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Delayed-Onset Superior Mesenteric Artery Syndrome Presenting as Oesophageal Peptic Stricture

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Key Words
Superior mesenteric artery syndrome · Oesophageal dilatation · Chronic acid reflux

Abstract
Superior mesenteric artery (SMA) syndrome is an infrequent cause of vomiting and weight loss due to compression of the third part of the duodenum by the SMA. We describe the case of a 17-year-old woman, admitted to our department for progressive dysphagia and severe weight loss due to an oesophageal peptic stricture, caused by chronic acid reflux secondary to duodenal compression by the SMA. Symptoms improved after (par)enteral nutrition and repeated oesophageal dilatation, thus supporting the role of intensive medical and endoscopic intervention as an alternative to surgery, at least in some cases.

Introduction

Superior mesenteric artery syndrome (SMA syndrome, also known as Wilkie's syndrome, aortomesenteric compression, aortomesenteric duodenal compression or duodenal vascular compression) is an uncommon cause of proximal intestinal obstruction, formally defined as trapping of the third portion of the duodenum between the SMA and the aorta secondary to bowel compression between the two vessels. The reported prevalence of this syndrome in the general population varies between 0.013 and 0.3% [1], although by applying stricter clinical or imaging criteria this rate may be

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reduced even further [2]. Females and young adults (18–35 years) are more likely to be affected by the condition, thought it can occur at any age [3–5]. This age and sex distribution may simply reflect the predisposing cause of the condition and, in particular, eating disorders. In this setting of patients, presenting with abdominal pain, nausea, anorexia, weight loss, and vomiting, the diagnosis of SMA syndrome should be considered. Patients may present acutely or more insidiously with worsening symptoms. In recent years, there have been numerous case reports of this condition, but in spite of this, its diagnosis is frequently delayed, resulting in ineffective symptomatic therapies and inappropriate investigations [6]. In fact clinicians needs a high degree of suspicion in order to diagnose SMA syndrome. The differential diagnosis includes megaduodenum [7] and other more common conditions, including chronic pancreatitis and peptic ulcer disease. The aim of this report is to describe the possible onset of SMA syndrome when the diagnosis is delayed, as happened in our young female patient with progressive dysphagia and severe weight loss. These symptoms result from gastric hypomotility and chronic acid reflux due to the 'trapping phenomenon' of the duodenum in the SMA syndrome, which progressively caused an oesophageal peptic stricture, as shown in the upper gastro-intestinal series and CT scan performed. After enteral nutrition and repeated oesophageal dilatation the patient showed improvement of symptoms and acquired an optimal nutritional status, thus avoiding surgical intervention.

Case Report

A 17-year-old female patient with somatic nonsyndromic hypoevolutism and dysplasia of the aorta was admitted to our department in November 2009 for progressive dysphagia for both solid and liquid nutrients, associated with weight loss (7 kg in the last 3 months) and occasional mild bloody vomiting. Her hypoevolutism, demonstrated through assessment of skeletal age with hand-wrist radiographs, had previously been evaluated through analysis of cariotype XX, malabsorption full screening including autoantibodies for coeliac disease, sweat test, study of the hypotalamus-hypophysis axis (growth hormone and gonadotropins), thyroid hormones and adrenal hormones (basal and after ACTH stimulus); however the cause of hypoevolutism remained unknown.

On physical examination the patient was severely malnourished (body weight 28 kg, body height 140 cm, BMI 14), and moderately hirsute. She had also a mid-systolic ejection murmur, heard best over the aortic area. Otherwise physical examination was negative. Blood pressure was 100/60 mm Hg, pulse rate 68 beats/min. Laboratory studies showed hyposideraemic anaemia (Hb 10.4 g/dl, sideraemia 23 μg/dl, ferritin 7 μg/l), hypotrigliceridaemia (39 mg/dl) and hypocholesterolaemia (100 mg/dl).

Upper gastro-intestinal series showed a marked stricture in the lower oesophagus associated with copious gastro-oesophageal reflux in the horizontal posture, reduction of gastric distension, and reduced gastro-duodenal peristalsis (**fig. 1**). A CT scan showed a 3-cm-long stricture in the lower oesophagus with pre-stricture dilatation and liquid stagnation, oesophageal wall thickening associated with hyperdensity and hypervascularity of peri-oesophageal fat, dilatation of the stomach and duodenum (until the third portion) with sharp calibre reduction of the fourth duodenal portion, around the aorto-mesenteric tract (**fig. 2**, **fig. 3**). Echocardiography showed moderate mitral valve insufficiency, mild tricuspidal valve insufficiency with normal pulmonary arterial pressure and an ejection fraction of 55%. Enteral nutrition with elementary diet and total parenteral nutritional were started, with improvement of her nutritional status (her body weight was 31 kg at discharge).

