Nascer e Crescer - Birth and Growth Medical Journal
2020;30(3): 171-174. doi:10.25753/BirthGrowthMJ.v30.i3.18823

CASE REPORTS

WEIGHT LOSS AS A CAUSE OF SUPERIOR MESENTERIC ARTERY SYNDROME

PERDA PONDERAL COMO CAUSA DE SÍNDROME DA ARTÉRIA MESENTÉRICA SUPERIOR

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ABSTRACT

Introduction and objective: Superior mesenteric artery syndrome (SMAS) is a rare condition in pediatric age, often presenting with non-specific gastrointestinal symptoms. The aim of this report was to highlight the importance of considering/excluding this diagnosis.

Case description: A 17-year-old female presented with slowly progressing epigastralgia, heartburn, early satiety, nausea and sporadic vomiting with one and a half years of evolution. She had lost 13% of her weight in the previous six months. On physical examination, the girl complained of pain on epigastrium palpation, with no further changes. Laboratory tests were normal. Esophageal, gastric and duodenal transit (EGDT) revealed a vertically elongated stomach, with the greater curvature projecting towards the pelvic cavity and a slight delay in gastric emptying. Abdominal computed tomography scan confirmed the diagnosis of SMAS and a conservative approach with a hypercaloric fractionated diet was initiated. The girl maintained multidisciplinary follow-up (Nutrition, Pediatrics and Pedopsychiatry), with marked improvement and without requiring surgical intervention.

Comments: Although rare, SMAS should be considered after exclusion of the most frequent underlying causes of persistent non-specific gastrointestinal symptoms.

Keywords: superior mesenteric artery; syndrome; weight loss

RESUMO

Introdução e objetivo: A síndrome da artéria mesentérica superior (SAMS) é uma patologia rara em idade pediátrica, frequentemente com sintomatologia gastrointestinal inespecífica. Este caso pretende alertar para a importância de considerar/excluir este diagnóstico.

Descrição do caso: Uma adolescente de 17 anos recorreu ao Serviço de Urgência com um quadro de epigastralgia, pirose, enfartamento, náuseas e vómitos esporádicos com um ano e meio de evolução. A adolescente tinha registado uma perda ponderal de 13% do peso nos últimos seis meses e apresentava dor à palpação do epigastro no exame objetivo, sem outras alterações. O estudo analítico era normal. O trânsito esofago-gastroduodenal revelou um estômago vertical alongado com a grande curvatura projetando-se para a cavidade pélvica e atraso no esvaziamento gástrico. Por suspeita de SAMS, foi efetuada tomografia computorizada abdominal, que confirmou o diagnóstico. Foi instituída uma abordagem conservadora, através de uma dieta hipercalórica fracionada e acompanhamento por nutricionista, pediatra e pedopsiquiatra, tendo a adolescente apresentado franca melhoria, sem necessidade de intervenção cirúrgica.

Comentários: Apesar de rara, a SAMS deve ser ponderada após exclusão das causas orgânicas mais frequentes associadas a sintomas gastrointestinais persistentes.

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Palavras-chave: artéria mesentérica superior; perda ponderal; síndrome

INTRODUCTION

Superior mesenteric artery syndrome (SMAS), also known as Wilkie's syndrome, was first described by Von Ronkitanski in 1861.[1-3] It is characterized by compression, with partial or total obstruction, of the third part of the duodenum by the superior mesenteric artery (SMA) (anterior) and the abdominal aorta (posterior). In most cases, the aortomesenteric angle is reduced below 25° (normal values lie between 38° and 65°) and the aortomesenteric distance below 8 mm (normal values lie between 10 and 28 mm) as the result of upper intestine obstruction.[1,2]

This condition is infrequent in pediatric age, with an incidence varying between 0.013 and 0.3%, being more frequent in female patients.[3] Significant weight loss is the most common cause, but it may also manifest through congenital or acquired anatomical variations.[1-5]

A conservative approach should be initially undertaken to alleviate symptoms and correct triggering factors, but surgical therapy may prove necessary in more severe or refractory cases.[1,2,4]

Herein is described the clinical case of a female adolescent with recurrent gastric and intestinal symptoms related to compression of the aortomesenteric angle and highlighted the importance of considering SMAS diagnosis after exclusion of the most frequent causes of these symptoms.

CASE PRESENTATION

A 17-year-old female with a medical history of asthma presented to the Emergency Department with epigastric pain, early satiety, heartburn, nausea and sporadic vomiting with approximately one and a half years of evolution and worsening in the three months before admission. Family history was relevant for positive *Helicobacter pylori* test in a paternal uncle and colorectal carcinoma diagnosed at the age of 45 in a paternal aunt. At presentation, anorexia and weight loss of 13% in the previous six months were noted. The clinical picture was initially mostly postprandial, with complaint relief in the genupectoral position. The girl was medicated with esomeprazole, without improvement. The diagnostic study included esophageal, gastric and duodenal transit (EGDT), which revealed a vertically elongated stomach with the greater curvature projecting towards the pelvic cavity, as well as a slight delay in gastric emptying (**Figure 1**).

