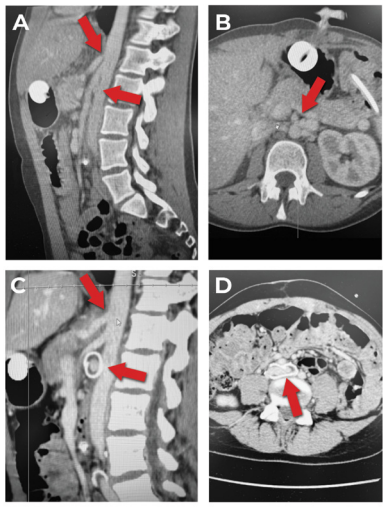

Figure 2: CT Abdomen and Pelvis with contrast. (A). Pretreatment: Visualization of celiac artery compression (top arrow), and reduced angle between aorta and SMA (bottom arrow). (B). Pretreatment: Compression of LRV between aorta and SMA (arrow). (C). Posttreatment: Resolution of celiac artery compression (top arrow). PTFE graft around LRV with increased angle between aorta and SMA (bottom arrow). (D). Posttreatment: PTFE graft around left iliac vein addressing MTS (arrow).

superior in a review of cases [10]. Additionally, neither approach accounts for the hyper-elasticity of spinal ligaments in patients with connective tissue laxity, causing a lordogenetic midline syndrome, which can lead to compression of the duodenum, due to strong lumbar lordosis [4]. Placement of an external PTFE graft around the left renal vein, which was originally published by R. W. Barnes to decompress the LRV for treatment of nutcracker syndrome, [8] has the same mechanodynamic effect to release compression of the third part of the duodenum, as in this case. The technique simply increased the gap between the aorta and SMA, preserving normal anatomy and reduces disruption to the bowel, which is important in a patient with a known connective tissue disorder. It was discussed with the patient that more traditional approaches could be done in the future, if necessary, but this initial method is less invasive.

Understandably, there is concern regarding the integrity of a PTFE graft, especially in young patients. In the experience of author Wilhelm Sandmann, by the technique described above used in more than 200 patients, there have been no known graft migrations. One patient did have collapse of the LIV graft, requiring subsequent repair, after very strong pressure to her lower pelvis three weeks postop.

We hypothesize that the patient presented here developed MALS first, followed by SMAS, due to the lordogenetic nature of compression,[4] due to the gradually increasing lumbar lordosis and exacerbated by weight loss, when her initial symptoms of early satiety, pain and abdominal distention led to decreased oral intake. This hypothesis is supported by her timeline of symptom development, since she began having nausea and bilious vomiting later in the disease process. There has been a connection between MALS in patients with EDS, as in this patient [4,5]. It is also becoming more apparent that AVPCS do co-occur, [2-5] since they all develop in front of the lumbar spine. In patients with EDS and gastrointestinal complaints, it is important to include AVPCS in the differential diagnosis, as timely diagnosis can impact success rates of surgery and prevent long-term complications.

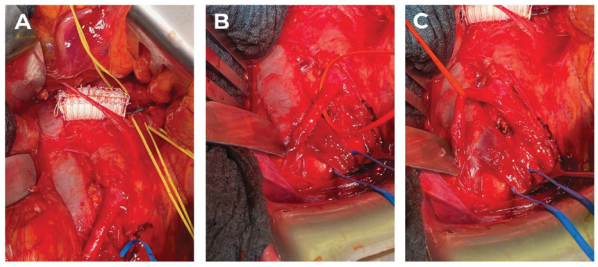

Figure 3: Intraoperative photos. (A). A 16 mm, ring-enforced PTFE graft around the LRV, enlarging the angle between the SMA and aorta, thereby, relieving the duodenal compression and simultaneously protecting the LRV. (B, C). Compression of the left iliac vein by the right iliac artery prior to graft placement.

It is also important to factor in coexisting comorbidities of EDS, such as abnormal colonic transit, gastric emptying, and autonomic dysfunction, [11-13] since these conditions may present simultaneously to the AVPCS and influence the improvement of symptoms. Important autonomic dysfunctions are deducible to AVPCS. Postural orthostatic tachycardia syndrome is a consequence of the impaired venous return to the heart by MTS, aggravated by orthostasis. As seen in this patient, surgery for MALS and SMAS greatly improved her gastrointestinal symptoms but she remains partially reliant on tube feeds for nutritional support. Additionally, the symptoms of orthostatic intolerance experienced prior to surgery are almost completely resolved, likely due to a combination of adequate nutrition and hydration and improved venous return to the heart.

Discussion of patient expectations for postsurgical outcomes is important, especially when other comorbid conditions are present. Outcome uncertainty should not prevent patients from getting surgery, as even partial improvement in symptoms could drastically enhance quality of life, as seen in this patient. Our hope is that as AVPCS become more known, patients receive a faster diagnosis, which can mitigate lingering post-surgery symptoms.

Conclusion

Multiple AVPCS are common in the EDS population and may present with gastrointestinal conditions. Gastrointestinal complaints are common in the EDS population, so AVPCS should be part of the primary differential diagnosis. Because it is common for patients to have more than one compression, a thorough functional workup is recommended with quantitative volume flow measurements in the affected veins and collaterals, Pixel Flux-measurements of pelvic organ congestion [14-17]. In addition, repeat examinations may be necessary while standing, along with postprandially standing and supine after direct observation of the duodenal transport of food across the aorta and frequently changing postprandial position of the SMA-before deciding on a surgical plan-to avoid poor outcomes. The measurement of the angle between the SMA and aorta, despite being widely used, may not correctly diagnose all patients with SMAS. Being aware of all contributing factors to patients' symptoms, including other gastrointestinal disorders, helps establish realistic expectations for improvement after surgical correction of AVPCS.

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