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Progressive Dysphagia and Chronic Abdominal Pain From Vascular Anomalies: A Case Report and Literature Review

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Median arcuate ligament syndrome and symptomatic aberrant right subclavian artery are uncommon in the pediatric population and are rarely found in the same individual. We present the case of a teenager with 2 rare vascular anomalies leading to chronic postprandial abdominal pain, dysphagia, and weight loss. The purpose of this case report is to raise awareness about these rare anomalies and their presentations in the pediatric population.

Key Words: median arcuate ligament syndrome, aberrant right subclavian artery, dysphagia, chronic abdominal pain, weight loss

INTRODUCTION

Median arcuate ligament syndrome (MALS), also known as celiac artery compression syndrome or Dunbar syndrome, is a rare disorder that was first described by Harolja in 1963 ([1]). It was first described in a patient who presented with abdominal pain attributed to mesenteric ischemia caused by compression of the celiac artery by a low-lying median arcuate ligament ([1]). The incidence of MALS is estimated at 2 per 100 000 patients and is said to primarily affect asthenic young women ([2]). In the general population, 10%–24% of people may have indentation caused by an abnormally low ligament; however, only a few of these patients have hemodynamically significant stenosis that causes symptoms ([1]). Aberrant right subclavian artery (ARSA) is the most common aortic arch abnormality, with an estimated prevalence of 0.4%–2% in the general population and is usually asymptomatic. Children usually present with recurrent lung infections or stridor, while adults more often present with dysphagia lusoria ([3]). There are no reports in the literature of a symptomatic pediatric patient with coexistence of both ARSA and MALS. We present case of an adolescent female with a rare presentation of dysphagia and chronic postprandial abdominal pain associated with significant weight loss, thought to be secondary to ARSA and MALS, respectively.

CASE DESCRIPTION

A healthy 17-year-old female presented to our emergency room moderately dehydrated with a history of dysphagia for solids and increasing nausea of 2-week duration. She also had a history of postnasal drip, congestion, and unintentional weight loss of approximately 12 lbs in 2 to 3 weeks and >40 lbs in 6 months before presentation. Medical history was significant for seasonal allergies and scoliosis requiring spinal rods. Examination revealed epigastric tenderness, signs of dehydration, allergic shiners, and swollen turbinates in bilateral nares. Routine laboratory results including complete blood count and comprehensive metabolic panel were normal except for mild metabolic acidosis secondary to dehydration. Esophagogram ordered in the emergency room was normal. She was admitted to the inpatient unit for further management and evaluation. Given her history of allergies, ongoing postnasal drip, and congestion with recent onset of dysphagia, eosinophilic esophagitis was suspected to be the cause of her symptoms and upper gastrointestinal endoscopy was ordered. It revealed pulsatile, shelf-like compression 18 cm from the pharyngoesophageal junction, suspicious for a mass compressing the lumen, with intact and normal mucosa (as seen in Fig. 1). The mucosal biopsies did not reveal any eosinophilic infiltration and showed normal morphology. Chest computed tomography (CT) with contrast, ordered to delineate the mass, showed an ARSA compressing the esophagus (as seen in Fig. 2). Echocardiography, ordered to rule out associated intracardiac anomalies, was normal. Her abdominal pain was presumed to be from non-ulcer dyspepsia at that time, and she was referred to cardiothoracic surgery for advice on management of ARSA which was potentially causing her dysphagia. The patient chose lifestyle modification (small, frequent meals, soft diet, and antacids) over surgical approach, in consultation with the cardiothoracic surgeon. At the follow-up visit 4 months later, her dysphagia had improved, and she had gained 4 lbs, but she continued to experience nausea and postprandial abdominal pain with food aversion from the fear of pain. The family knew an athletic teenager who had recently been diagnosed with MALS after having an extensive evaluation for chronic postprandial abdominal pain. Given the similarity of symptoms, the family requested magnetic resonance angiography (MRA) abdomen to rule out MALS. MRA could not be done due to the presence of posterior spinal stabilization rods, placed for correction of lumbar scoliosis; hence, a functional ultrasound of aorta was ordered. The ultrasound showed a short segment (3 mm) narrowing in the celiac trunk with poststenotic dilatation and turbulent blood flow. An increase in deflection angle from 66 to 74 was noted at the end of expiration. Elevated peak systolic velocity (PSV; >200 cm/s) was noted in both phases of respiration and postprandially (as seen in Fig. 3), with the resolution of all findings in the erect position. These findings supported the diagnosis of MALS and in conjunction with her clinical presentation, MALS was presumed to be the cause of the patient's chronic postprandial abdominal pain. CTA was ordered to confirm the diagnosis, but the family wanted to have her further evaluated by an adult gastroenterologist where she would soon be relocating for college. We have been unable to discuss further workup with our patient since she transitioned care.

