Case Report

A Rare Case of Wilkie's Syndrome or Superior Mesenteric Artery Syndrome

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Abstract

Superior Mesenteric Artery (SMA) syndrome (known as Wilkie's syndrome) is a rare cause of upper gastrointestinal obstruction. It is an acquired disorder, causes compression of the third part of the duodenum between the SMA and the aorta, due to loss of fatty tissue as a result of a variety of debilitating conditions. We report a case of 18 year old female presented with history of intermittent abdominal pain, vomiting of 6 months duration followed by significant weight loss for which she is hospitalized. Patient underwent extensive invasive and non-invasive tests, thereafter revealed her diagnosis. This case emphasizes the challenges in the diagnosis of SMA syndrome and the need for increased awareness of this entity.

Key words

Wilkie's syndrome, Superior Mesenteric Artery Syndrome, Case Report.

Introduction

Superior Mesenteric Artery syndrome is an unusual cause of proximal intestinal obstruction. Superior mesenteric syndrome is a rare pathology with an incidence estimated at 0.1% to 0.3%. SMA syndrome preferentially occurs in adolescents and young adults with a general age range of 10 to 39 years old but can ultimately occur at any age. It occurs more commonly in females over males with a ratio of 3:2. No ethnic predisposition has been described, but familial cases have been reported [1, 2, 3].

SMA syndrome was first described by the Austrian professor Carl Freiherr von Rokitansky in 1861 as an autopsy finding [4]. It has been referred by a variety of other names, including Cast syndrome, Wilkie

syndrome, arterio mesenteric duodenal obstruction, and chronic duodenal ileus.

The third portion of the duodenum is cradled in an angle of approximately 45 degrees formed by the root of the superior mesenteric artery and the wall of the aorta. When this angle is narrowed to less than 25 degrees, the superior mesenteric artery impinges on the duodenum, thereby leading to gastric and intestinal obstruction, a condition referred to as Wilkie's syndrome or the superior mesenteric artery syndrome.

In adults, clinical SMA syndrome manifestations appear if the angle drops below 20°, and it is believed that values of this angle may be lower for pediatric patients [5]. Thus, in the appropriate clinical context, detailed history as well as imaging findings should highly raise the clinical suspicion for the diagnosis of SMA syndrome.

A delay in this diagnosis can potentially lead to many complications, such as electrolyte imbalance, catabolic wasting, peritonitis and gastric perforation. Conservative therapy mainly consists of weight gain achieved orally or parenterally, with the aim of restituting the mesenteric fat pad and increasing the aortomesenteric angle [6]. If this non-invasive approach fails, surgical therapy may be the next approach, with duodenojejunostomy being the currently preferred treatment [7].

However, advances in imaging, such as in computed tomography (CT) and magnetic resonance imaging, have tremendously helped with clear visualization of the angle between the aorta and the SMA and thus improved the diagnostic rate [8].

Case report

We present here a case of 18 year female who presented with history of abdominal pain of 6 months duration. Pain was diffuse, constant, non-radiating, squeezing type, not related to food intake, partially relieved by medication. Associated with vomiting since 6 months which were non-projectile, non-bilious, non-blood tinged, 4-5 episodes/day, content – food particles. She was admitted for the above complaints 6 months ago for two days at a local hospital following which vomitings temporarily subsided. There was an associated history of loss of appetite and loss of weight. No history of fever, loose stools. She was cachectic with a BMI of 10.32 kg/m^2. No co-morbidities were present. Vitals were stable.

On examination pallor was present, diffuse abdominal tenderness was noted. Other system examination was normal.

