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Diagnosis and treatment of Dunbar Syndrome in a pediatric patient

Diagnóstico e tratamento de Síndrome de Dunbar em paciente pediátrica

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ABSTRACT

Median arcuate ligament syndrome or Dunbar's Syndrome is a rare pathology, not very prevalent in the population, whose nonspecific symptoms contribute to diagnostic difficulty, presenting severe abdominal pain in a marked way and attributing disability to the affected individuals. Together, postprandial abdominal pain, weight loss, and abdominal murmur characterize the symptomatological triad. Contrast-enhanced imaging methods are the best choice for identifying the pathology, especially in the angiography modality. To treat this disease, Dunbar suggested surgery in 1965 and to this day it persists as the best method of resolutive treatment, whether open or laparoscopic.

Keywords: Dunbar's syndrome, Median arcuate ligament, Celiac compression.

INTRODUCTION

With the name of Median Arcuate Ligament Syndrome, the so-called Dunbar syndrome is a rare condition, whose diagnosis consists of exclusion within the conditions of abdominal pain that include cholelithiasis, esophagitis, food intolerance, etc. It is a vascular compression

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syndrome of surgical resolution, characterizing a clinical entity that has a high impact on the affected individuals, since it is responsible for severe recurrent and postprandial pain, leading to their incapacity in the presence of the pathology.

Dunbar's syndrome was initially described in 1965 by Dunbar J D et al and the pathophysiology is not very well understood, but technically it is called celiac artery compression syndrome precisely because it presents coercion of this artery by the median arcuate ligament and diaphragmatic crura.

The median arcuate ligament (LAM) is a fibrous arch that connects the right and left crura of the diaphragm at the level of the T12-L1 vertebrae. Celiac artery compression may occur in a more cephalic origin of the artery and/or abnormally caudal insertion of the diaphragm. The literature reveals few meta-analyses that contain information about the presence of these abnormalities in various physiological individuals, which brings to light that not necessarily carriers of this anomaly have developed Dunbar's syndrome, that is, they will be symptomatic. This factor contributes to the misunderstanding of the etiological origin, but it is known that ischemia generated in the celiac artery is the factor of symptomatic origin, since this compression alters the artery at the cellular level, causing hyperplasia of the intimal layer, making the symptoms chronic. Studies also reveal that artery compression can be affected by several physiological factors, including respiratory movement.

The symptomatology therefore consists of the artery's ischemic factor, generating epigastric pain, nausea, vomiting, weight loss, and postprandial and/or exercise-induced abdominal pain, usually resulting from a compression of more than 50% of the arterial lumen. It is commonly seen in young women aged between 30 and 50 years, in a ratio of 3:1 or 4:1 varying according to the literature, with some pediatric cases described.

The diagnosis of this pathology is a challenge due to the non-specificity of the symptoms and has shown a high error rate in the various reports from which relevant information was extracted. Due to the rarity of the disease, there is no consensus or specific guideline for diagnosing the syndrome, however, because it is a case of mesenteric ischemia, angiography is indispensable. Even more advanced imaging methods can be subject to errors, such as computed tomography of the abdomen, which is why the time elapsed from symptom onset to diagnosis is so prolonged, leading to complications.

Among the treatments, the definitive resolution is surgical and, with the advancement of technology, laparoscopic arterial decompression and release of the arcuate ligament has become standard, capable of reestablishing flow and controlling pain by neurolysis. Percutaneous

transluminal angioplasty may be a preoperative adjuvant option for symptomatic relief, but its benefit disappears when chosen alone. In addition, for patients with cellular alteration of the intimal layer of the artery, with a tendency to chronification of the condition, celiac angioplasty and stencid implants are viable options.

The diagnosis and treatment of Dunbar's syndrome remains arduous for the medical team and the scarcity of metaanalytical data contributes to the altercation of the literature regarding the specific methods for this purpose. In this way, the most relevant information is taken from reports, which aims to assist in the scientific construction of this relatively recent theme within medical history.

METHODOLOGY

This is a case report type, in which information was collected through a review of medical records. At the same time, to support the ideas discussed in this article, a literature review was carried out in scientific databases such as PubMed and Scielo. The production of this scientific article followed the regulations proposed by the National Research Council (CONEP).

