

Wilkie's syndrome: A rare cause of vomiting and weight loss

Ali Coşkun, Deniz Uçar, Erdem Barış Carti, Erkan Oymaci, Mehmet Yıldırım, Nazif Erkan

ABSTRACT

Introduction: Superior mesenteric artery (SMA) syndrome, also known as Wilkie's syndrome, is extremely rare and is characterized by postprandial epigastric pain, nausea, vomiting and loss of appetite, with subsequent weight loss, which aggravates the condition of the patients. The syndrome is caused by compression of the third part of the duodenum in the angle between the aorta and the superior mesenteric artery. Herein, we presented a patient with Wilkie's syndrome and discussed the diagnostic difficulties and surgical treatment options.

Case Report: We report a case of 13-year-old female who was diagnosed with SMA syndrome. There was no history of recent trauma, surgery, prolonged immobilisation or neurological illness. Her weight loss was gradual. The patient was initially diagnosed as anorexia nervosa due to her symptoms of vomiting and anorexia without any clinical cause, but later on, she underwent computed tomography scan and was diagnosed with SMA syndrome. Her SMA syndrome, resolved after successful nonoperative management based on accepted guidelines for nutritional therapy, thus avoiding the need for operation. One year follow-up was uneventful.

Conclusion: Superior mesenteric artery syndrome is a rare cause of intestinal obstruction and its first-line treatment is usually conservative with jejunal or parenteral nutrition for restoration of the aortomesenteric fatty tissue. If conservative management fails, surgical options, open or laparoscopic duodenojejunostomy or duodenal mobilization. Its recognition is important because early diagnosis of a partial obstruction may allow for medical rather than surgical intervention, as exemplified by our case.

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usually conservative with jejunal or parenteral nutrition for restoration of the aortomesenteric fatty tissue. If conservative management fails, surgical options, open or laparoscopic duodenojejunostomy or duodenal mobilization. Its recognition is important because early diagnosis of a partial obstruction may allow for medical rather than surgical intervention, as exemplified by our case.

Keywords: Superior mesenteric artery (SMA) syndrome, Wilkie's syndrome, Weight loss, Vomiting, Enteral feeding

How to cite this article

Coşkun A, Uçar D, Carti EB, Oymaci E, Yıldırım M, Erkan N. Wilkie's syndrome: A rare cause of vomiting and weight loss. International Journal of Case Reports and Images 2013;5(2):150–154.

doi:10.5348/ijcri-2014-02-461-CR-13

INTRODUCTION

Superior mesenteric artery (SMA) syndrome which is also known as Wilkie's syndrome, is a rare cause of upper gastrointestinal obstruction [1–3]. Superior mesenteric artery syndrome was first described in 1861 by von Rokitansky, who proposed that its cause was obstruction of the third part of the duodenum as a result of arteriomesenteric compression [4]. Later, Wilkie provided a more detailed clinical and pathophysiologic description in a series of 64 patients and suggested treatment approaches. This is usually associated with conditions that cause significant weight loss, such as anorexia nervosa, malabsorption, or hypercatabolic states such as burns, major surgery, severe injuries, or malignancies as fat loss causes direct compression of third part of duodenum with superior mesenteric

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Received: 02 August 2013
Accepted: 19 September 2013
Published: 01 February 2014

artery anteriorly [3]. Despite diagnostic confusion with intestinal dysmotility syndrome, conservative therapy with nutritional supplementation is the initial approach and duodenojejunostomy is favored, if non-surgical treatment fails.

CASE REPORT

A 13-year-old female who was initially diagnosed as anorexia nervosa due to her symptoms of vomiting, anorexia and weight loss for 12 months, admitted to our emergency service. There was no history of recent trauma, surgery, prolonged immobilisation or neurological illness. Her weight loss was gradual. Abdominal examination revealed a distended abdomen, mild epigastric tenderness, and hyperactive bowel sounds. There was no palpable organomegaly. Her hemoglobin was 10 g/dL, leucocyte count was $8.0 \times 10^3/\text{mm}^3$. Serum albumin was 2.5 g/dL and rest of the investigations were normal. Abdominal radiograph revealed a dilated stomach with a prominent air fluid level. Subsequent abdominal computed tomography (CT) scan revealed dilatation of stomach, first and second part of duodenum. Third part of duodenum was compressed between superior mesenteric artery (SMA) and superior mesenteric vein (SMV) anteriorly and aorta and vertebrae posteriorly. On prone position, the narrowing of fourth part opens up but dilatation of third part persists. These features were suggestive for SMA syndrome (Figures 1 and 2). The small bowel distal to the SMA was decompressed. Endoscopic examination of the upper gastrointestinal tract revealed mild esophagitis, dilated stomach and proximal duodenum, and narrowing of the third part of the duodenum due to a pulsating extrinsic compression. She was hospitalized and total parenteral nutrition was administered with Daily 950 kcal (35 kcal/kg/d) of oliclinomel N7 (Baxter, Eczacıbaşı, Turkey). After 15 day of admission, she tolerated oral nutrition. She gained 4 kg weight before discharge and she has been following without any complication and complain for one year.

