WASTING AWAY – SUPERIOR MESENTERIC ARTERY SYNDROME IN A YOUNG GIRL – A CASE REPORT

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ABSTRACT

The case of an 18-year-old girl with Superior Mesenteric Artery syndrome is presented here. She presented to us with complaints of anorexia, nausea, vomiting, and weight loss for the past year. She was cachectic and volume-depleted. All investigations and preliminary imaging were inconclusive. Upper GI endoscopy revealed normal structure up to the second part of the duodenum. Contrast-enhanced Computerized Tomography of the abdomen and pelvis did not reveal any intra- or extra-luminal pathology. The patient was unwilling to undergo surgical correction and opted for conservative treatment. This case highlights the importance of considering SMA syndrome in the differential diagnosis of patients presenting with anorexia and weight loss and emphasizes the changing epidemiology of SMA syndrome.

Keywords: Superior Mesenteric Artery Syndrome; Anorexia Nervosa; Weight Loss; Wilkie's syndrome

INTRODUCTION

Superior Mesenteric Artery Syndrome, also known as cast syndrome, which refers to a rare condition characterized by the compression of the third part of the duodenum by the superior mesenteric artery. The SMA traverses the anterior wall of the duodenum, acuity of the aforementioned angle can lead to external compression of the lumen leading to symptoms of upper gastrointestinal obstruction.[1] Clinical presentation varies between acute and chronic forms of intermittent abdominal pain and vomiting leading to a vicious cycle of weight loss and worsening obstruction.[2] The initial weight loss that leads to the propensity of the SMA to obstruct the duodenum can have several causes however, as this case would subsequently demonstrate, a cause may not always be readily apparent. The first step towards the diagnosis of Superior Mesenteric Artery Syndrome is to suspect it in at risk individuals. A conclusive diagnosis is established radiologically.[3] Management is either conservative with nutritional support or surgical with duodenojejunostomy being the procedure with the highest success rate, however, due to the rarity of the condition, follow-up studies are sparse.[4]

CASE REPORT

An 18-year-old girl presented to us with the major symptoms of anorexia (loss of appetite), nausea, and vomiting for the past month. She had been having these symptoms for the past year; however, they had worsened in the previous month. The family reported significant weight loss associated with these complaints. She had severe nausea when attempting to eat food and could only tolerate small amounts of liquid diet. The vomiting was non-bilious, non-projectile, contained semi-digested food and had no blood. She did not endorse dysphagia, odynophagia, abdominal pain, diarrhea, or constipation. She denied fevers, night sweats, myalgias, arthralgias, headaches, and visual blurring. She had a normal menstrual cycle of 5/28 days. The rest of the systemic review was unremarkable. The past medical history of the patient was a necessary step for admission to a nearby hospital a month earlier for the same complaints, the records of which were not available. She did not have any past surgeries and had no history of prescription or recreational drug use. She did not have any drug or food allergies. She is single and lives with her family which includes her

parents and four siblings. She studies at a local public school upon examination, she was severely cachectic with a BMI of 13.3 kg/m2. With a resting tachycardia of 120 bpm with a blood pressure of 90/60 mmHg. She was afebrile and maintained saturations while breathing ambient air. She did not have pallor, cyanosis, jaundice, or lymphadenopathy. Oral cavity examination was significant for aphthous ulcers and poor dental hygiene. A cardiovascular examination revealed tachycardia but no added sounds. There was no organomegaly and no masses were palpated. There was

no distension, flank dullness, or succussion splash. Bowel sounds were normal. Central nervous system examination including gait was normal. Peripheral pulses were palpable and there were no rashes or edema.

Initial investigations include a complete blood count, renal function tests, serum electrolytes, liver function tests, coagulation profiles, and thyroid profiles were all within range. Viral serologies for hepatitis B, C, and HIV were negative. Random serum cortisol was high. Anti-tissue transglutaminase IgA antibodies were negative. A pregnancy test was also negative. Serum calcium, serum ferritin, and vitamin D levels were normal. EKG showed sinus tachycardia, a chest radiograph was normal and so was the abdominal ultrasound. A psychiatric evaluation was taken to rule out anorexia nervosa. They were unable to find any signs suggestive of an eating disorder. An upper gastrointestinal endoscopy was planned. The endoscopy revealed mild esophagitis and a stomach that was full of partially digested food material. The procedure was thus abandoned. A nasogastric tube was passed and the stomach decompressed. Meanwhile, she was started on intravenous fluids and a CT Abdomen was planned. The Upper GI endoscopy was repeated and revealed normal stomach and pylorus. The first and second part of the duodenum was also normal however the third part appeared compressed. The CT Abdomen did not reveal any intra or extra-luminal pathology but was suggestive of Superior Mesenteric Artery syndrome. A surgical consultation was taken and a laparoscopic duodenojejunostomy was planned however the patient declined and decided to try the conservative treatment. She was advised to lie in a lateral recumbent position after meals to allow decompression of the duodenum and food to pass through. A nutritionist was consulted for gradual up-titration of calories to avoid refeeding syndrome. At one month follow-up, she is doing well, however weight gain has not been substantial. She does not report any further anorexia or vomiting. Informed consent was taken from the patient.

DISCUSSION

Superior Mesenteric Artery Syndrome was initially described more than a century ago by Bohemian pathologist and physician Carl Rokitansky. He described it in autopsy reports of people who had succumbed to the condition. The first formal description of SMA syndrome came from Wilkie in 1927 when he described 75 patients diagnosed with the disorder.[5] Subsequently, more than 400 cases have been described in the literature in the form of case reports, case series, and reviews.[6] SMAS is one of several rare gastrovascular compression syndromes, others being median arcuate ligament syndrome, May-Thurner syndrome, nutcracker syndrome, portal biliopathy, and ureteropelvic junction obstruction.[7] The syndrome has been described in all age groups however most patients are between the ages of 10 and 39 years. Females are much affected and the estimated prevalence is between 0.013 % to 0.3 % in various reports.[9]

Clinical presentation of SMA syndrome is non-specific and depends upon the acuteness and chronicity of the illness. Acute SMAS usually presents with abdominal cramps and a feeling of vomiting. Chronic SMAS presents with anorexia, weight loss, and intermittent episodes of nausea, epigastric pain, reflux, and vomiting which may at times be bilious. Symptoms may resolve in the left lateral decubitus position or the knee-chest position after meals. Physical examination may well reveal severe cachexia and abdominal distension or a succussion splash.[4]

The diagnosis of SMAS is radiological. The gold standard for diagnosis is CT or MR arteriography which provides information regarding the aortomesenteric distance as well as the angle and assessment of the mesenteric fat pad. A review of cases described in the local and international literature reveals that almost all cases of Superior Mesenteric Artery Syndrome are associated with a predisposing condition such as surgery[10] or anorexia nervosa.[11] A thorough psychiatric assessment of the patient in our case was done to rule out anorexia nervosa. Other causes were similarly ruled out based on history, examination, and basic testing.

CONCLUSION

Superior mesenteric artery syndrome is a clearly defined radiological entity however diagnosis relies upon consideration of the syndrome in at-risk populations. The case that we have described is unique in that a cause for the initial weight loss that perpetuated the cycle of vomiting, anorexia, and more weight loss could not be ascertained. This indicates that SMAS may be idiopathic in some patients.

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Author's Contribution

AR and JS received the case and contributed in the write-up of the manuscript. FN and AN helped in editing and proofreading of the manuscript. Authors agree to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Conflict of Interest

Authors declared no conflict of interest

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None

Data Sharing Statement

The data that support the findings of this study are available from the corresponding author upon reasonable request.