Investigations

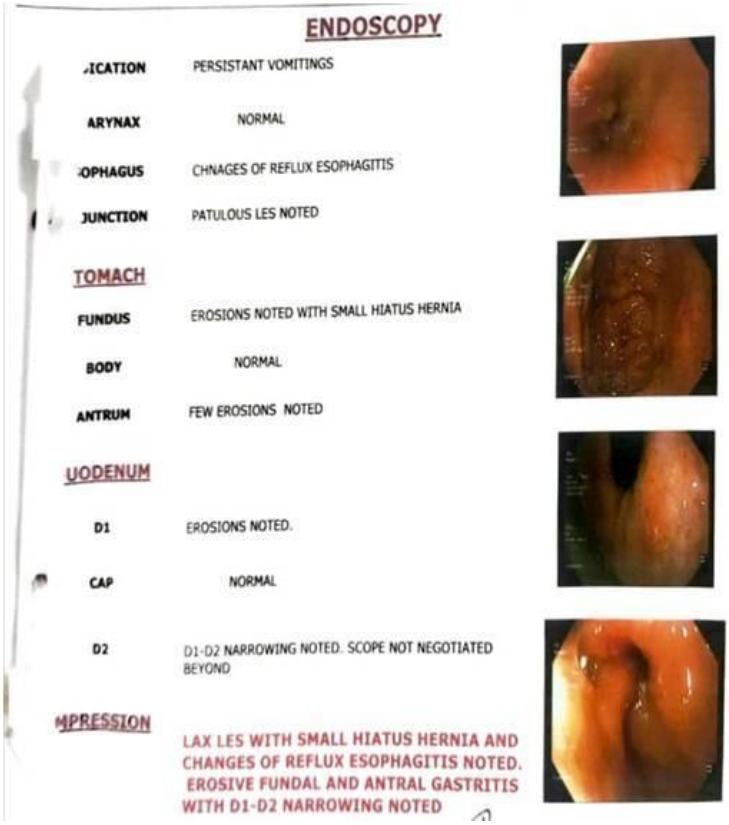
Hb - 8.1 g/dL (MCHC)
ESR – 50 mm/hr
LFT: Protein - 5.2 g/dl
Albumin – 2.9 g/dl
RFT – Normal
Viral serology – Negative
Chest X-Ray, X - RAY erect Abdomen – Normal
Based on the patients history, general examination and USG Abdomen findings of Mesenteric lymphadenopathy largest measuring 9 x 4 mm a diagnosis of abdominal tuberculosis was contemplated. It was ruled out by a normal colonoscopy finding.

Further upper GI endoscopy was done in view of persistent vomiting it showed – D1-D2 Narrowing noted (**Figure – 1**).

CECT Abdomen showed dilated stomach duodenum up to D2. Significant narrowing at D3, appears to be compressed between SMA and Aorta - SMA syndrome (**Figure – 2**).

Patient was diagnosed to have SMA syndrome. Conservative management with nasogastric tube placement, high protein, nutritious diet was chosen, clinical improvement was noticed and patient was discharged and asked to follow up.

Figure - 1: UGIE showing D1-D2 Narrowing.



<u>Figure - 2A</u>: Contrast enhanced computed tomography of the abdomen in Venous phase showing compression of third part of the duodenum between superior mesentery artery and aorta.
Blue arrow- aorta, red arrow – superior mesenteric artery,
Green arrow – 3rd part of duodenum