DISCUSSION

MALS is more prevalent in women (4:1 ratio) between the ages of 30 to 50 years and in patients with a thin body habitus ([1]). There is no consensus on the underlying mechanism but there are various pathophysiologic mechanisms proposed in the literature including, mesenteric ischemia caused by extrinsic compression

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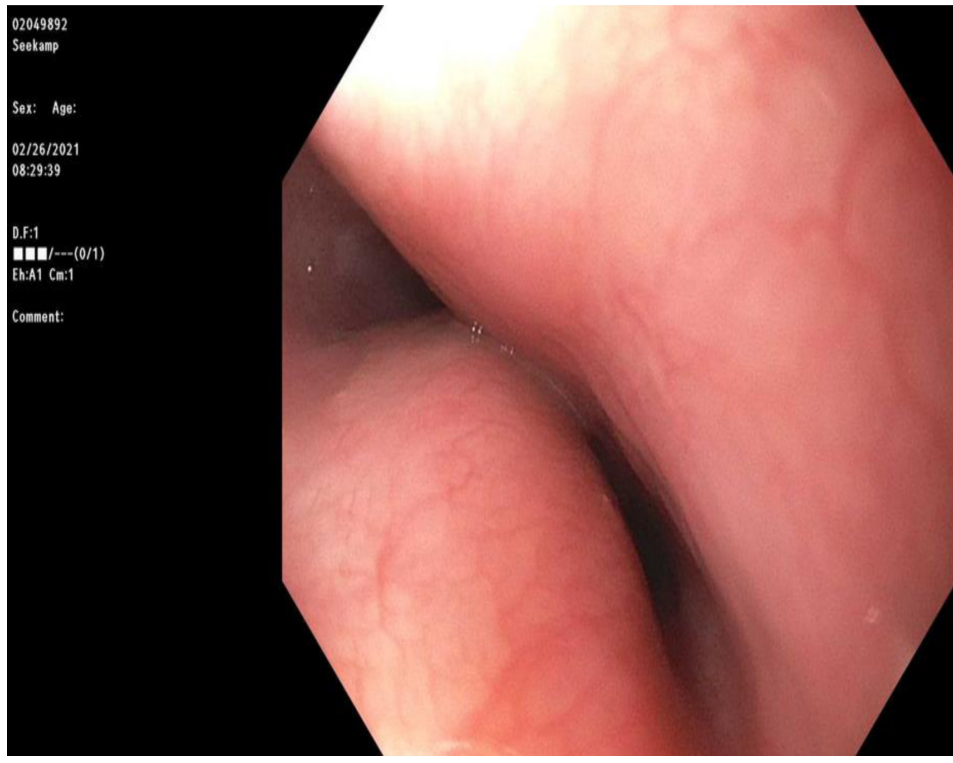

FIGURE 1. Esophagogastroduodenoscopy picture showing compression of esophageal lumen from outside.