The patient underwent upper endoscopy, which revealed a small sliding hiatal hernia without other macroscopic or histological changes, and a *Helicobacter Pylori* test, which was negative. Laboratory tests (hemogram, coagulation tests, and renal, liver, and pancreatic function tests) were normal. This study ruled out common diseases like esophagitis, peptic ulcers, *Helicobacter Pylori* gastritis, achalasia, pancreatitis and cholecystitis.

Upon admission and objective examination, the girl weighed 50 kg (25th percentile) and displayed good overall condition and no signs of dehydration or malnutrition folds. Signs of peritoneal irritation were absent and no organomegaly or masses were present, but palpation of the epigastric region was painful. The remaining examination revealed no relevant changes.

The patient was admitted to the Pediatric Ward for further study and medicated with esomeprazole (due to absence of a diagnosis and short treatment period) and erythromycin (as a prokinetic agent), due to the delay in gastric emptying and SAMS suspicion, with clear symptom improvement.

An abdominal Doppler ultrasound was performed to exclude other causes of gastric or intestinal obstruction, which was inconclusive regarding SAMS but helpful in excluding intestinal malrotation. Contrast computerized tomography (CT) scan of the abdomen was performed, revealing a 4-mm distance between the superior mesenteric artery and the aorta (**Figure 2**) and an aortomesenteric angle of 12.2° (**Figure 3**), confirming the diagnosis of SAMS. A hypercaloric fractionated diet adjusted to the patient's tolerability with regular Nutrition and Pediatrics follow-up was undertaken, enabling recovery of the lost weight. This conservative approach aims to recover the lost fat pad supporting the SMA, preventing it from falling and compressing the third portion of the duodenum. As the girl displayed significant anxiety, Pedopsychiatry consultation was also required and she was medicated with sertraline, with symptom improvement.

Six months after discharge, the girl has gained 6.5 kg (equivalent to the weight lost in the last six months) and remained asymptomatic. Control abdominal CT scan revealed an increase of the aortomesenteric angle, now at 25° (**Figure 4**).

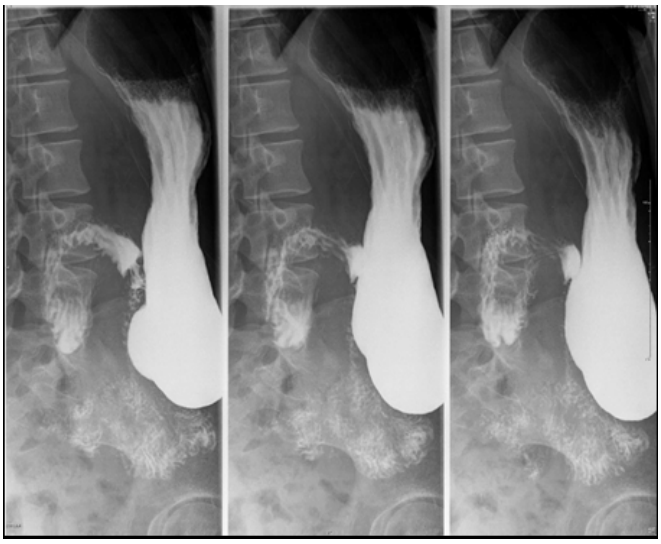

Figure 1 - Esophageal, gastric, and duodenal transit evidencing delayed gastric emptying, affecting the shape of the third part of the duodenum.

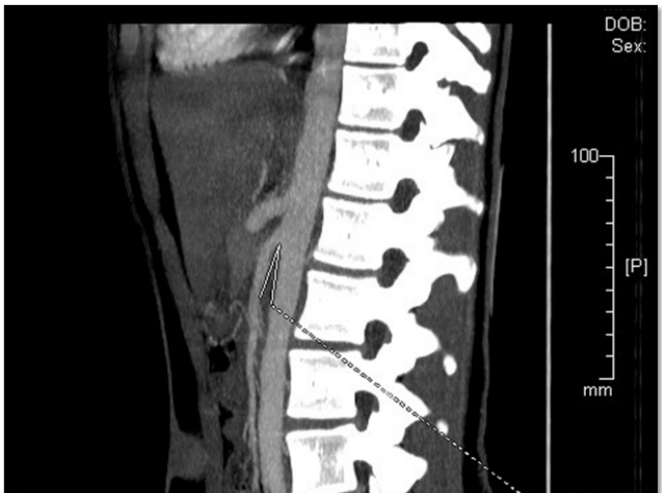

Figure 3 - Abdominal CT scan (profile) revealing a 12.2° angle between the aorta and the superior mesenteric artery.