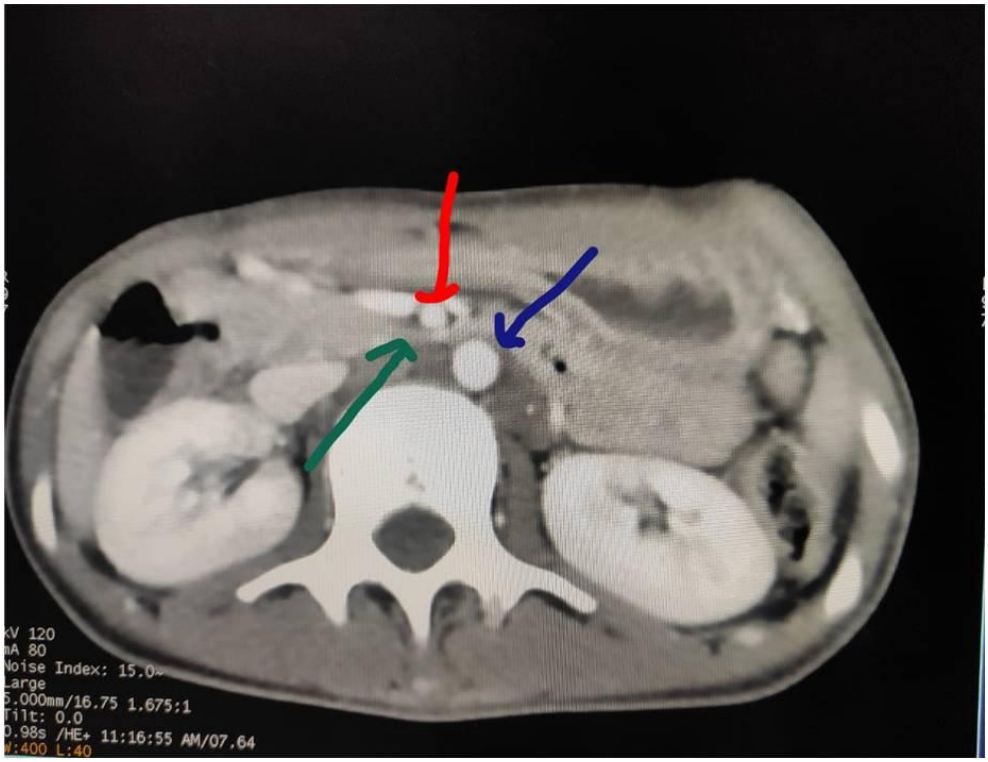
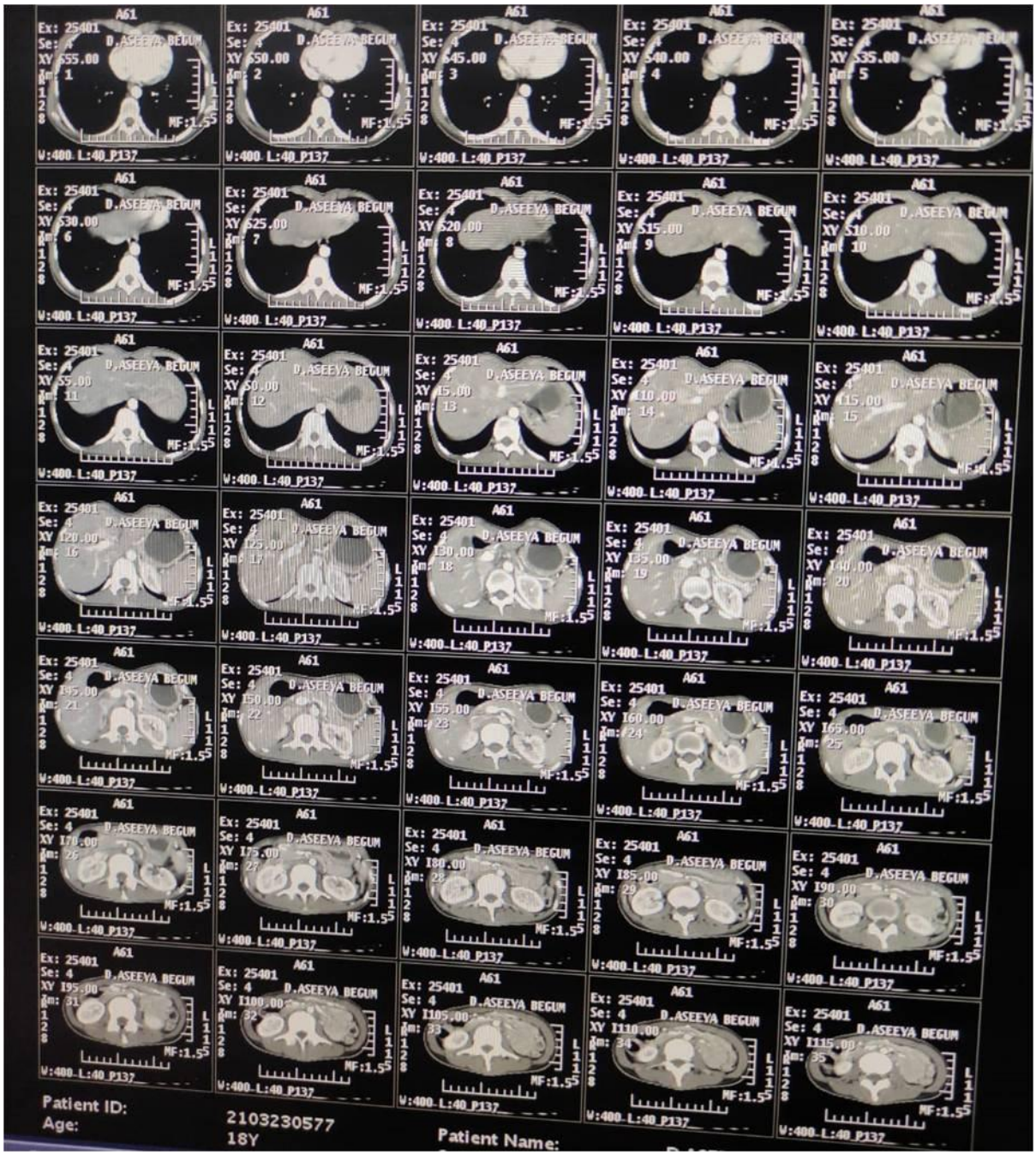


Figure - 2B: Contrast enhanced computed tomography of the abdomen revealed compression of third part of the duodenum between superior mesentery artery and aorta.