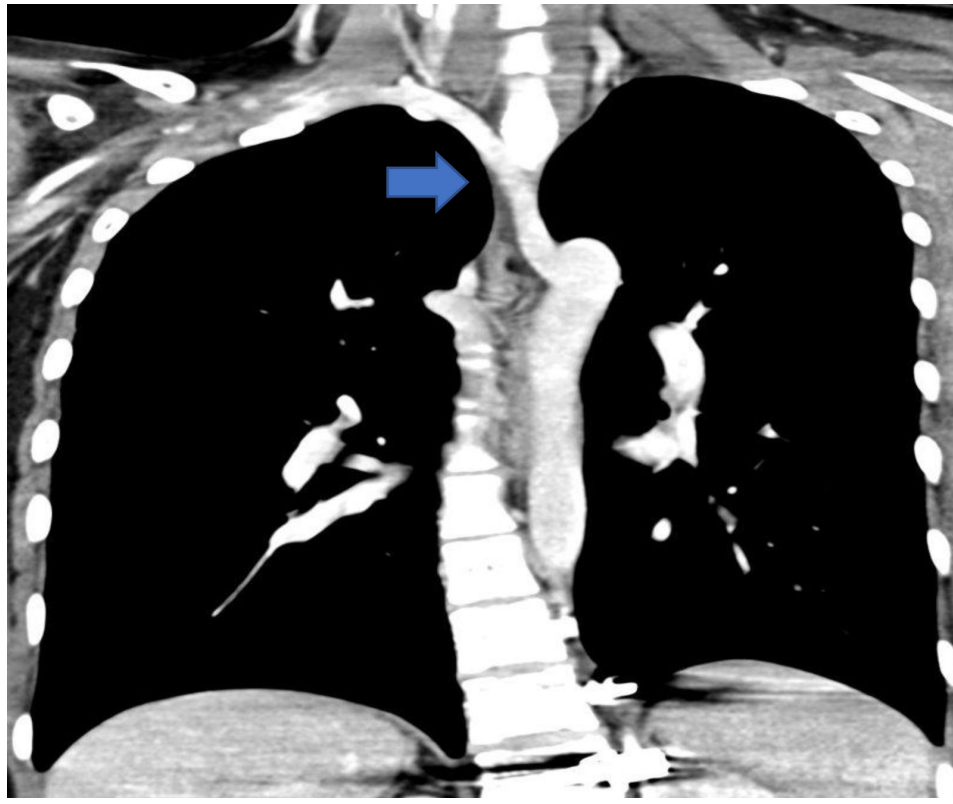

FIGURE 2. Coronal CT chest with contrast showing retroesophageal aberrant right subclavian artery as shown in the image by the arrow. CT = computed tomography.

of the celiac artery at its branching point from the median arcuate ligament or a neurogenic process that might result directly from compression of celiac ganglia by the median arcuate ligament ([4]). However, the most recent studies suggest a neurogenic process over vascular ischemia as the underlying cause of symptoms ([5,6]). Presenting symptoms include recurrent postprandial nausea, vomiting, and weight loss, with aversion to food ([7]) but can rarely present as exercise-related transient abdominal pain in young athletes ([8]). Physical examination may reveal epigastric tenderness or a bruit that is amplified with expiration, but these are nonspecific and observed in only 35% of symptomatic patients ([1]).

The diagnosis of MALS is challenging and is often a diagnosis of exclusion, as the symptoms are very nonspecific and overlap with many other foregut pathologies. After other causes of abdominal pain have been excluded by initial imaging and endoscopic studies, further investigations such as abdominal duplex ultrasound, CT angiography, and MRA abdomen should be considered ([9]). Doppler ultrasound is a noninvasive technique to measure the rate of blood flow through the celiac artery on inspiration and expiration. Doppler flow velocity measurements made at the compressed segment of the celiac artery reveals a variation in the PSV during respiration with PSV >200 cm/s in expiration. The ratio of PSV greater than 3:1 in celiac artery in expiration as compared to PSV in the abdominal aorta immediately below the diaphragm is another criterion to diagnose MALS on ultrasound. In addition, all the measurements should return to normal in erect position to make the diagnosis of MALS ([9]). Selective catheter angiography is the gold standard diagnostic method for diagnosing the condition, and it should be performed during both inspiration and expiration, in the lateral position ([9,10]).

Surgical decompression of the celiac artery and celiac plexus by division of the median arcuate ligament fibers, first described by Harjola and Dunbar in the 1960s, remains the treatment of choice in patients with symptomatic MALS. This can be done by open, laparoscopic, or robotic-assisted approaches. Celiac ganglia block can also provide temporary pain relief in these patients. A large cross-sectional study analyzing the trend of operative management of MALS from 2010 to 2022 showed that out of 33 951 cases identified in the last 12 years, only 2.4% had operative management, with a shift toward the laparoscopic approach due to minimal postoperative complications and decreased hospital stay. In properly selected cases, symptom relief has been reported at 96% postoperatively ([11,12]).

Retroesophageal right subclavian artery also known as ARSA is the most common congenital aortic arch abnormality. It is usually asymptomatic but can rarely be associated with dysphagia, also known as dysphagia lusoria. Digestive or respiratory symptoms appear when posterior compression of the esophagus is present. Progressive dysphagia and severe weight loss have also been reported in patients with this anomaly ([13]).