DISCUSSION

Superior mesenteric artery syndrome is an uncommon but well recognized clinical entity characterized by compression of the third, or transverse, portion of the duodenum against the aorta by the SMA, resulting in chronic, intermittent, or acute, complete or partial duodenal obstruction [1]. The precise incidence of this entity is unknown. In a review of literature, approximately 0.013–0.3% of the findings from upper gastrointestinal tract barium studies support a diagnosis of SMA syndrome [3]. More females are affected by SMA syndrome. The SMA usually forms an angle of approximately 45° (range $38\text{--}56^\circ$) with the abdominal aorta, and the third part of the duodenum crosses posteroinferiorly to the origin

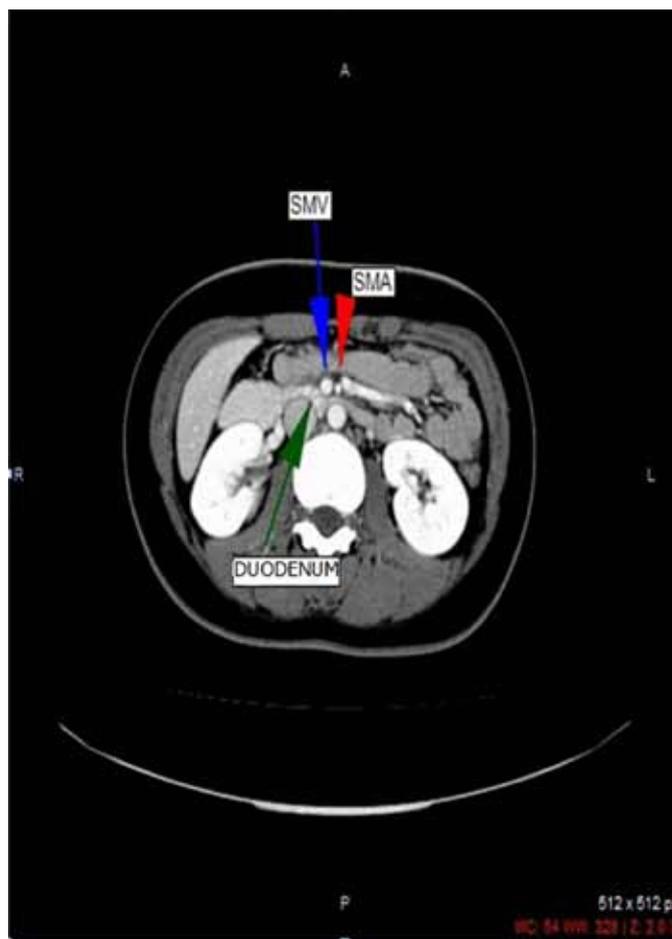


Figure 1: Computed tomography scan of the abdomen showing compression of the third part of the duodenum between the abdominal aorta and superior mesenteric artery.

of the SMA, coursing between the SMA and aorta [1, 4, 5]. Any factor that sharply narrows the aortomesenteric angle to approximately $6\text{--}25^\circ$ can cause entrapment and compression of the third part of the duodenum as it passes between the SMA and aorta, resulting in SMA syndrome. In addition, the aortomesenteric distance in SMA syndrome is decreased to 2–8 mm (normal is 10–20 mm). Important aetiologic factors that may precipitate narrowing of the aortomesenteric angle and recurrent mechanical obstruction include, thin body build, exaggerated lumbar lordosis, visceroptosis and abdominal wall laxity, depletion of the mesenteric fat caused by rapid severe weight loss due to catabolic states such as cancer, surgery, burns, or psychiatric problems. Severe injuries, such as head trauma, spinal disease, deformity, or trauma leading to prolonged bedrest, dietary disorders such as anorexia nervosa and malabsorption may cause loss of fat [4, 6]. Our patient was having symptoms of anorexia nervosa and was treated for it and later on due to severe weight loss she developed SMA syndrome.

