Superior Mesenteric Artery Syndrome: An Unusual Cause of Intestinal Obstruction

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Abstract

Superior mesenteric artery (SMA) syndrome is an uncommon cause of proximal intestinal obstruction, frequently occurring in patients who have had an important weight loss. The diagnosis can be difficult and usually is made by exclusion. The most characteristic symptoms are postprandial epigastric pain, fullness, voluminous vomiting and eructation. These symptoms are due to the compression of the third portion of the duodenum against the posterior structures by a narrow-angled SMA and surgical management is necessary. We report a case of SMA syndrome in a 23 year old patient, with a long history (since childhood) of voluminous vomiting, epigastric pain and an important weight loss. We performed small bowel enteroclysis, upper gastrointestinal series and endoscopy, biopsy of gastric and duodenal mucosa, abdominal computer tomography and ultrasonography to establish the diagnosis. Finally, the patient successfully underwent duodenojejunal anastomosis with a postoperative favourable outcome.

Key words

SMA syndrome - chronic duodenal ileus - arterio-mesenteric duodenal compression syndrome

Rezumat

Sindromul arterei mezenterice superioare reprezintă o cauză neobişnuită de ocluzie intestinală înaltă, ce apare mai frecvent la pacienţii care au suferit o scădere ponderală importantă. Diagnosticul poate fi dificil şi este de obicei unul de excludere. Simptomele caracteristice sunt reprezentate de dureri epigastrice postprandiale, senzaţie de plenitudine, vărsături importante cantitativ şi eructaţii. Aceste simptome sunt cauzate de compresiunea celei de a III-a porţiuni a duodenului pe structurile posterioare de către un unghi îngust al arterei mezenterice superioare. Tratamentul de elecţie este cel chirurgical. Comunicăm cazul unui pacient tânar, 23 ani, cu istoric de vărsături abundente din copilărie, dureri epigastrice şi scădere ponderală importantă, la care au coroborat în susţinerea diagnosticului radiografiile gastro-duodenale seriate, endoscopia, biopsiile din mucoasa gastrică şi duodenală, ecografia şi computer tomografia. În final, pacientul a fost supus intervenţiei chirurgicale cu efectuarea anastomozei duodeno-jejunale, cu evoluţie postoperatorie favorabilă.

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Introduction

Superior mesenteric artery (SMA) syndrome is an uncommon condition thought to be caused by the compression of the third or transverse portion of the duodenum against the aorta, resulting in chronic, intermittent or acute, complete or partial duodenal obstruction (1,2). Since it was first described by Von Rokitansky in 1861, about 400 cases have been reported in the medical literature, but many have doubted its true incidence.

Case report

A 23-year old man presented with a long history, since childhood, of abdominal cramps, voluminous vomiting, once a week, with his own description "I eat cherries in June and I vomit them at Christmas". There was no history of an eating disorder, nor medication taken and no remarkable family history. Apart from a marked tenderness and distension of the epigastrum and dehydration, the physical examination was normal. He had been in the emergency unit several times and he had received (years ago) treatment for a presumed duodenal ulcer.

On small bowel enteroclysis, a duodenograma showed an important dilatation of the duodenum and a massively distended stomach (Figs.1,2).

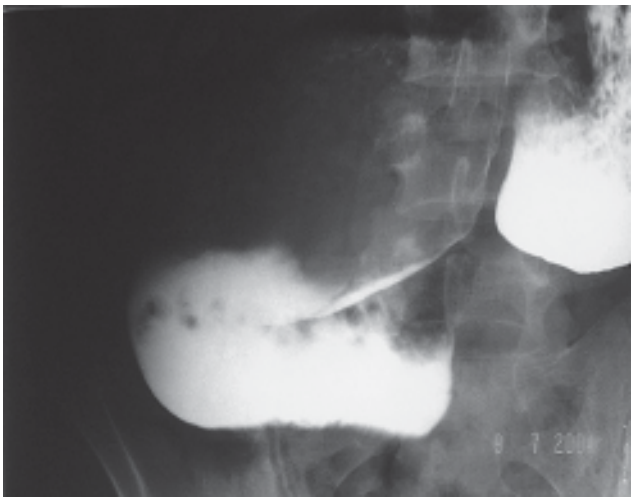

Fig1 X ray: Distended stomach and dilated duodenum.

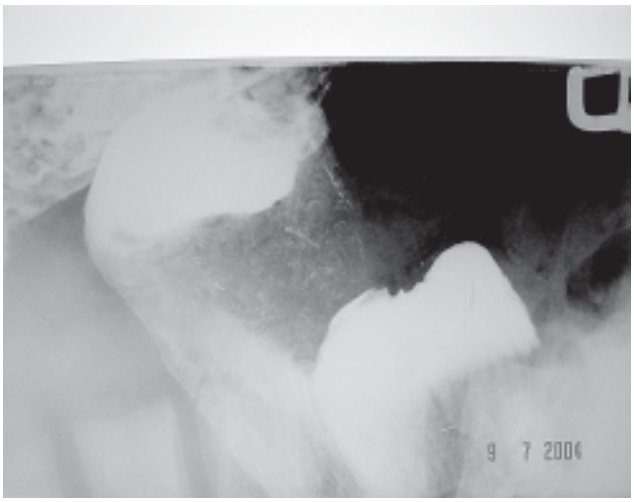

Fig.2 X ray: Dilatation of the duodenum.