To improve her physiological oral feeding, a first endoscopic dilatation of the oesophageal stricture was performed in October 2009, under anaesthesiologic assistance, using a 10-mm-large Rigiflex Through The Scope (TTS) (Boston®) balloon, allowing for an oesophageal canalization with a residual channel 6 mm large. The subsequent oesophagography with contrast showed the

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maintenance of the oesophageal canalization (fig. 4) as seen in the radiographic series performed during endoscopic dilatation (fig. 5). Thus it was confirmed that the stricture arose from the chronic acid reflux due to gastric hypomotility and was sustained by the duodenal stenosis due to the SMA syndrome. A second endoscopic dilatation of the oesophageal stricture obtained an oesophageal residual channel 8 mm large and allowed endoscopic exploration of the duodenum. This showed duodenal compression in the presumed site of the SMA (fig. 6, fig. 7, fig. 8). After total parenteral nutrition and antibiotic therapy with ceftriaxone she started oral feeding again, and her body weight increased by 3 kg.

In January, February and September 2010 three further endoscopic oesophageal dilatations were performed, using up to 20 mm width. Histology showed the presence of heterotopia of gastric mucosa in the oesophagus and mild chronic gastritis associated with pancreatic heterotopia into the antrum. After the resumption of physiological oral feeding the patient showed a marked rise in her body weight (to 35 kg). The programme of repeated endoscopic dilatations of the oesophageal stricture allowed a reduction of the duodenal trapping in the aortomesenteric space (possibly due to the increase of the mesenteric fat pad after normalization of the patient's body weight) and then improvement of the gastro-duodenal peristalsis, as shown in the following upper gastrointestinal series.

Discussion

SMA syndrome is an uncommon and controversial form of upper intestinal obstruction in which usually the third part of the duodenum is compressed by the overlying SMA, due to compression between the aorta and the SMA, usually due to reduction of the mesenteric fat pad [8]. SMA syndrome has been described mostly in patients with severe and debilitating illnesses, malignancies and malabsorption syndromes, but also in other disorders associated with rapid weight loss such as anorexia nervosa, trauma or burns, spinal cord injury or paraplegia, and prolonged bed rest [9, 10]. Moreover it can be observed in younger patients after surgical correction of scoliosis, as a result of surgical lengthening of the spine, which may cause SMA cephalad displacement with a decrease in its lateral mobility [11].

Rokitansky is credited with the earliest description of SMA syndrome in 1861 [12], but the first case series was reported by Wilkie in 1927 [13]. The controversy of this entity is due to the fact that its signs and symptoms were not regarded as unique, because they could be found in other circumstances, such in neoplastic and inflammatory disease or electrolyte imbalance [8]. Patients may present acutely or more insidiously with gradual or progressive symptoms, consistent in both cases with proximal small bowel obstruction, such as postprandial epigastric pain and early satiety in case of mild obstruction, or severe nausea, loss of weight and emesis in case of severe obstruction [14]. Its principal complication are electrolyte imbalance, gastric perforation and duodenal bezoar formation [15, 16].

Although a case of congenital SMA syndrome presenting as gastro-oesophageal reflux has been previously described [17], its clinical presentation was not characterised by oesophageal stricture. In the present case we hypothesised that a delayed-onset SMA syndrome presenting as a misdiagnosed vomiting syndrome might have caused gastric dilatation and hypomotility with chronic acid reflux followed by oesophageal stenosis; this sequence of events was responsible for the chronic hyponutrition which in turn caused progressive worsening of the duodenal trapping and of the whole syndrome.

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A definitive diagnosis may be obtained with upper gastrointestinal series, which usually shows a marked delay in the passage of the contrast from the duodenum to the distal small bowel, with concomitant dilatation of the stomach and proximal duodenum, and also to and fro peristalsis, as showed in our patient [18]. Similar findings can be seen with CT, which, most importantly, can demonstrate by a tridimensional reconstruction the SMA superimposed on the duodenum, also providing further information on the amount of intra-abdominal and retroperitoneal fat [19, 20].

