Wilkie’s syndrome: Review of eight cases

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Acta Gastroenterol Latinoam 2013;43:240-247

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Summary
Background. The superior mesenteric artery (SMA) syndrome is a rare entity presenting with upper gastrointestinal tract obstruction and abrupt weight loss. Usually seen in tall individual with low body mass index (BMI), the weight loss may be primary or secondary to a systemic disease or to the syndrome itself. Compression of the duodenum between the abdominal aorta and the superior mesenteric artery due to loss of the duodenal pad of fat causes the obstruction. Early recognition prevent complications. Imaging and endoscopy is diagnostic. Aorto-mesenteric angle and SMA aorta distance is lower than 22 degree and eight millimeter respectively. If nutritional support fails, surgery is required (e.g. jejunocolostomy). Studies to determine the optimal methods of diagnosis and treatment, especially in a suburban hospitals with limited expertise are essential.

Case report. We retrospectively evaluated 8 cases presented over three years. Six (75%) patients were males, age ranged from 19 to 70 years and weight from 40 to 55 kg. The mean BMI was 18.7 kg/m² (range 16.42 to 25.11 kg/m²). Mean weight loss before diagnosis was 9.88 kg (range 6 to 12 kg). Symptoms developed between 8 to 180 days (median 12 days). Commonest presentation was epigastric pain, vomiting and nausea. Four patients had pre-morbid conditions and the syndrome was idiopathic in the other 4. Median aorto-mesentric angle was 16.5 degree and SMA-aorta distance was 5.15 mm. Four patients required operation. The rest improved on conservative treatment.

Conclusion. Early recognition, institution of the appropriate conservative measures and timely selection of a definite surgical method are critical to prevent severe complications and death.

Key words. Superior mesenteric artery syndrome, duodenal obstruction, Wilkie’s syndrome.

Síndrome de Wilkie: Revisión de ocho casos

Resumen
Antecedentes. El síndrome de la arteria mesentérica superior (AMS) es una entidad poco frecuente que presenta obstrucción del tracto gastrointestinal superior y pérdida de peso brusco. Por lo general se ve en personas altas con un índice de masa corporal (IMC) bajo y la pérdida de peso puede ser primaria o secundaria a enfermedades sistémicas o al síndrome en sí mismo. La compresión del duodeno entre la aorta abdominal y la arteria mesentérica superior debido a la pérdida de la almohadilla de grasa duodenal provoca la obstrucción. El reconocimiento temprano permite prevenir las complicaciones. Las imágenes y la endoscopía son diagnósticas. El ángulo aorto-mesentérico y la distancia aorta-AMS es inferior a 22 grados y 8 mm, respectivamente. Si falla el soporte nutricional, se requiere la cirugía (por ejemplo, duodenoyeyunostomía). Son esenciales los estudios para determinar los métodos eficaces de diagnóstico y tratamiento, especialmente en algunos hospitales suburbanos con experiencia limitada.

Presentación de los casos. Evaluamos retrospectivamente 8 casos que se presentaron durante tres años. Seis (75%) pacientes eran varones. La edad osciló entre 19 y 70 años y el peso entre 40 y 55 kg. El IMC medio fue 18.7 kg/m² (rango 16.42 a 25.11 kg/m²). La media de pérdida de peso antes del diagnóstico fue 9.88 kg (rango 6 a 12 kg). Los síntomas se desarrollaron entre 8 y 180 días (mediana 12 días). La presentación más común fue dolor epigástrico, vómitos y náuseas. Cuatro pacientes tenían condiciones pre-mórbidas y en 4 el síndrome era idiopático. La mediana del ángulo aorto-mesentérico fue 16.5 grados y la distancia AMS-aorta 5.15 mm. Cuatro pacientes requirieron operación. El resto mejoró con tratamiento conservador.

Conclusión. La detección precoz, la institución de las medidas conservadoras adecuadas y la selección oportuna de un método quirúrgico definitivo son fundamentales para prevenir las complicaciones graves y la muerte.

Palabras claves. Síndrome de la arteria mesentérica superior, obstrucción duodenal, síndrome de Wilkie.
The superior mesenteric artery (SMA) syndrome is an unusual form of gastrointestinal obstruction. Usually it presents with symptoms of mid to upper gastrointestinal obstruction due to compression of the duodenum between the abdominal aorta posteriorly and the SMA anteriorly. This occurs due to the loss of the duodenal pad of fat that cushions, holding the SMA off the spine and protecting the duodenum from compression.