CASE REPORT

A 13-year-old female adolescent patient with no comorbidities and/or continuous use of medications was admitted to the children's emergency room of a referral hospital in the West of São Paulo with continuous pain in the epigastric region radiating to the dorsum for 2 years, with progressive worsening for 8 months, intensified in the last 4 days, associated with episodes of vomiting and dyspnea. no improvement or worsening factors and progressive weight loss. She reported having sought emergency care in the city of origin, being medicated with opioids, with no improvement, seeking a referral service due to difficulty in walking followed by pain. She denied urinary symptoms and other complaints.

The patient reported having been assisted by the Urology team after identifying kidney stones, treating them and not resolving the pain. Ultrasonography of the total abdomen and urinary tract showed anomaly in the right renal rotation and renal ectopy on the left. The urotomography revealed the presence of adnexal nodular formation to be clarified, and the gynecology and obstetrics team was activated, which guided outpatient follow-up. Chest X-ray and laboratory tests showed no abnormalities.

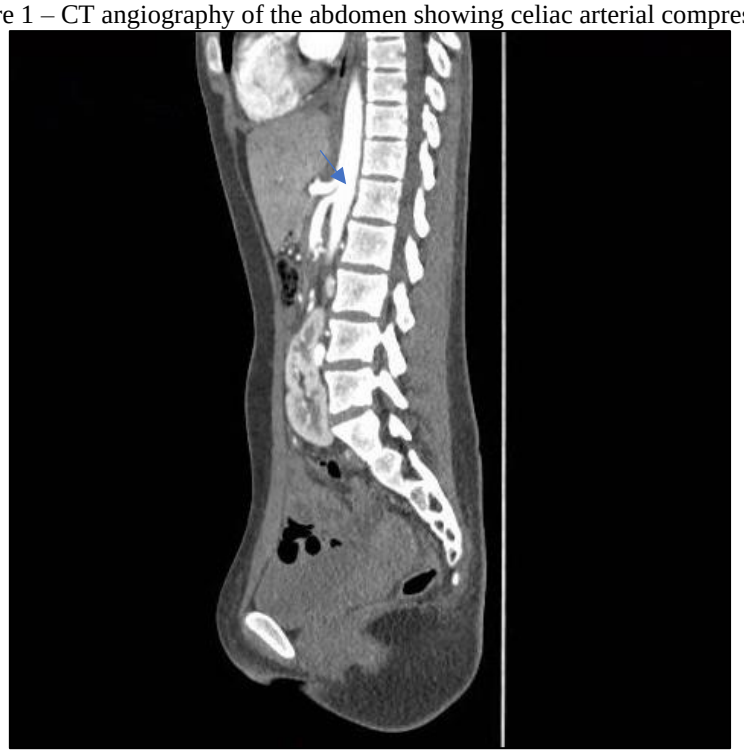
The patient was admitted to an intensive care unit bed with worsening of the pain, classifying it at 09 on the pain scale, globose abdomen, painful on superficial and deep palpation,

evacuation absent for 4 days, flatulence present. She reported that as she worsened, she noticed the evolution of postprandial pain, chest pain, radiating to the left upper limb and neck.

The general surgery team requested CT angiography of the abdomen, which showed marked stenosis (above 70%) at the origin of the celiac trunk, determined by the arcuate ligament. Its branches proved to be patent without significant stenosis. The surgical schedule made it possible to perform exploratory laparotomy, and a section was performed around the celiac trunk. The patient was hemodynamically stable and had good acceptance of the diet 1 day after surgery, with improvement in pain. The patient was discharged with outpatient follow-up on the 5th day and postoperatively.

Currently, the patient reports persistence of postprandial pain, but at low intensity, resolving with analgesics.

Figure 1 – CT angiography of the abdomen showing celiac arterial compression



Source: The authors.

DISCUSSION

Median arcuate ligament syndrome, also called celiac artery compression syndrome or Dunbar's syndrome, is a symptomatological entity caused by compression of the celiac trunk, whose vessel originates directly from the aortic artery. The celiac trunk originates just below the median arcuate ligament (fibrous ligament that connects the diaphragmatic crurae) and branches into the hepatic artery, splenic artery, and left gastric artery, which is responsible for supplying a large part of the digestive system.