Usually the initial treatment of SMA syndrome is aimed at correcting fluid and electrolyte imbalance, decompressing the obstruction via a nasogastric tube, and establishing nutrition. Frequently, enteral nutrition is initially needed to achieve the main therapeutic goal, which is reversal of weight loss [21], then oral nutrition can be gradually started once significant weight gain is observed, such as in our patient. When a conservative approach fails, or in case of longstanding disease or complicated peptic ulcer disease secondary to biliary stasis and secondary reflux, surgical intervention, such as duodenojejunostomy, gastrojejunostomy, lysis of the ligament of Treitz or laparoscopic duodenojejunal bypass, may be successful [18]. Our patient represents a case in whom a complicated peptic oesophageal stricture, due to chronic acid reflux, worsened by gastric hypomotility and by duodenal trapping, was successfully treated through an endoscopic approach which allowed physiological oral feeding, thus obtaining improvement of her nutritional status and general well-being.

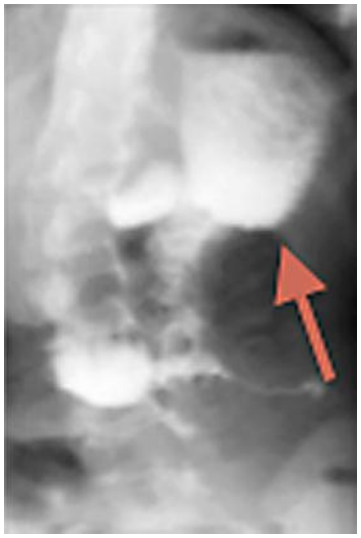

Fig. 1. Upper gastrointestinal series showing a reduction in gastric distension.

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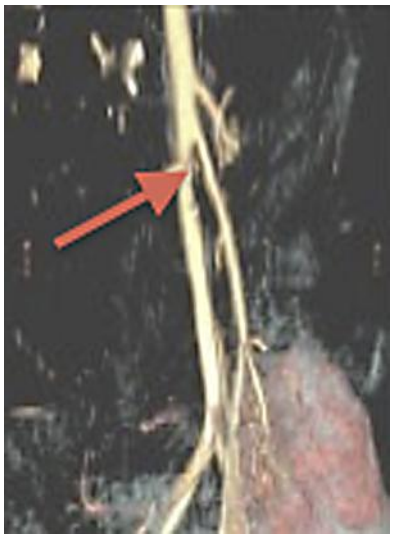

Fig. 2. 3D CT scan showing the narrow angle between the aorta and the SMA.

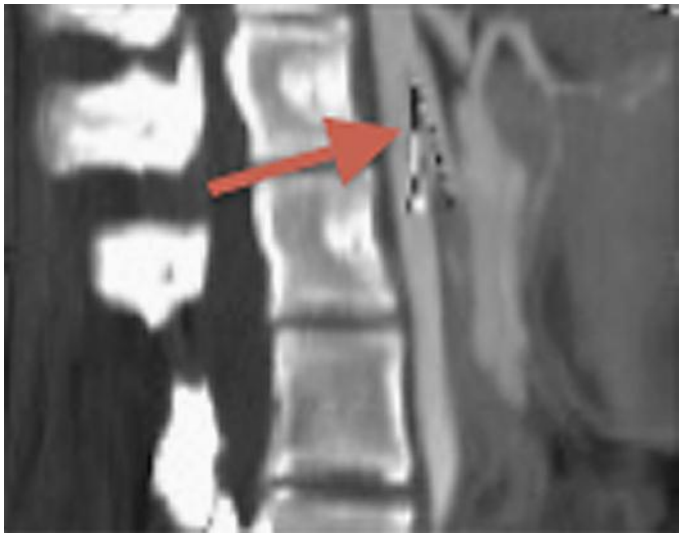

Fig. 3. CT scan focused on the narrow angle between the aorta and the SMA.

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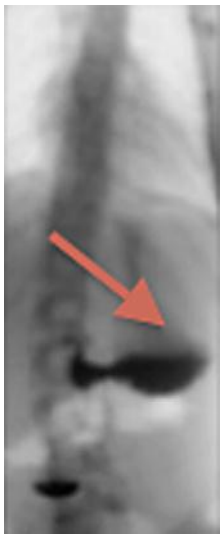

Fig. 4. Oesophagography showing the oesophageal canalization.

