

A Rare Case of Aortomesenteric Clamp Syndrome in a 14-Year-Old Adolescent

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Abstract:

Aortomesenteric clamp syndrome (SPAM) or Wilkie syndrome results from compression of the third duodenum between the superior mesenteric artery and the aorta; it is a rare entity, especially in children. The symptomatology resulting from this duodenal obstruction is that of a high occlusion. The diagnosis is based on CT angiography which may or may not be coupled with ingestion of water (or iodinated contrast product). The treatment is primarily medical, it is based on the installation of a nasogastric tube; correction of hydro electrolyte disorders and parenteral or enter jejunal nutrition. Surgery is only indicated if medical treatment fails. In this work, we report our experience in caring for a 14-year-old adolescent with SPAM.

Key words: aortomesenteric clamp syndrome, Wilkie syndrome, duodenal obstruction.

Introduction:

Aortomesenteric clamp syndrome (AMCS) or Wilkie syndrome results from compression of the third duodenum between the superior mesenteric artery and the aorta [1-7]; it is a rare entity, especially in children [1, 2, 8]. It is a multifactorial condition. Significant weight loss reducing the fatty mass protecting the duodenum [3, 9], deformation of the lumbar spine [9, 10] and anatomical anomalies (a short Treitz ligament, a low insertion of the superior mesenteric artery on the aorta) are the most frequently found contributing factors [1].

The symptomatology resulting from this duodenal obstruction is that of a high occlusion [4, 8, 11]; associates epigastralgia, postprandial vomiting, nausea, anorexia, and weight loss [6, 10, 12, 13].

The treatment is first medical, its failure requires recourse to surgery [7].

In this work, we report our experience in the care of a 14-year-old adolescent with AMCS and we discuss the diagnostic and therapeutic means.

Patient and observation:

This is a 14-year-old adolescent from a non-consanguineous marriage with no history admitted to our service in the month of Ramadan for the treatment of an occlusive syndrome, his history of the disease dates back 10 days. marked by the appearance of late postprandial vomiting following each eating attempt associated with a weight loss estimated at 03 kg after 10 days; all evolving in an afebrile context. On clinical examination the child was in moderately preserved general condition; mucocutaneous pallor; digestive transit was preserved; abdomen supple on palpation; not painful; rectal examination revealed a full rectal blister.

The biological analyses revealed microcytic hypochromic anaemia with a slightly disturbed ionogram.

The X-ray of the abdomen without preparation showed a rarefaction of digestive aeration (Figure 1); an ultrasound was done which showed stenosis syndrome without signs of gastric struggle; we completed the radiological assessment with a CT scan which concluded that there was significant

duodenogastric distension upstream of a disparity in duodenal calibre at the level of D3 located between the abdominal aorta behind and the mesenteric artery in front with an angle aortomesenteric estimated at 13.8° associated with a disappearance of perivascular fat thus confirming the diagnosis of aortomesenteric clamp syndrome (Fig 2).

Our course of action in this patient was the placement of a nasogastric tube to decompress the distended stomach and duodenum with parenteral nutrition, and correction of ionic disturbances.



Figure n° 1 : ASP debout



Figure n° 2

The evolution was marked by an improvement in the general condition; the nasogastric tube no longer brought back; a digestive transit taken up in the form of matter; a correction of the ionogram and oral nutrition resumed on day 8 of hospitalization.

The patient was discharged on day 13 of hospitalization with a follow-up appointment accompanied by a follow-up scan which came back normal.

Discussion:

It was Rokitansky who made the first description of aortomesenteric clamp syndrome in 1861 [12], Wilkie in 1927 presented the pathophysiology and treatment based on a series of 75 cases in adults [1, 8, 10]. It is a rare condition [1, 8], sometimes little known [1].

Its pathophysiology is linked to a reduced aortomesenteric space, less than 8 mm at the height of D3 associated with an aortomesenteric angle thus becoming less than 20° [4, 8].

Certain contributing factors are most often found: rapid weight loss leading to a reduction in the thickness of the adipose tissue at the level of the aortomesenteric space, spinal hyper lordosis, correction of scoliosis, anatomical anomalies (shortness of the ligament of Treitz, low origin of the superior mesenteric artery on the aorta), a systemic disease (scleroderma), an intervention on the aorta, cerebral palsy and recently a genetic factor has been incriminated after the description of a mesenteric clamp syndrome in members of the same family [1, 3-6]. In our case, there was significant weight loss during the month of Ramadan.

Symptoms are variable and non-specific. The installation can be acute or evolve insidiously depending on the aetiology and the importance of the duodenal obstruction [7], The acute form causes a severe high obstruction, with acute dilatation of the stomach [8], the Clinical symptomatology essentially boils down to repeated postprandial bilious or food vomiting [1, 10] as was the case in our patient. Other clinical signs are epigastric distension, weight loss and nutritional disorders [1].

The diagnosis is based on CT angiography which may or may not be coupled with ingestion of water (or iodinated contrast product). It shows localized dilatation involving the gastric and duodenal segments located upstream of the aortomesenteric clamp [1]. It can accurately measure the aortomesenteric space and angle [3, 4, 8, 10]. In recent studies, it has been demonstrated that the reduced aortomesenteric angle can be accurately visualized and assessed using colour Doppler ultrasound[2], Endoscopy provides proof of stenosis at the level of the 3rd duodenum: pulsatile extrinsic compression[9], Gastrointestinal transit highlights indirect signs, namely gastric and duodenal dilatation with incomplete linear stop of the contrast product at the level of D3[1, 9], in our patient it was thanks to an abdominal CT scan that the diagnosis was made.

The treatment is primarily medical. It is based on the placement of a nasogastric tube left in gentle suction to cause decompression of the stomach and duodenum [10], correction of hydro electrolytic disorders and parenteral or entero-jejunal nutrition [1, 8, 10], The goal is to increase the retroperitoneal fat mass and open the

aortomesenteric clamp. Postural measures by placing the patient in left lateral decubitus [8, 10] can also allow the clamp to open and therefore relieve the patient. Wearing a corset (in scoliotic patients) should be avoided until normal transit resumes [9]. Surgery is only indicated if medical treatment fails [1, 8, 9]. In our patient the medical treatment was carried out successfully with a favourable evolution and improvement in the general condition of the child, disappearance of vomiting and resumption of transit, a CT scan was done remotely, and which confirmed the effectiveness of our treatment medical

Conclusion:

Mesenteric clamp syndrome is a rare entity, especially in children; we must have the reflex to mention it when faced with a clinical sign of a high occlusion.

The injected abdominal scanner is a simple and effective examination to make the diagnosis. First-line treatment is always medical. Surgery should only be considered if medical treatment fails.

Conflicts of interest:

The authors declare no conflict of interest.

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