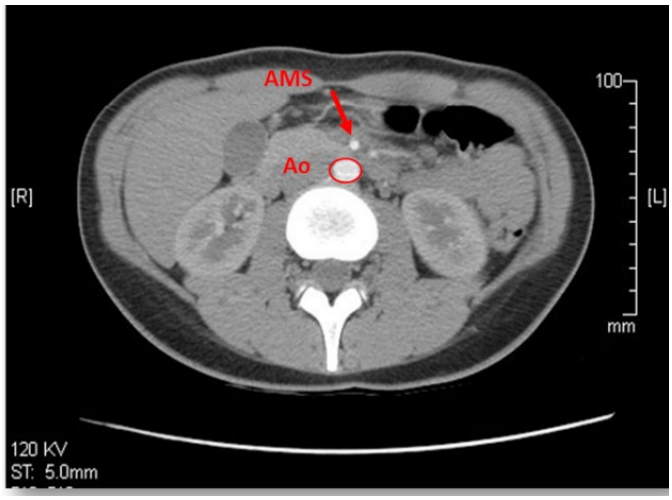

Figure 2 - Abdominal CT scan (axial cut) revealing a shortened distance (4 mm) between the aorta (Ao) and the superior mesenteric artery (SMA).

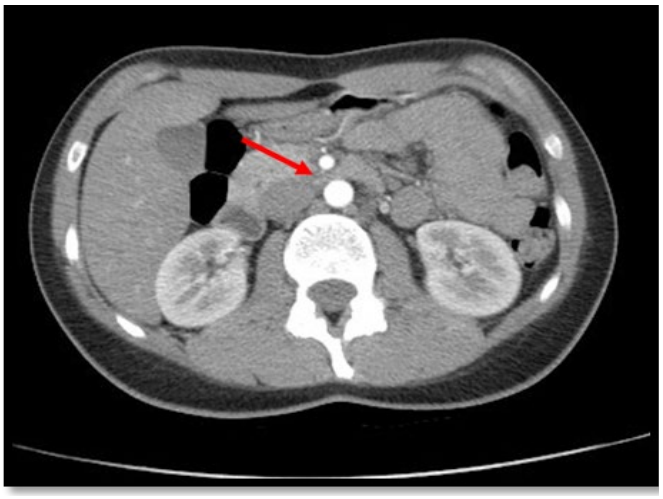

Figure 4 - Control abdominal CT scan, showing an increase in the aortomesenteric angle, now at 25°.

DISCUSSION/CONCLUSION

In Pediatrics, recurrent abdominal pain, among other gastric and intestinal symptoms, are frequent Emergency Department presentations. The high number of differential diagnoses poses a challenge for establishing the correct diagnosis.

In the present case, weight loss associated with slowly progressing symptoms and postprandial prevalence motivated the referral to General Pediatrics clinic and follow-up. The diagnostic study began with laboratory tests and contrast radiography (EGDT), which, despite low sensitivity, may direct the subsequent clinical management if showing, for example, dilation of the proximal duodenum or delayed gastric emptying.[4] In this case, EGDT was abnormal, with dilation of the distal portion of the second duodenal portion and absence of contrast in the third duodenal portion, suggesting an obstruction at this level.

As symptoms persisted, an upper digestive endoscopy was performed, which excluded esophageal and gastric lesions. Despite

being operator- dependent, abdominal ultrasound may potentially identify the underlying causes associated with gastrointestinal symptoms, evaluate SMA anatomy, and identify anatomical variations of the aortomesenteric angle.[3,4,6-8] However, in this case it was also inconclusive, motivating the subsequent abdominal CT scan. As SMAS was suspected, the aortomesenteric angle and distance were assessed (12.2° and 4 mm, respectively), enabling to establish the diagnosis according to imaging criteria and clinical data.

Although several invasive exams were performed in this case, excluding other common underlying causes (esophagitis, peptic ulcers, cholecystitis, pancreatitis, etc.), the diagnosis remained inconclusive. Only CT scan provided an unambiguous assessment of the relevant anatomy.

The patient's favorable evolution was possible through a multidisciplinary approach including anxiety disorder management, resulting in weight recovery and progressive improvement (including lasting symptom reversal) and precluding the need for surgical treatment. The significant weight loss observed probably constrained the aortomesenteric angle decrease, due to loss of protective and/or cushioning adipose tissue. By managing the anxiety disorder and introducing a hypercaloric diet, weight recovery was accomplished within six months. The marked clinical picture improvement was objectively confirmed through a control CT scan performed eight months later, which showed an increase in both the aortomesenteric angle and distance.

SMAS requires a high index of suspicion and is, therefore, a challenging diagnosis. Delayed diagnosis may lead to complications, such as acute gastric dilatation, intestinal pneumatosis, and electrolyte imbalances. CT scan is the exam of choice, as it is a non-invasive diagnostic tool that provides important and detailed anatomical information, allowing a detailed evaluation of the aortomesenteric angle and distance.[3,4,6-8]

Initial treatment should be conservative and surgery reserved for refractory cases or those associated with anatomic malformations (short Treitz ligament, inferior origin of SMA, and extrinsic obstruction of third duodenal portion). Duodenojejunostomy is currently the procedure of choice.[1-4,6-9]

AWARDS AND PRESENTATIONS

Study presented at the 8th National Congress of Adolescent Medicine (8º Congresso Nacional de Medicina do Adolescente), where it was awarded 'Best Clinical Case'.

ACKNOWLEDGEMENTS

The authors acknowledge Dr. Víctor Pardal, head of the Department of Radiology of Centro Hospitalar de Leiria, for image selection.

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Received for publication: 04.11.2019
Accepted for publication: 09.10.2020