Discussion

SMA syndrome was first described by the Austrian professor Carl Freiherr von Rokitansky in 1861 as an autopsy finding [4]. Later, Wilkie provided a more detailed clinical and pathophysiologic description in a series of 64 patients and suggested treatment approaches [5]. After that, a controversy regarding the actual existence of this syndrome started, especially because of the lack of specificity of the symptoms and the long list of differential diagnoses

- Anorexia bulimia
- Anorexia nervosa
- Chronic idiopathic intestinal pseudo-obstruction
- Collagen vascular conditions
- Diabetes mellitus
- Mega duodenum and duodenal ileus
- Peptic ulcer disease

Duodenal compression is usually due to the loss of the intervening mesenteric fat pad between the aorta and SMA, which in turn, results in a narrower angle between the vessels. The fat pad cushion functions to hold the SMA off the spine and protect it from duodenal compression. A normal aortomesenteric angle is 38 to 65 degrees; however, decreasing the angle less than 25 degrees will decrease the distance to less than 10 mm and cause compression to the third part of the duodenum. Decreases in the aortomesenteric angle can be either congenital or acquired. SMA syndrome is associated with significant weight loss including situations of hypermetabolism (trauma and burns) dietary conditions (anorexia nervosa and malabsorptive diseases) and cachexia causing conditions (AIDS, cancer, paraplegia). Other risk factors include surgical correction of scoliosis, congenitally short or hypertrophic ligament of Treitz, peritoneal adhesions, duodenal malrotation, Ladd's bands, abdominal aortic aneurysm, lumbar hyperlordosis, and mesenteric root neoplasm.

Patients with SMA syndrome may present acutely, with chronic insidious symptomatology, or with an acute exacerbation of chronic symptoms. Acute presentation is usually characterized by signs and symptoms of duodenal obstruction. Chronic cases like our patient may present with long-standing vague abdominal symptoms or recurrent episodes of abdominal pain, associated with vomiting. Other less common symptoms are esophageal reflux, early satiety with a sensation of fullness owing to increased gastroduodenal transit time, and gastric distension [9, 10, 11, 12, 13].

Diagnosis is based on interpreting clinical symptoms alongside radiological testing which can confirm its presence. Various imaging modalities that can be used include plain film x-ray, barium x-ray, endoscopy, computed tomography (CT), Doppler ultrasound, and magnetic resonance angiography (MRA). Plain radiographs may reveal a dilated stomach and diminished distal bowel gas. Endoscopy and barium studies can be used but are often nonspecific and not available in an emergency setting. Endoscopy, rather, can be used to investigate complications of the disease including gastric stasis, biliary reflux, gastritis and duodenal ulcers and to rule out other cause of the duodenal compression. CT scan is helpful in diagnostics in that it allows for measurement of aortomesenteric (AO) angle which aids in confirmation of SMA syndrome and has thus replaced MRA as the standard for diagnosis. The normal AO angle is between 38 to 65 degrees and has a distance of 10 to 28 mm.

In a study which reviewed 8 cases of SMA syndrome, a reported AO angle cut off of 22 degrees revealed a 42.8% sensitivity and 100% specificity, and a distance of 8 mm was both 100% sensitive and specific for the condition.

Laboratory tests are usually non-diagnostic and it is noted that electrolyte disturbances as well as protein and albumin levels can still be normal despite associated weight loss. While it is rare, SMA syndrome is important to consider because the delay in diagnosis can result in significant morbidity and mortality from malnutrition, dehydration, electrolyte abnormalities, gastric pneumatosis and portal venous gas, gastrointestinal hemorrhage and gastric perforation [14, 15, 16].