The CT scan showed a massively distended stomach and proximal duodenum. The third portion of the duodenum was compressed between the SMA and the aorta. The superior mesenteric vein was also compressed.

The upper GI endoscopy showed marked narrowing of the third portion of duodenum as it passes over the aorta. The duodenal mucosa is similar with gastric mucosa, and the compression is seen at the level of the third segment of the duodenum as a very narrow portion, without any other pathology (Figs.3,4).

The patient was referred for surgery and a duodeno-jejunal anastomosis was performed successfully .

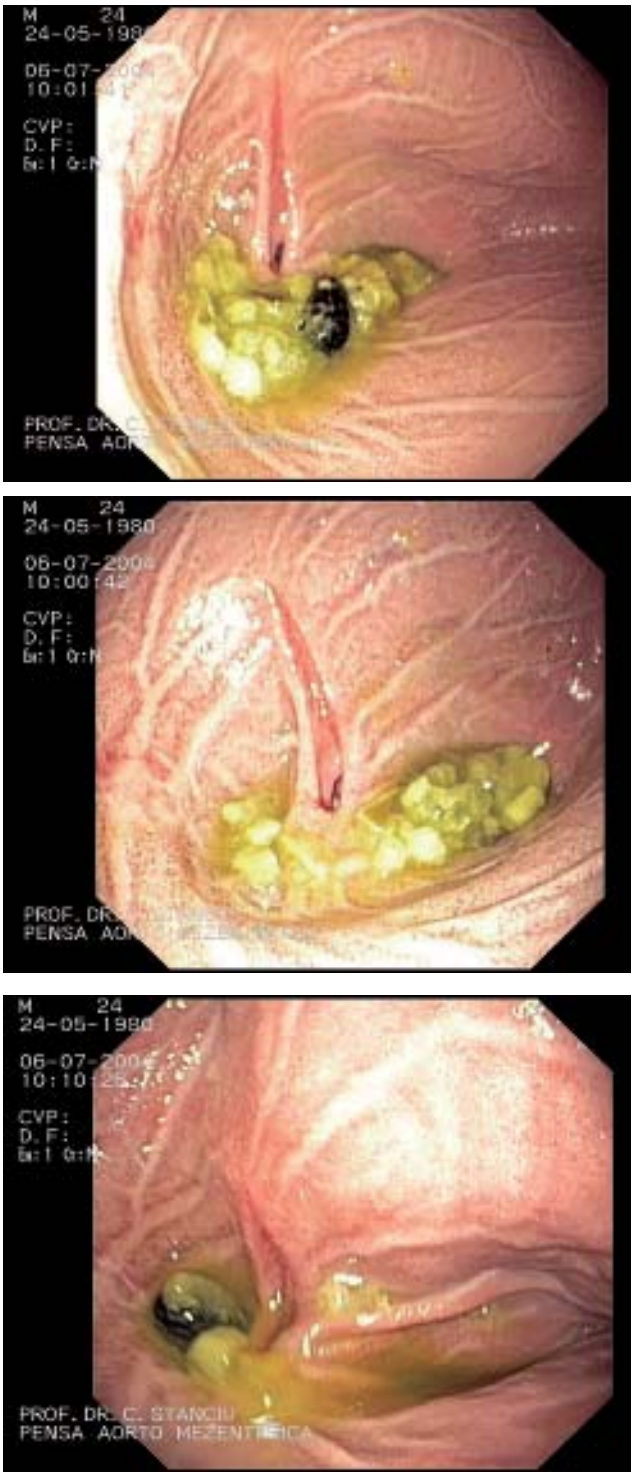

Fig.3 Endoscopic view: narrowing of the third duodenal portion.

Discussion

The SMA usually forms an angle of 45° with the abdominal aorta, and the third part of the duodenum crosses caudally to the origin of the SMA, coursing between the SMA and aorta. Any factor that sharply narrows the aortomesenteric angle causes the compression of the third part of duodenum, resulting in the SMA syndrome. In addition, the aortomesenteric distance in SMA syndrome is decreased to 2-8mm (normal 10-20mm). Many causes have been suggested, as a high insertion of the duodenum at the ligament of Treitz, a congenitally low origin of the SMA and compression of the duodenum caused by peritoneal adhesions, due to duodenal malrotation (3-5).

