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CASE REPORT

SUPERIOR MESENTERIC ARTERY SYNDROME - WILKIE SYNDROME: REPORT OF TWO CASES

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ABSTRACT

Introduction: Superior Mesenteric Artery Syndrome / Wilkie Syndrome is characterized by the compression of the third portion of the duodenum, due to the reduction of the aorta angulation with the superior mesenteric artery. A rare disease, with few cases described in the medical literature. Case 01: A 13-year-old male patient, diagnosed with Diamond-Blackfan Anemia and with acute intestinal occlusion for 2 days with previous recurrent episodes of intestinal occlusion. CT showing gastrectasis and dilation of slender loops up to the third portion of the duodenum. Case 02: 18-year-old female patient, without previous comorbidities, with high intestinal obstruction for 7 days, with CT showing duodenal obstruction and reduction of aorto-mesenteric angulation. Discussion: The most prevalent age group occurs between 18 and 35 years. Associated with mainly weight loss and Cast Syndrome. The diagnosis is given by clinical suspicion followed by imaging tests such as upper gastrointestinal tract radiography and contrast abdominal CT. Treatment may be conservative with weight gain or surgical measures, the duodenojejunoanastomosis is the technique of choice. Diamond-Blackfan anemia despite being related to staturo-ponderal alterations no association was identified in the medical literature with Wilkie Syndrome. Conclusion: Differential diagnosis should be made in young patients with repetitive vomiting. Upper Gastrointestinal Endoscopy may delay the diagnosis.

KEYWORDS: UPPER MESENTERIC ARTERY SYNDROME; WILKIE SYNDROME; BLACKFAN-DIAMOND ANEMIA.ATRIAL APPENDAGE.

INTRODUCTION

Rokitansky described the Superior Mesenteric Artery Syndrome (SMA Syndrome) in 1842. Wilkie, in 1927, carried out a review containing 75 cases, and from then on it was known by his name, Wilkie Syndrome.[1,2]

It is characterized by compression of the third portion of the duodenum, due to the reduction of the angulation of the aorta with the superior mesenteric artery (SMA) .1,2,3 In normal individuals, the distance between the aorta and SMA is about 10-34 mm with an angle of 28-65o. In Wilkie Syndrome, this distance is usually less than 8 mm and the angle is less than 25 o.[3,4,5]

In this series of cases, two clinical cases of young patients diagnosed with SAMS are reported, one of whom has Diamond-Blackfan Anemia (DBA) and the other of a young patient with no known comorbidities, highlighting their clinical characteristics and treatment. It is a rare disease, with just over 500 cases described in the medical literature.[2,6,7]

CASE 01

Patient S.F.S, male, 13 years old, was attended at the Hospital de Urgências de Goiânia (HUGO) in February 2017 with a suggestive picture of acute intestinal occlusion for about 2 days. The patient reported onset of the condition with recurrent and unquantified episodes of vomiting preceded by nausea and diffuse abdominal pain, poorly located, like colic. He said that since then he has stopped the elimination of gases and feces and a progressive increase in abdominal volume.

Patient was in good general condition, oriented, Glasgow coma scale 15. Inaudible abdominal hydro-air noises, slightly distended abdomen, with diffuse pain on abdominal palpation, with no palpable viscera on physical examination, with no signs of peritoneal irritation.

He reported that, over the past 12 months, he had had recurrent episodes of vomiting and stopped the elimination of gases and feces lasting less than 2 days, with spontaneous resolution using scopolamine, without medical

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advice. He underwent digestive endoscopy (EDA), which showed no changes. He also mentioned that, in the same period, he had a statural growth of 10 cm (current height of 165 cm) maintaining the same body weight of 45 kg, which represents a reduction in the Body Mass Index from 18.7 to 16.5.

He reported a diagnosis of Diamond-Blackfan Anemia, diagnosed at 3 months of age, in follow-up at the Hematology Service of Santa Casa de Misericórdia de Goiânia. He reported that, since his diagnosis, he had undergone several non-quantified blood transfusions and continued use of corticosteroids until he was 8 years old. He also reported that he underwent a surgical procedure at 7 months of age in Belém - PA, for the treatment of gastroesophageal reflux (GERD). He denied other comorbidities and surgical procedures.

He underwent abdominal computed tomography (CT), which showed gastrectasis and dilation of slender loops up to the third portion of the duodenum, with no liquid passage after that area, and upper digestive endoscopy, showing megaduodene without an evident obstructive factor.