First described by von Rokitansky in 1842, Wilkie formally characterized the SMA syndrome in a series of 75 patients in 1927. Classically there is a history of acute weight loss preceding the typical symptoms of chronic intermittent abdominal pain, vomiting (occasionally biliious), early satiety and anorexia.

Diagnosis is usually confirmed by barium meal X-ray supplemented by endoscopy, computed tomography (CT), doppler ultrasound (US) and magnetic resonance angiography (MRA). A conservative treatment with an enhanced nutritional program is tried and surgical correction is attempted when it fails. In some cases diagnosis is confirmed only at laparotomy.

It was observed that most of the comprehensive studies and case series dated back to 1960–1980, while recent studies were majorly limited to case-reports. This manuscript presents a series of 8 patients with SMA syndrome treated at a single institution over a duration of 36 months, being one of the largest series reported in over 30 years, after Merret et al. We have included a literature review to raise awareness of the condition. The aim was to analyze the clinical presentation, the diagnosis and the management of SMA syndrome in adults, especially in a suburban hospital.

Case report

This retrospective evaluation was performed after obtaining authorization from our institutional review board and informed consent was not required for individual patients. The extracted data included demographics, presenting symptoms and their duration, past medical and surgical records, examination findings, methods of diagnosis, treatment and outcome. We tabulated them in a Microsoft Excel spreadsheet and used standard analysis. Long-term follow up was obtained in all patients.

Between August 2007 and July 2010, 8 cases were analyzed. One of the patients was referred from outside the normal drainage area. Six patients (75%) were males. The age range was 19 to 70 years (median 42.5 years) and the weight range 40 to 55 kg (median 48 kg). The mean body mass index (BMI) was 18.7 ± 2.71 kg/m² (range 16.42 to 25.11 kg/m²). The mean weight loss before diagnosis was 12.37 ± 4.9 kg (range 6 to 20 kg). Unlike some large series where few patients did not have any change in weight, all our patients had considerable weight loss. Seven patients (87.5%) lost more than 10% of their body weight over a median of 135 days.

Symptoms developed between 8 to 180 days (median 12 days), with 6 cases presenting acutely. Usual presentation included multiple symptoms: pain, nausea, vomiting, early satiety and anorexia. The most frequent constellation was epigastric pain, vomiting and nausea, occurring in 50% of patients, with or without early satiety. The vomit contained bile-stained material and was triggered by eating and drinking in all patients. In 2 patients vomiting was associated with significant electrolyte abnormalities, particularly hypokalemia requiring intravenous replacement. Esophageal reflux was absent in all patients.

Four patients had pre-morbid conditions. One patient had abdominal tuberculosis with enlarged abdominal lymph nodes and 3 had a recently diagnosed gastric carcinoma, non-obstructing and resulting in severe weight loss that reduced the aorto-mesentric angle. Four patients (50%) had idiopathic SMA syndrome. One patient had a previous gastrojejunostomy. Eating disorders, psychiatric problems, drug abuse or endocrinopathies were absent in all patients.

US and straight X-ray of the abdomen were done as the first imaging investigations in all patients (Figures 1 and 2). X-ray showed a dilated fluid-gas filled stomach and diminished distal bowel gas in all the cases. US supplemented by color doppler when deemed
appropriate suggested the diagnosis in 4 (50%) cases. Furthermore, US suggested mass lesions in 50% of patients, which was investigated by contrast enhanced CT scan. CT, performed in 6 cases, showed gastric carcinoma in 3 patients, abdominal tuberculosis in 1, and SMA syndrome in all. In the 4 cases of idiopathic SMA syndrome, barium meal X-ray was done and was positive (Figure 3). Of note, diagnosis was missed by US in 2 of these 4 cases and achieved by barium studies. Upper gastrointestinal endoscopy was done in 5 cases. In 3 patients endoscopy revealed the non-obstructive gastric cancer which was the cause of cachexia leading to SMA syndrome. Two patients underwent upper gastrointestinal endoscopy after non-confirmatory US but the study did not contribute to the diagnosis. As these 2 patients were severely symptomatic, we immediately performed an exploratory laparotomy and diagnosis was confirmed intra-operatively. In the remaining cases, CT, X-rays and US were enough for the diagnosis and endoscopy was not required. None of the patients showed signs of reflux or gastritis in endoscopy. In spite of the marked weight loss, all patients had serum albumin and proteins within the normal limits. Two patients had electrolyte abnormalities.