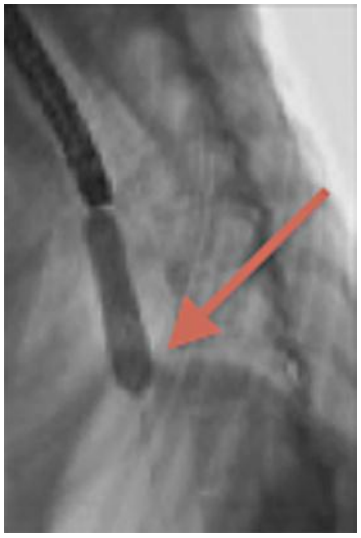

Fig. 5. Radiographic series performed during oesophageal dilatation.

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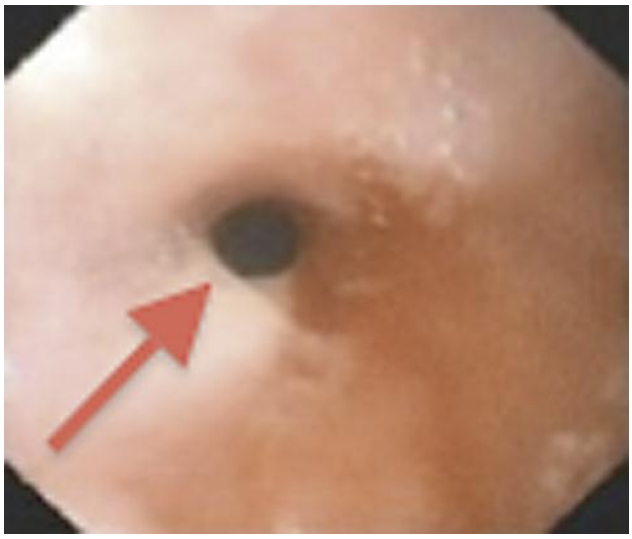

Fig. 6. Oesophageal lumen before the dilatation.

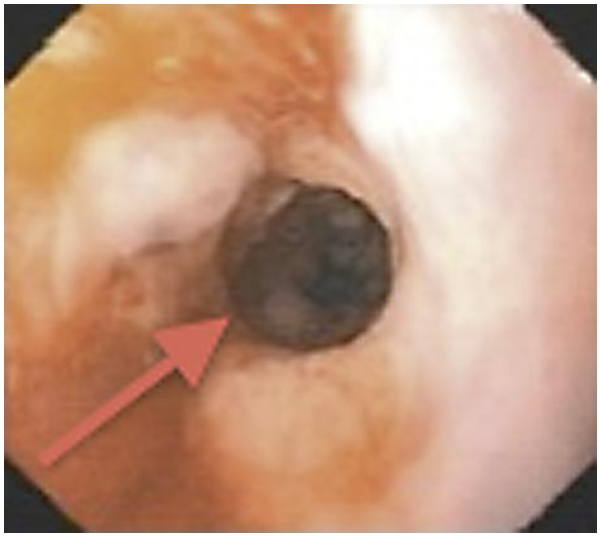

Fig. 7. Oesophageal lumen after the dilatation.

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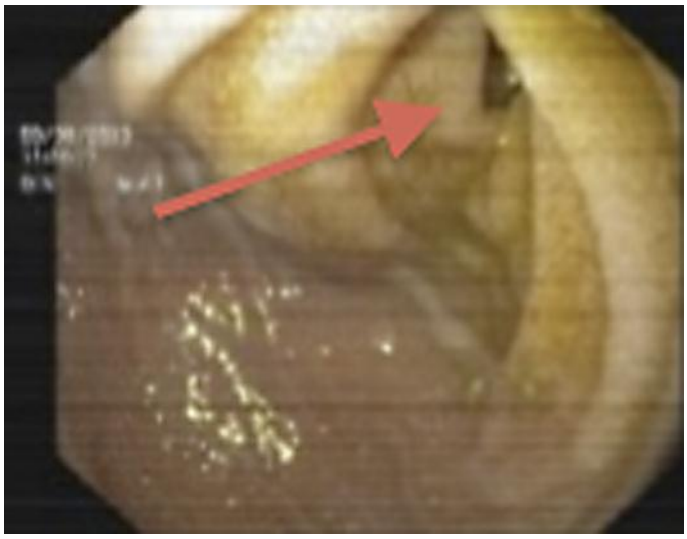

Fig. 8. Endoscopic image of the narrow duodenal lumen due to the 'trapping phenomenon'.

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