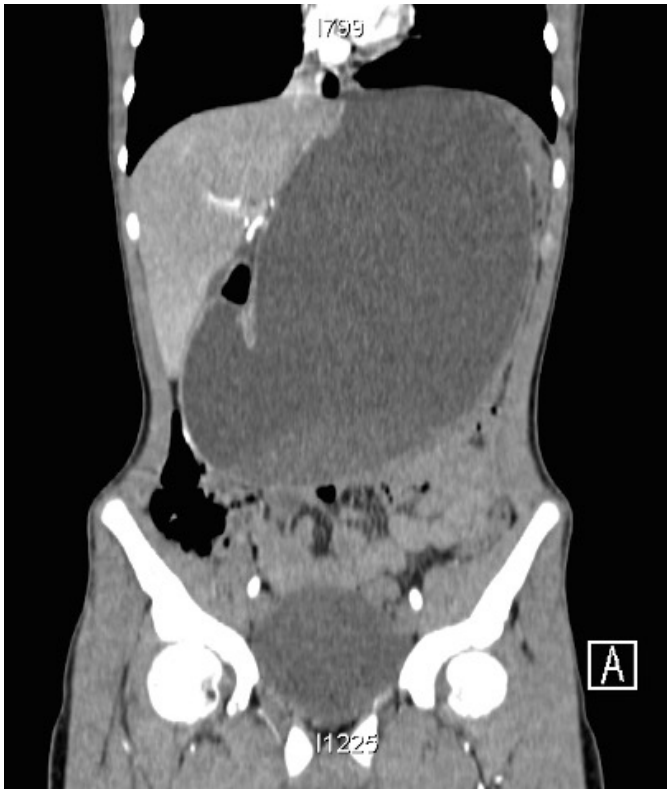

Image 1 - CT scan of the coronal abdomen showing gastrectasis.

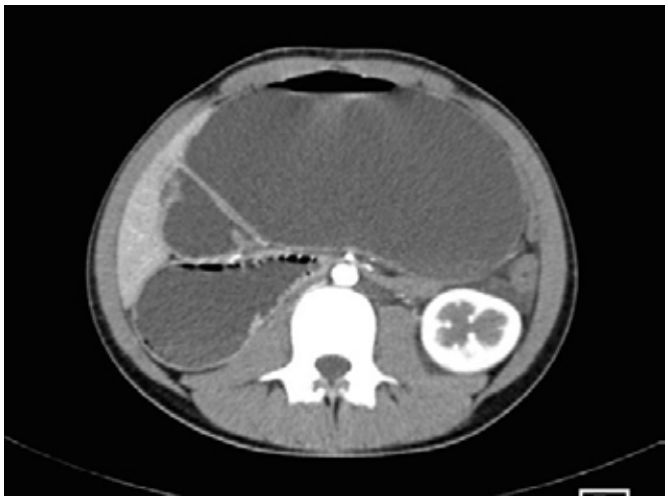

Image 2 - Axial abdominal CT showing gastric and duodenal dilation with duodenal compression point between aorta and superior mesenteric artery.

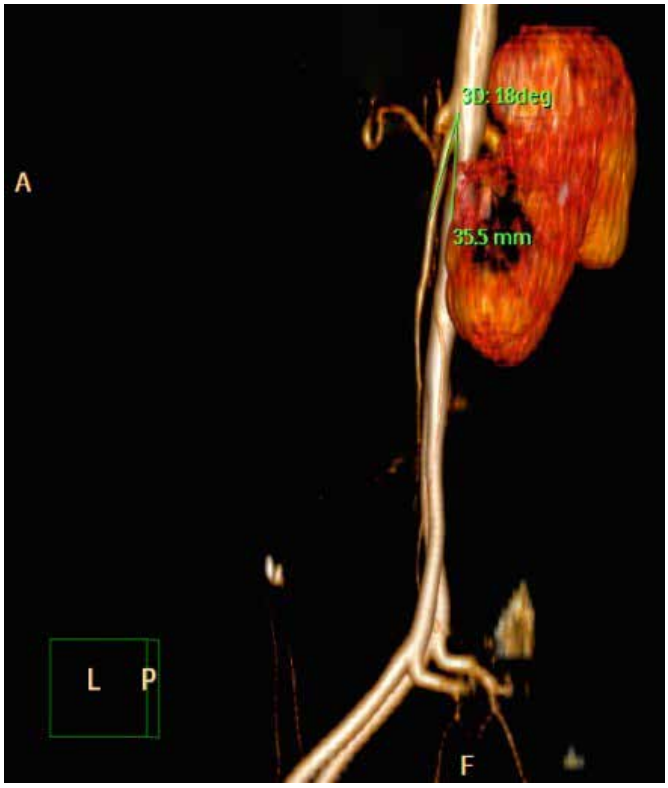

Image 3 - 3D sargital cut CT showing aorto-mesenteric angulation of approximately 18 o.