Traditionally treatment has consisted of conservative measures such as gastric decompression, parenteral nutrition and/or post-pyloric feeding when possible, followed by oral diet as tolerated [6]. Posturing maneuvers during meals and motility agents may be helpful in some patients. No time limit has yet been defined for the medical treatment. Surgery may be considered if conservative treatment fails [7]. Duodenojejunostomy is the operation of choice to relieve the obstruction, with a success rate up to 90% [7]. Another less invasive surgical option, known as Strong's procedure, involves lysis of the ligament of Treitz with mobilization of the duodenum; however, this operation had a failure rate of 25% [10]. Gastrojejunostomy, a previously reported surgical treatment, has been

abandoned because of increased postoperative complications like blind loop syndrome and recurrence of symptoms due to non-decompression of the duodenum.

Conclusion

We feel that SMA syndrome is under diagnosed. High clinical suspicion is of utmost importance; especially in patients with severe weight loss and symptoms of gastric distension Interdisciplinary teamwork provides the most beneficial diagnostic and therapeutic result in this often underestimated disease. Computed tomography angiography is considered as the gold standard for diagnosis and in case of failure of medical treatment, open duodenojejunostomy and laparoscopic duodenojejunostomy have been described as the best surgical treatment option.

References

1. Zhang ZA. Superior mesenteric artery syndrome: a vicious cycle. BMJ Case Rep., 2018 Oct 24; 2018.
2. Ehlers TO, Tsamalaidze L, Pereira L, Stauffer J. Laparoscopic Duodenojejunostomy for the SMA Syndrome. Zentralbl Chir., 2018 Oct; 143(5): 461-463.
3. Hillyard J, Solomon S, Kaspar M, Chow E, Smallfield G. Gastrointestinal: Reversal of superior mesenteric artery syndrome following pregnancy. J Gastroenterol Hepatol., 2019 Mar; 34(3): 486.
4. Rokitanski CV. Vienna, 3rd edition, Vol. 3, Braumulller and Siedel: 1861. Lehrbuch der Pathologische Anatomie; p. 187.
5. Wilkie D. Chronic duodenal ileus. Am J Med Sci., 1927; 173: 643–649.
6. Welsch T, Buchler MW, Kienle P. Recalling superior mesenteric artery syndrome. Dig Surg., 2007; 24: 149–156.
7. Mandarry M, Zhao L, Zhang C, Wei Z. A comprehensive review of superior mesenteric artery syndrome. Eur Surg., 2010; 42: 229–236.
8. Unal B, Aktaş A, Kemal G, Bilgili Y, Güliter S, Daphan C, Aydinuraz K. Superior mesenteric artery syndrome: CT and ultrasonography findings. Diagn Interv Radiol., 2005; 11: 90–95.
9. Mandarry M, Zhao L, Zhang C, Wei Z. A comprehensive review of superior mesenteric artery syndrome. Eur Surg., 2010; 42: 229–236.
10. Merrett ND, Wilson RB, Cosman P, Biankin AV. Superior mesenteric artery syndrome: diagnosis and treatment strategies. J Gastrointest Surg., 2009; 13: 287–292.
11. Ahmed AR, Taylor I. Superior mesenteric artery syndrome. Postgrad Med J, 1997; 73: 776–778.
12. Tsirikos AI, Anakwe RE, Baker AD. Late presentation of superior mesenteric artery syndrome following scoliosis surgery: a case report. J Med Case Rep., 2008; 2: 9.
13. Reynolds EW, Kinnard TB, Kriss VM, Perman JA. Superior mesenteric artery syndrome: an uncommon cause of feeding intolerance in infancy. J Pediatr Gastroenterol Nutr., 2008; 46: 92–95.
14. Ganss A, Rampado S, Savarino E, Bardini R. Superior Mesenteric Artery Syndrome: a Prospective Study in a Single Institution. J Gastrointest Surg., 2019 May; 23(5): 997-1005.
15. Silva G, Moreira-Silva H, Tavares M. Iatrogenic superior mesenteric artery syndrome. Rev Esp Enferm Dig., 2018 Nov; 110(11): 742-743.
16. Young A, Kinnear N, Hennessey D, Kanhere H, Trochsler M. Intermittent superior mesenteric artery syndrome in a patient with multiple sclerosis. Radiol Case Rep., 2018 Dec; 13(6): 1108-1111.