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Case Report

Superior Mesenteric Artery Syndrome in an Adolescent Female

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Abstract

Superior Mesenteric Artery (SMA) syndrome is a relatively rare diagnosis. In this case, report we describe an adolescent female who presented to our Emergency Department with chronic abdominal pain and weight loss. We describe the etiologies, relevant radiologic findings, treatment of SMA syndrome.

Keywords: Wilkie's syndrome; Aorto mesenteric angle; Radiographic investigations; Anorexia nervosa

Case Presentation

A 14-year-old girl was referred to our Emergency Department (ED) for possible intestinal obstruction. She had history of chronic abdominal pain for two months. The pain was localized to the epigastric region, without radiation, and worsened with eating. As a result, she started eating less and endorsed a weight loss of between 20-30 pounds over the two month period. She developed emesis two days prior to presentation, which progressed to bilious emesis, which prompted the ED visit. She denies fever, diarrhea and blood in bowel movements, urinary symptoms, rash, or other systemic symptoms. Of note, she reported that she had not had her menstrual period for over four months and endorsed a feeling of constantly feeling cold. Diagnostic studies including complete blood count and comprehensive metabolic panel was within normal limits. Serum metabolic panel showed hypernatremia, hypokalemia with mild metabolic alkalosis. Computerized Tomography abdomen (Figure 1) was obtained which showed a collapsed duodenum with a superior mesenteric artery angle of 10 degrees diagnostic for superior mesenteric artery syndrome. Thereafter, she was transferred to our institution for further evaluation and management. Her vital signs in our ED were significant for bradycardia, with heart rate ranging 49-60 beats/minute and low normal blood pressures ranging 86-95/43-50 mmHg. Her physical examination was significant for a cachetic adolescent who was undernourished with weight of 30.3 kilograms (<3rd percentile) and BMI 13.9 kg/m^2. Her abdominal exam was remarkable for epigastric tenderness without any masses, hepatosplenomegaly or peritoneal signs. X-ray abdomen showed a fluid filled distended stomach but the bowel gas pattern was non-obstructive. Pediatric surgery was consulted and an upper GI study (Figure 1) was obtained to rule out intestinal malrotation and it was normal. The patient's SMA syndrome was determined to be secondary to anorexia and weight loss. She was started on nasogastric feeds and was subsequently transitioned to oral feeds with a slow increase in the calorie count to prevent refeeding syndrome. At discharge, she was referred to an outpatient comprehensive treatment program for eating disorders.

Discussion

SMA syndrome was first reported in 1861 by Von Rokitansky, but was described in detail by Wilkie in 1927 in a case series of 75 patients. It is a rare occurrence with an estimated incidence between 0.1-0.3 %. It is characterized by compression of the third portion of the duodenum between the aorta and the SMA resulting in an acute partial or complete duodenal obstruction [1]. When viewed laterally, the SMA is seen to run ventrally and caudally from the anterior aspect of the aorta, enveloped by lymphatic tissues and mesenteric pad of fat. The acute downward angle ranges from 380 to 650 and the aortomesenteric distance is usually 10 to 28 mm. Any factor decreasing the intervening space around the duodenum or altering its relation to the surrounding anatomical structures will result in external compression or occlusion of the duodenum, by decreasing the aorto-mesenteric angle and distance to about 60 to 160 and 2 mm to 8 mm respectively [2]. SMA syndrome is associated with hypermetabolic conditions such as trauma and burns, dietary conditions such as anorexia nervosa and malabsorptive diseases and malnourishment conditions such as AIDS and malignancy, reduced peristalsis from diabetes, connective tissue diseases such as scleroderma, and low muscle tone disorders such as myotonic dystrophies [3]. Other risk factors include surgical correction of scoliosis, congenitally short or hypertrophic ligament of Treitz, peritoneal adhesions, duodenal malrotation with Ladd's bands and aneurysm of abdominal aorta.

Patients typically present with chronic epigastric abdominal pain and vomiting along with significant weight loss as hallmark of the disease [4]. The symptoms are chronic lasting anywhere between

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8-28 months prior to diagnosis. Diagnosis is often made with Upper GI series or CT abdomen. Key findings include the reduced aortomesenteric angle and distance, gastroduodenal distention and bowel caliber narrowing at the takeoff of the SMA from the aorta. Upper GI can also show delayed gastric emptying or positional obstruction [5]. Management is directed at underlying etiology i.e. nutritional support and electrolyte monitoring for those patients whose SMA syndrome is secondary to weight loss. Surgical intervention is an option for those patients refractory to medical management and in those whose SMA syndrome is secondary to physical compression [3]. Complication stem both from the underlying diagnosis, as well as from treatment. Re feeding syndrome is perhaps the most well known complication and occurs when nutrition replenishment is rapidly implemented. Patients may experience electrolyte disturbances such as hypokalemia, hypomagnesaemia, and hypophosphatemia as these intracellular ions are rapid utilized [6] and will require close monitoring for the same.

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