Based on clinical and imaging findings, the diagnostic hypothesis of Wilkie Syndrome was suggested.

He was initially treated with the passage of a nasogastric tube, showing partial improvement in abdominal distension and abdominal pain. In addition to the use of antiemetics, analgesics, anticholinergics, H2 blockers, and venous hydration.

The patient remained without bowel movements and eliminations, without new episodes of vomiting for 72 hours. Surgical treatment was indicated, with duodenojejunoanastomosis.

The patient evolved well in the postoperative period, an oral diet was introduced on the first postoperative day with good acceptance and progressive evolution of food consistency. He was discharged on the fourth postoperative day with physiological eliminations present and asymptomatic.

CASE 02

Patient A.C.M.L, female, 18 years old, was seen at the Hospital de Urgências de Goiânia (HUGO) in March 2019 with recurrent vomiting and nausea for about 7 days. She presented progressive worsening of the condition with abdominal pain in the upper abdomen, malaise and asthenia. She complained about having stopped the elimination of gases and feces 3 days before.

She reported that, over the past 3 years, she had presented recurrent episodes of nausea, vomiting, gastric fullness and malaise, with partial improvement of the condition after induction of vomiting and with the use of antiemetics without medical advice.

She stated that the onset of symptoms, 3 years ago, coincided with growth spurt and episodes of depression, associated with a stressful event in adolescence, reporting a marked loss of body weight. She denied other comorbidities and surgical procedures.

On physical examination, the patient was dehydrated, emaciated, Glasgow 15 coma scale. The abdomen was flaccid, with no signs of peritoneal irritation, with pain on deep palpation of the upper abdomen. BMI of 18.07 kg / m2. Laboratory tests showed leukocytes of 12000 / **µ**L, platelets of 250000 **µ**L, hemoglobin of 13.9 g / dL, amylase of 89 U / L, with no other evident laboratory alterations.

The patient underwent abdominal CT scan, which showed moderate gastrectasis and occlusion point in the third portion of the duodenum. The arterial phase of the imaging examination showed a reduction in the angle between the superior mesenteric artery and the aorta (11.85 °) causing duodenal obstruction. No other obstructive factors were identified. SMA Syndrome was diagnosed.

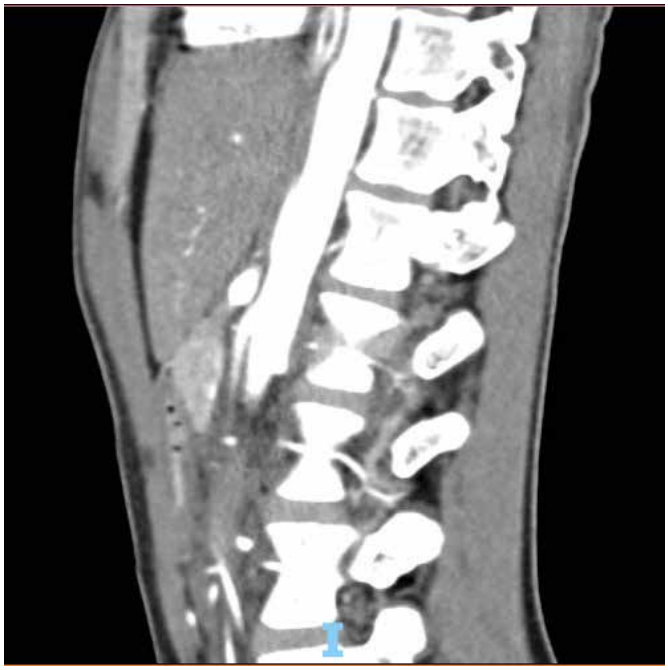

Image 4 - CT of the abdomen arterial phase, in sargital section, showing the emergence of the superior mesenteric artery of the abdominal aorta, with reduced angulation between the two vessels.