We advised transpyloric jejunal feeding past the point of obstruction (via gastrojejunostomy tube or nasojejunal tube) in the 6 patients with a pre-operative diagnosis. Conservative management was successful in four patients, namely those with tuberculous mesenteric lymphadenitis and carcinoma. In these cases antitubercular drugs (ATD) and neo-adjuvant chemotherapy, respectively, along with nutritional supplementation, alleviated the symptoms. However, 2 of these cases failed to respond after three weeks of conservative therapy and had to undergo surgery. Two patients had a relatively acute presentation and an exploratory laparotomy was performed without further delay.

Four patients were operated upon. We performed a retrocolic side-to-side duodenojejunostomy in 2 patients and a Roux-en-y duodenojejunostomy in 2. One patient in the first group initially had a gastrojejunostomy but failed to improve after the operation due to persistent pathology. A revision surgery with retrocolic side to side duodenojejunostomy without disturbing the loop gastrojejunostomy was done three weeks after the first procedure. After surgery, recovery was complete and uneventful in all patients. Mean length of hospitalization was 37.5 days (range 14 to 90 days). Mean duration of follow up was 12.38 months. All patients had an excellent clinical outcome gaining between 5 and 15 kg at a six-month review. The patient’s weight gain stabilized at the 6-month period with no patient recording any major weight gain after this period. The patient with tuberculous lymphadenopathy completely responded to ATD and those with malignancy improved symptomatically on chemotherapy. Two of the cancer patients required subsequent surgery and the third was managed with palliative support.

Patient characteristics are described in Table 1.
Table 1. Patient Characteristics

<table>
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<th>Case (number)</th>
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<th>2</th>
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<td>Weight loss [%]</td>
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<td>10 (16.4)</td>
<td>6 (9.8)</td>
<td>12 (23.1)</td>
<td>10 (17.2)</td>
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<td>A M angle (°)</td>
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<td>15</td>
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<td>Non-op</td>
<td>Non-op</td>
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<td>41</td>
<td>32</td>
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Discussion

Although somewhat controversial, mechanical compression of the duodenum between the SMA and aorta has been postulated as the basic pathogenesis of SMA syndrome. There are underlying predisposing factors that contribute to the final common pathway in which perimesenteric and retroperitoneal fat is depleted. These predisposing factors include wasting diseases (burns, cancer, endocrine disorders), severe injuries (head or spinal trauma, application of a body cast), dietary disorders (anorexia nervosa, malabsorptive diseases), and postoperative states (treatment for scoliosis, total proctocolectomy and ileal pouch anastomosis). Additionally, aortomesenteric narrowing can occur owing to extrinsic compression from a mass, abnormally high fixation of the ligament of Treitz, increased lordosis or low origin of the SMA. Choi and Pfalzer suggested that the course of the SMA running anterior and parallel to the aorta, and not the narrow aorto-mesenteric angle and the distance alone, is the critical factor. Others have refuted the theory of vascular compression and suggested that the compression of the duodenum is caused by peritoneal adhesions which are a result of duodenal malrotation or inflammatory thickening of the mesenteric root secondary to acute pancreatitis, duodenal ulcer or infarction of the bowel. The SMA syndrome should be differentiated from the SMA-like-syndrome (mega-duodenum), found in several neuropathic and connective tissue disorders. The majority of published studies of this condition is limited to case reports and small series.

In the general population, the prevalence of SMA syndrome was estimated in around 0.013% to 0.3% based on upper gastrointestinal barium studies, underlining its rarity. Most single institution studies reported an average incidence of one case per year. Our tertiary referral center, with a local drainage population of 6 million people and 8 cases in three years, has a higher incidence rate. We emphasize the importance of identifying those patients at risk and maintaining a high index of suspicion. Paying attention to the various risk factors that may be present before surgery may be beneficial to the understanding and treatment of SMA syndrome. According to most previous studies, the SMA syndrome is more common in females and young adults between 10 and 39 years of age. However, our study depicted a higher prevalence among males with only 37.5% of the patients falling in the above-mentioned age group. This may be a plausible error due to the small sample size or because a high proportion of the cases had SMA syndrome secondary to systemic causes like gastric carcinoma, which is prevalent among elderly males. Of course, the affected age and sex are closely linked to the underlying predisposing condition. The SMA syndrome is typically found in slender persons with acute weight loss. Munns et al identified high-risk patients as “those with a thin, asthenic habitus”. The average BMI of an adult Indian is 22 to 23 kg/m² (male: 21.8 kg/m², female: 22.7 kg/m²). In our sample, pre-symptomatic mean BMI was 22 kg/m² but due to se-
were weight loss in all patients, mean BMI became 18 kg/m², thus producing significant symptoms. Our results do not support the findings of Trisiskos et al who reported the association between pre-symptomatic low BMI and SMA syndrome.29 However, all patients underwent significant weight loss thus confirming the findings of Munns. In 50% of the patients, weight loss was due to co-morbid conditions like carcinoma and tuberculosis. Weight loss could not be explained in the rest of the patients. In idiopathic cases, it was not clear whether the weight loss was due to SMA syndrome or vice-versa. In our series, all patients who developed SMA syndrome were also disproportionately taller. The height was above the mean of a normal population with the percentile ranging from above 50 to above 90. The weight was below the mean of a normal population with the weight percentile ranging from 5% to 25% after weight loss. Our findings agree with previous studies showing that the height percentile higher than 50% and the weight percentile lower than 25% (after weight loss) might be potential risk indicators for SMA syndrome, but we did not find that BMI previous to weight loss was below normal or that patients were disproportionately “thin” by habitus.30