Upper gastrointestinal endoscopy may show prominent aortic pulsation; however, this is not necessary for the diagnosis. CT or magnetic resonance imaging angiography is considered the gold standard for the diagnosis. It not only confirms the diagnosis but also helps to plan surgery and exclude aneurysm of the aorta or presence of other associated anomalies ([13]). Echocardiography is often used in conjunction for comprehensive evaluation of intracardiac anatomy and function. In the presence of respiratory symptoms, the evaluation usually begins with a chest radiograph; in the presence of noisy breathing, stridor, or brassy cough, flexible bronchoscopy is the procedure of choice ([14]).

In moderate cases, lifestyle changes and education on eating behavior can provide significant symptomatic relief. The use of a surgical approach is still controversial and has no clear indications, but generally depends on the urgency of the situation and to provide relief of symptoms ([15,16]).

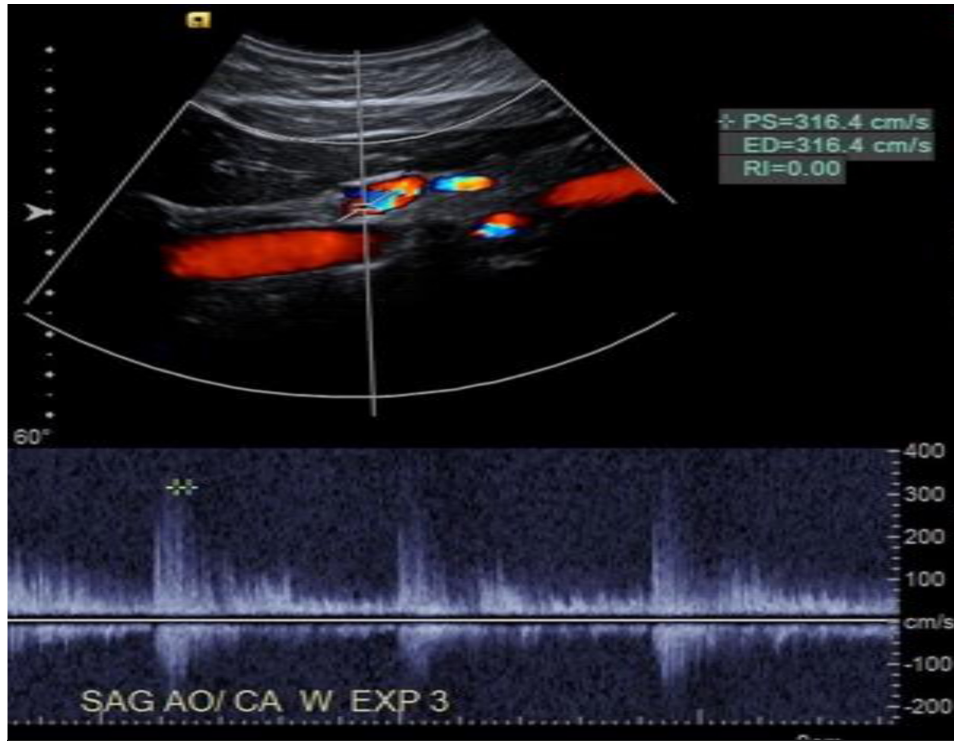

FIGURE 3. Longitudinal duplex ultrasound of aorta with spectral analysis showing elevated expiratory peak systolic velocities in celiac axis in supine position of 316 cm/s which resolved in erect position (normal peak systolic velocities in celiac axis is 50–160 cm/s).

There has been no literature to suggest a relationship between these 2 rare vascular anomalies or any underlying genetic or connective tissue disorder to explain the coexistence in a single individual. There are several reports of MALS being associated with Myhre syndrome, Ehlers Danlos syndrome, postural orthostatic tachycardia syndrome, and other visceral vascular anomalies ([17,18]). Similarly, ARSA has been reported in association with Marfan syndrome which points toward a possible pathophysiological relationship, but more studies are needed to establish this relationship ([19]).

CONCLUSIONS

Chronic abdominal pain in the pediatric population is often diagnosed as functional abdominal pain (like functional dyspepsia or abdominal migraine) in the absence of any demonstrable infectious, inflammatory, or anatomical abnormality. MALS has a similar presentation and therefore often goes undiagnosed ([13]). ARSA is a rare cause of dysphagia or respiratory symptoms (stridor, respiratory distress, or cough) in children which can often go undiagnosed until adolescence or adulthood when compression symptoms often present ([13]). Although rare, patients with these anomalies may benefit from surgical intervention when symptomatic, which is why it is important to consider a broader differential in patients with dysphagia and/or chronic abdominal pain. Hence, the purpose of this case report is to raise awareness about these vascular anomalies and their clinical presentations.

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