The patient was admitted to hospital where a nasogastric tube was passed for decompression, corrected the hydroelectrolytic disorders eventually identified, showing improvement in abdominal distension, abdominal pain, nausea and vomiting. The nasogastric tube was removed after 24 hours and a restricted liquid diet was introduced. After 12 hours of starting the diet, she developed new episodes of vomiting. Surgical treatment was indicated, with duodenojejunoanastomosis.

The patient evolved well in the postoperative period, with acceptance of the restricted liquid diet 24 hours after the surgery. She tolerated the progression of the diet well on subsequent days, being discharged on the 5th postoperative day, without complications. She continued outpatient follow-up with a general surgery team.

DISCUSSION

These reports describe the case of a 13-year-old patient and an 18-year-old patient diagnosed with SMA Syndrome, with the age group with the highest prevalence of such involvement occurring between 18 and 35 years. Women are most commonly affected.[8,9]

The history of intra-abdominal adipose tissue loss has an important correlation with Wilkie Syndrome, which

correlates with the cases presented. It can also be associated with a series of medical conditions such as malabsorption syndrome, eating disorders and bariatric surgeries.[8,9,10] Other associated conditions are Cast Syndrome (due to orthopedic plaster immobilization of the trunk and abdomen), peptic ulcer, thickening inflammation of retroperitoneum after pancreatitis, regional enteritis and scleroderma.[10,11]

In case 01, the patient had Diamond-Blackfan Anemia as a previous pathology, which consists of pure congenital aplasia of the erythroid lineage. It is a rare disease, with an incidence of 4-7/105 live births.[12,13] It can manifest as a severe anemia and can present, up to 25% of the time, congenital malformations, especially craniofacial and lower limbs. The most common manifestations are related to anemia, being mainly pallor and poor height-weight evolution. [12,14,15,16] The association between DBA and SMA Syndrome, although not identified in previous medical reports, are two rare conditions diagnosed in the same patient. In case 02, the patient did not report any previous comorbidities.

SMA Syndrome's clinical manifestations are variable, with vomiting and epigastric pain being the most common manifestations. Postprandial fullness, early satiety and anorexia are other manifestations found. Antalgic positions can be assumed as the genupeitoral and the left lateral decubitus. [7,8,17,18]

The diagnosis was established with careful clinical analysis and generally consists of a diagnosis of exclusion of other abnormalities of the gastrointestinal tract. Radiographic examination of the stomach and the duodenum (RESD) is one of the imaging tests that can be ordered, which would show dilation of the 1st and 2nd portions of the duodenum with abrupt termination of the dilations with or without gastric dilations, suggesting extrinsic compression.[4,5,7,19] Abdominal ultrasound can be used to anatomically assess the aorto-mesenteric angle. Contrast computed tomography has become the exam of choice because it is non-invasive and provides detailed information on the anatomy, level of obstruction and position of the SMA and its angle with the aorta. [6,7,19] UGE has little diagnostic value.[4,8,20]

Treatment can be conservative or surgical depending on the intensity of symptoms and clinical evolution. Conservative treatment is based on decompression with gastric aspiration and nutritional support to favor weight gain and increase in mesenteric fat, contributing to unblocking. Such support can be given by feeding via a nasoenteral tube passing the obstruction site, or via total parenteral.1,4,9,20 In addition, correction of hydroelectrolytic disorders must be considered and positional measures such as left lateral decubitus and genupeectoral position can be adopted to establish a greater amplitude of the aorto-mesenteric angle. [6,7,11,17]

The surgical approach can be performed using the Strong procedure, which consists of the division of the Treitz ligament, by gastrojejunostomy and duodenojejunostomy. [3,4,7,20] The latter was described in 1908 by Stavely, and is currently considered the technique of choice for presenting results superior to the others.[3,4,11] In the reported cases, this technique was chosen for the treatment of the disease .

CONCLUSION

Wilkie Syndrome is a rare cause of intestinal obstruction that affects mainly young adults with a history of weight loss and non-specific symptoms. It should be considered as a differential diagnosis of recurrent vomiting, especially in young individuals.

The diagnosis must be confirmed by imaging tests such as RESD and Computed Tomography. Duodenojejunostomy is the surgical treatment of choice in cases refractory to clinical measures.

No reports were found in the literature regarding the association between DBA and SMA Syndrome so far.

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