Patients with SMA syndrome may present symptoms of gastrointestinal obstruction, such as upper abdominal distension and epigastric tenderness, usually relieved



Figure 2: The low aorto-mesenteric angle on magnetic resonance angiography.

by posture changing. The acute presentation is usually characterized by signs and symptoms of duodenal obstruction. Chronic cases may present with long-standing vague abdominal symptoms, early satiety and anorexia, or recurrent episodes of abdominal pain, associated with vomiting. Delay in the diagnosis of SMA syndrome can result in malnutrition, dehydration, electrolyte abnormalities, and even death. Our patient was having chronic symptoms.

The differential diagnosis includes anorexia nervosa and bulimia. In addition, SMA syndrome should be differentiated from other causes of megaduodenum, including diabetes mellitus, collagen vascular conditions, and chronic idiopathic intestinal pseudoobstruction. The diagnosis of SMA syndrome is difficult. The diagnosis of SMA syndrome should be considered in patients with rapid weight loss who develop atypical, recurrent obstructive symptoms not to other common causes. Confirmation usually requires radiographic studies, such as an upper gastrointestinal endoscopy, hypotonic duodenography, and CT scan. Contrast-enhanced CT scan and magnetic resonance angiography (MRA) is useful in the diagnosis of SMA syndrome and can provide diagnostic information, including aorta-SMA distances and duodenal distension [4]. Also, it can be used to assess intra-abdominal and retroperitoneal fat. Both these procedures are noninvasive and are probably equivalent

to angiography, which has previously been suggested as the reference standard for establishing the diagnosis [5]. Computed tomography scan was the diagnostic in our patient as it revealed the narrow aortomesenteric angle and compression of third part of duodenum. Upper gastrointestinal study with barium revealed characteristic dilatation of the first and second parts of the duodenum, with an abrupt vertical or linear cut-off in the third part with normal mucosal folds. Once radiologic studies established diagnosis, first-line treatment is usually conservative with jejunal or parenteral nutrition for restoration of the aortomesenteric fatty tissue. Conservative initial treatment is recommended in all patients with SMA syndrome. This includes adequate nutrition, nasogastric decompression, and proper positioning of the patient after eating (i.e., left lateral decubitus, prone, or knee-to-chest position) [2, 5, 7, 8]. Enteral feeding using a double lumen nasojejunal tube passed distal to the obstruction under fluoroscopic assistance is an effective adjunct in treatment of patients with rapid severe weight loss and also eliminates the need for intravenous fluids and the risks associated with total parenteral nutrition [2]. Our patient's symptoms had been resolved by nutritional support without any surgical treatment. Total parenteral nutrition (TPN) had revealed all the symptoms and we did not need any surgical interventions. Total parenteral nutrition is not definitive treatment for Wilkie's syndrome.

Surgical intervention is indicated when conservative measures are ineffective, particularly in patients with a long history of progressive weight loss, pronounced duodenal dilatation with stasis, and complicating peptic ulcer disease [1, 2, 9]. Duodenojejunostomy is the most frequently used procedure, and it is successful in about 90% of cases. The use of laparoscopic surgery that involves lysis of the ligament of Treitz and mobilization of the duodenum has been reported [7]. Our patient was responded to the conservative management. The only medical treatment was administered TPN without any surgical treatment. She gained 4 kg before discharge and she has been following without any complication and complain for one year.

CONCLUSION

The treatment of Wilkie's syndrome is aimed at the precipitating factor, which usually is related to weight loss. Therefore, conservative therapy with nutritional supplementation is the initial approach, and surgery is reserved for those who do not respond to hyperalimentation. Its recognition is important because early diagnosis of a partial obstruction may allow for medical rather than surgical intervention, as exemplified by our case. The diagnostic challenges of identifying vascular constriction between the aorta and superior mesenteric artery have been answered by advances in the field of computed tomography. Despite diagnostic

confusion with intestinal dysmotility syndrome, conservative therapy with nutritional supplementation is the initial approach and duodenojejunostomy is favored, if non-surgical treatment fails.

Author Contributions

Ali Coşkun – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Deniz Uçar – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Erdem Barış Carti – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Erkan Oymaci – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Mehmet Yıldırım – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Nazif Erkan – Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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Article citation: Coşkun A, Uçar D, Carti EB, Oymaci E, Yıldırım M, Erkan N. Wilkie's syndrome: A rare cause of vomiting and weight loss. *International Journal of Case Reports and Images* 2013;5(2):150–154.



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