SMA syndrome can present in both acute and chronic forms, although prevalence in a chronic-care hospital (0.965/1,000 admissions) was significantly higher than that in acute general hospitals (0.0108-0.0520/1,000 admissions).31 Similar to previous publications, 75% of the patients in this series presented acutely with a median duration of symptoms of only 12 days.32,33 However, on scrutiny, it was found that weight loss and milder forms of the symptoms were present for much longer duration with a median of 135 days. The rural catchment area with less health awareness may explain the phenomenon. Acutely, patients usually present with symptoms and signs of duodenal obstruction, namely nausea, vomiting, abdominal pain, distension, tenderness and abnormal bowel sounds. These symptoms are aggravated by eating. Manifestations of electrolyte imbalance may occur.9,10,23,24,34 All our patients presented with typical symptoms. Pain however, was not relieved by prone, knee-chest or left lateral decubitus position in any case, as documented by Yang.35 The chronic cases may present many times, over many years, for investigation of intermittent abdominal pain associated with vomiting, early satiety and anorexia.36 Food intolerance promotes ongoing weight loss, which may further reduce intraabdominal adipose tissue and exacerbate the problem, resulting in a vicious cycle.20,23,36

Upper gastrointestinal series, CT scans, MRA, conventional angiography, ultrasonography and endoscopy have all been used for diagnosis.7,26,37,40 Conventional barium studies play an important role in the diagnosis of SMA syndrome according to radiographic criteria described by Hines et al.19,41 However, as suggested by few recent studies, this study revealed that US supplemented by color Doppler suggested the diagnosis in 62.5% of cases.7,42 As it is a commonly available cheap investigation, frequently done in the emergency setting, we performed US in all patients with upper gastrointestinal symptoms. Santer et al and Applegate et al were the first researchers who introduced CT findings for the syndrome.43 CT scan is rapid, safe, and relatively non-invasive, and has become one of the preferred investigations, which is considered as a reference standard for establishing the diagnosis.8,44-46 The advantage of CT over US is that it can provide an overall assessment of the abdominal cavity as well as the amount of intra-abdominal and retroperitoneal fat. As suggested by the literature, upper gastrointestinal endoscopy confirmed underlying pathology and ruled out peptic ulcer disease that might be secondary to reflux or constitute a primary pathology mimicking SMA syndrome.9 It also revealed that in the three cases of carcinoma of the stomach, the antral growth did not result in gastric outlet obstruction. However, endoscopy could not suggest SMA syndrome in patients presenting with symptoms of upper gastrointestinal obstruction.34 Two patients (25%) were diagnosed at surgery, one for the malfunction of a recently placed gastrojejunostomy and the other after abdominal US and esophago-gastro-duodenoscopy failed to reveal the actual pathology.

The third portion of the duodenum is retroperitoneal and runs between the ventrally oriented SMA and the aorta. The vascular abnormalities are well delineated by fine slice CT imaging with vascular reconstruction, US and MRA, allowing to measure the aorto-superior mesenteric artery angle. The normal aorto-mesenteric angle ranges from 38° to 56°, with an SMA-aorta distance of 10 to 28 mm, where the third duodenum crosses the aorta.17,38 Unal et al reported cutoff values of 22° (42.8% sensitivity and 100% specificity) for the aorto-mesenteric angle and 8 mm (100% sensitivity and specificity) for at least one symptom of SMA syndrome.40 In the present study, the median value of aorto-mesenteric angle was 16.5° (range 12 to 22°) and the SMA-aorta distance was 5.15 mm (range 4.4 to 6.4 mm) which is consistent with the above mentioned studies.