The pathology was initially reported by Harjola and later described by Dunbar, who characterized the set of signs and symptoms that arise when the artery is implanted superiorly or when the ligament is more caudal due to genetic and/or environmental factors.

As elucidated in the literature, this syndrome most commonly affects young women in the age group of 20 to 40 years, but few pediatric cases have been described, having a rare attribute, affecting 2 out of every 100,000 people.

Pain is the most important symptom of the pathology. The one presented by this patient is pertinent to the anatomical pathological particularity, which refers to arterial and/or neuropathic compression, responsible for composing the triad of the disease: postprandial pain, weight loss and abdominal murmur. Although the murmur was not observed, it is noteworthy that the progressive worsening of the condition led to weight loss and pain was the most striking symptom, initially nonspecific, evolving to postprandial worsening, which made it difficult for the young woman in this report to eat food. This factor, associated with delayed gastric emptying, is responsible for the weight loss observed in all studies, including the one reported here.

The origin of the pain, as previously stated, may be adjacent to ischemia or neuropathic. As for ischemia, it is clearly justified by arterial mechanical compression, but neuropathic pain results from celiac plexus compression, whose origin is found adjacent to the median arcuate ligament and originates from the preganglionic splanchnic nerves, with somatic bundles of the phrenic nerve and the vagus nerve. As described in this case, it allows the appearance of pain of great intensity even outside the postprandial period.

All the data studied highlight the diagnostic difficulty of median arcuate ligament syndrome and the non-specificity of the symptoms is a major contributing factor to such altercation. The patient in this report took a period of 2 years to be correctly diagnosed, as other entities were first considered, such as appendicitis, gastritis, ovarian cyst and even kidney stones, which have been shown to be more prevalent in this age group. Several imaging modalities can be used to rule out differential diagnoses, making Dunbar's DS a diagnosis of exclusion9.

For this diagnosis, ionizing images with contrast, in the angiography category (either by computed tomography or magnetic resonance imaging) remains the best method of choice for this pathology, whose hook sign evidences the compression of the celiac artery. Doppler ultrasonography can also be used. According to the availability of hospital resources to which this patient was submitted, CT angiography was the decisive modality for the diagnosis to be

unraveled, and the image of this examination evidenced this finding, which is mentioned in the literature.

Chronic abdominal pain is a common concern among children and adolescents and is usually responsible for causing psychological comorbidities. According to the meta-analysis by Stiles-Shields, Colleen et al, 2021, 30% of patients undergoing surgery for the treatment of median arcuate ligament syndrome have persistent pain. When evaluating pediatric patients, this same study brings to light that this generates psychological consequences, triggering depressive and anxious syndromes in children and adolescents. Fortunately, the patient in this report did not present the development of any related comorbidity in the course of her pathology, but reported pain persistence at low intensity, especially postprandial.

As for the treatment, just as Dunbar proposed the surgery in 1965, there is currently no other modality that better meets the problem-solving needs of the condition. In 2000, the first laparoscopic procedures for the release of the celiac trunk were described, this method provides a less invasive alternative, reducing hospitalization time, chances of infection, less bleeding, etc., being the best choice to treat the pathology. Once again, the availability of resources at the hospital in question did not allow this choice, forcing the medical team to opt for open surgery with section of the structures, which allows better visualization of the vessel and greater possibility of hemorrhage control.

Other surgical alternatives include: arterial reconstruction and graft interposition (in cases of severe arterial obstruction or concomitant formation of aneurysms) and retroperitoneal endoscopic release, as well as placement of *stends* to release the lumen from the vessel.

Few meta-analyses have been performed on this subject, making the literature on Dunbar's Syndrome mostly isolated reports. Thus, there is no guideline to guide the medical professional regarding the diagnosis and treatment, which makes it difficult to identify the disease, making it an essential part of the professional's discernment and study. Median arcuate ligament syndrome should be considered in all patients who present with signs of chronic mesenteric ischemia or unwarranted severe abdominal pain.

Conflicts of interest

The authors affirm that there is no potential conflict of interest that could compromise the impartiality of the information presented in this scientific article.

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