There are some important etiologic factors which may precipitate the narrowing of the aortomesenteric angle and

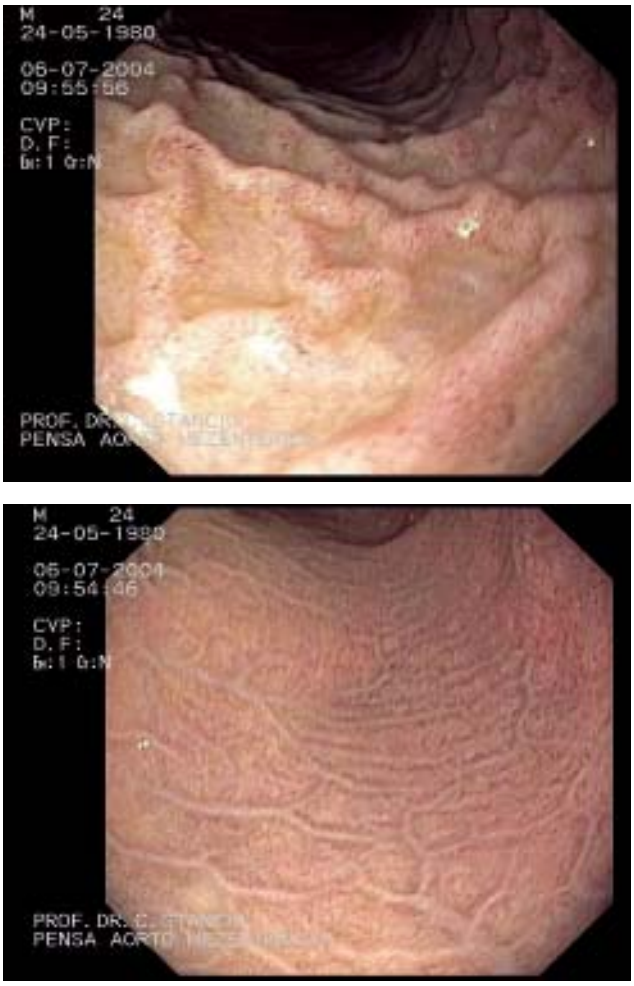

Fig.4 Endoscopic view: duodenal mucosa.

lead to chronically recurrent mechanical obstruction such as: constitutional factors (6,7) – slight body build, exaggerated lumbar lordosis, visceroptosis and abdominal wall laxity, depletion of the mesenteric fat caused by rapid severe weight loss due to catabolic states such as cancer and burns (8), severe injuries, such as head trauma leading to prolonged bed rest, dietary disorders such as anorexia nervosa (9,10), malabsorption, spinal disease, deformity or trauma (11,12), the use of body cast in the treatment of scoliosis or vertebral fractures, rapid linear growth without compensatory weight gain, particularly in adolescents.There are some unusual causes such as traumatic anevrism of the SMA, familial SMA syndrome, vascular compression of duodenum in association with peptic ulcer disease and traumatic mesenteric arteriovenous fistula following abdominal aortic aneurysm repair (1).

Our diagnosis was supported by age, clinical features and the long history of the patient. We considered abnormalities such as pancreas divisium, chronic idiopathic intestinal pseudoobstruction, or a SMA syndrome, from the beginning.

SMA syndrome occurs in females more often than in males, and some authors believe that SMA syndrome is not so rare, but is a matter of degree, claiming that many people with only mild compression do not present symptoms.

The clinical diagnosis can be confirmed by radiologic studies in 95% of cases (13). The following radiologic criteria have been established for the diagnosis of SMA:

- dilatation of the first and second portions of the duodenum, with or without gastric dilatation;
- abrupt vertical and oblique compression of the mucosal folds;
- antiperistaltic flow of barium proximal to the obstruction, producing to-and-fro movement;
- delay of 4 to 6 hours in transit through the gastroduodenal region;
- relief of obstruction when the patient is placed in a position (prone or knee-chest) that diminishes the drag of the small-bowel mesentery.

CT is also useful in demonstrating both the duodenal distention, the anatomy and the relationships of the superior mesenteric vessels and excluding other pathology.

By the use of selective SMA arteriography against a barium-filled duodenum it is possible to demonstrate extrinsic compression and to measure the aortomesenteric angle and the distance from the aorta.

The initial treatment of the SMA syndrome is generally conservative, with multinutritional support and postural therapy, in patients with a short history and a relatively minor degree of duodenal stasis shown radiologically.

Surgery is indicated if there is a long history of vomiting, progressive weight loss, pronounced dilatation and stasis of the duodenum. Procedures used are: duodeno-jejunostomy from the second portion of the duodenum to the jejunum (the best choice), gastrojejunostomy, Roux en Y duodenojejunostomy and anterior transposition of the third part of duodenum.

The outcome of the disease is excellent if it is diagnosed early and if the patient receives appropriate therapy.

In conclusion, SMA syndrome is a rare entity with severe consequences if not treated. Appropriate history and physical examination will facilitate the diagnosis.

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