Treatment of SMAS is usually conservative. Acute management is focused on bowel decompression, main-
tenance of the fluid and electrolyte balance and nutritional rehabilitation. Once stabilized small frequent high caloric oral feeds, changes in feeding position (knees to chest or right lateral decubitus) help in bypassing the obstruction. If symptoms persist, a nasojejunal tube placed past the obstruction to allow for continuous enteral feeding may be required. Parenteral nutrition is useful when enteral feedings are not tolerated. In children and in adults with a short history, this may have a reasonable prospect of success; but in the chronically symptomatic adult patients, conservative treatment is often a prolonged in-hospital therapy with a low success rate and frequently requires surgery. In the present study, all the patients diagnosed pre-operatively were started on medical treatment, but only those four patients with an underlying cause responded. Symptoms persisted in those with idiopathic SMA syndrome and those who presented with chronic symptoms. Jejunal feeding was the most frequent intervention. Oral diet was not tolerated and facility for parenteral therapy was unavailable. All patients required hospital admission. Main surgical options include Strong’s procedure, duodenojejunostomy with or without division of the fourth part of the duodenum, and gastrojejunostomy. Duodenojejunostomy as a treatment for this condition was first described by Stavely in 1908 and has become the preferred surgical procedure by most surgeons, with a success rate of 90%. However, duodenojejunostomy without division of the fourth part of the duodenum carries a risk of blind loop syndrome. Gastrojejunostomy allows gastric decompression but the failure to relieve the duodenal obstruction may result in recurrent symptoms requiring a second procedure. Also, the unrelieved obstruction may result in dumping syndrome, blind loop syndrome, gastric bile reflux and ulceration. Strong’s procedure (lysis of ligament of Treitz) has the advantage of maintaining bowel integrity, but has a failure rate of 25%, presumably due to short branches of the inferior pancreatic-duodenal artery that do not allow the duodenum to fall inferiorly.

Laparoscopic Strong procedures and laparoscopic duodenojejunostomy have also been reported by several centers. World’s first robotically assisted intestinal bypass surgery for SMA syndrome was reported in 2008 at London Health Services Centre. Kudo et al reported the anterior transposition of the third part of the duodenum. The risk of recurrence because of inadequate drainage of a grossly dilated duodenum is eliminated. Some authors have promoted the resection of the abnormal duodenum rather than bypassing the duodenal third part, as they have postulated that SMA syndrome is a variant of a motility disorder rather than a true mechanical obstruction. However, there is little evidence to support this postulate. We performed duodenojejunostomy without mobilization and division of the fourth part of the duodenum. Fortunately, there were no evidences of a blind-loop syndrome in any of these case. As stated before, one of them (who had gastrojejunostomy as the first procedure) had a revision surgery as symptoms persisted after initial operation. Usually encirclage operation is recommended in such cases but we avoided the gastric resection and made an additional duodenojejunoanostomy along with the already done gastrojejunostomy. To date, there are no data to guide us to an optimal period or indications for preoperative or postoperative nutritional support either by enteral or by total parenteral nutrition (TPN). Although it is well recognized that significant preoperative weight loss is associated with an increased risk of postoperative complications, all of our patients had uneventful recoveries. No patient required TPN preoperatively. We infer that, in the presence of normal serum proteins, surgery may be safely performed with preoperative nutritional supplementation, especially TPN being used selectively on an individualized basis. Similarly, the possibility of refeeding syndrome in the postoperative period may need to be considered, but it did not occur in our series.

Complications of SMA syndrome include dehydration, metabolic imbalance, gastroparesis after corrective surgery, persistent vomiting causing esophageal tear, and spontaneous bleeding of the gastrointestinal tract. Respiratory distress syndrome and gastric perforations have also been described. Rarely death can occur. Most of the reported mortality involved patients in whom the diagnosis was markedly delayed or completely missed. With treatment, all the patients in this series showed gradual but uneventful recovery. This study is consistent with other reports supporting the favorable outcome of SMA syndrome.

We conclude that the SMA syndrome is a very rare entity associated with a wide range of predisposing factors, presenting with features of upper gastrointestinal tract obstruction and recent abrupt weight loss. Early recognition of the condition, appropriate conservative measures and proper timely selection of a definite surgical method are critical to prevent the development of severe complications and death. However, profound studies are still needed to better define the optimal methods of diagnosis and treatment of this condition.

No funding was received for this work.
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