Superior mesenteric artery syndrome: an uncommon cause of abdominal pain mimicking gastroenteritis

腸繫膜上動脈綜合徵：一個酷似腸胃炎的不常見腹痛成因

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Abdominal pain and vomiting are frequently encountered in the emergency department. We report a 54-year-old man with an uncommon cause of intestinal obstruction — superior mesenteric artery syndrome — who presented with epigastric pain and vomiting. Diagnosis is clinical with radiological confirmation by upper gastrointestinal series or computed tomography scan. Most patients respond to conservative and supportive treatment. A minority may need surgical intervention. (Hong Kong j.emerg.med. 2008;15:235-239)

急症室常遇上腹痛及嘔吐。本文報告一名54歲男子腸梗塞的不常見成因：腸繫膜上動脈綜合徵，而呈現上腹痛及嘔吐。臨床診斷以上腸胃道放射造影系列或電腦掃描確定。保守性及支持性治療對大部份病人是有效的；少數可能需要外科介入。

Keywords: Abdominal pain, duodenal obstruction, intestinal obstruction, superior mesenteric artery syndrome, vomiting

關鍵詞：腹痛、十二指腸梗塞、腸梗塞、腸繫膜上動脈綜合徵、嘔吐

Case report

A previously healthy 54-year-old gentleman attended our emergency department (ED) one morning in August 2007 (Day 1) for one-day history of repeated vomiting (10 times of undigested food) associated with vague abdominal pain. Bowel opening was normal. He denied any fever, weight loss or change in bowel habit. Clinical assessment revealed no significant abnormalities. He was treated and discharged as gastroenteritis. He returned about twelve hours later for persistent vomiting. Clinical examination, electrocardiogram and X-ray chest showed no abnormalities. He was managed in the emergency medicine ward.

His complete blood count was normal except mild hypochromic microcytic anaemia (Hb 13.1 g/dL, MCV 62.9 fl). The serum amylase, liver and renal function tests were normal. Electrocardiogram and serial cardiac troponin T (cTnT) were normal. The chest radiograph showed clear lungs with no free gas under the diaphragm.
When the patient was reassessed in the morning round on the next day (Day 2), he had persistent vomiting and he could not tolerate oral intake. His vital signs remained stable with satisfactory hydration status. There was no diarrhoea despite the persistent vomiting. When the abdomen was examined, succussion splash was found. He was then kept nil by mouth with intravenous fluid supplementation. The presence of bilious gastric aspirate via the nasogastric tube and abnormal duodenal fluid levels on the abdominal radiograph (Figure 1) made the diagnosis of pyloric obstruction unlikely. He was transferred to the surgical ward for further evaluation and treatment.

He was managed in the surgical ward with intravenous hydration and nasogastric tube aspiration. Oesophagastroduodenoscopy (OGD) was performed on Day 3. The stomach and duodenum were filled with greenish substances. The scope could be passed down to the second part of duodenum (D2). The provisional diagnosis was intestinal obstruction.

On Day 4, a water soluble contrast follow through study was performed. There was dilatation of D1 and D2 with an extrinsic vertical linear compression defect in D3 as it crossed the midline (Figures 2a & b). There was transient hold up of contrast at D3. Compression

**Figure 1.** Abdominal radiograph on Day 2 of admission showing 2 air-fluid levels in the duodenal area with non-dilated distal bowel – suspicious of upper gastrointestinal tract obstruction.

**Figure 2.** Contrast upper gastrointestinal study AP (a) and oblique (b) spot radiographs demonstrating dilatation of the 1st and 2nd part of duodenum with vertical linear compression defect in the 3rd part of duodenum.
was relieved by postural change into the knee-elbow position.

The patient was further evaluated by contrast computed tomography (CT) scan of the abdomen and pelvis (Figures 3 & 4). The stomach, D1, D2 and D3 were grossly dilated down to a region of extrinsic vascular compression by the superior mesenteric artery (SMA) at the aortomesenteric compartment. D4 and the distal small bowel were not dilated. The angle between the SMA and abdominal aorta was less than 5 degrees. Thin subcutaneous fat was noted over the trunk of the patient’s body. There was no exaggerated lumbar lordosis. No other abnormalities were identified. The findings were compatible with the SMA syndrome causing obstruction at D3.

The patient gradually improved with conservative management and nutritional support with advice from the dietitian. A second look OGD showed diffuse gastritis (confirmed by biopsy and histology). Stool was negative for occult blood. Coagulation studies and thyroid function test were normal. Haemoglobin pattern analysis revealed beta-thalassaemia trait. He was discharged on Day 12. When he was seen in subsequent follow-up sessions, there was no more nausea or vomiting. His body weight was recorded as 52.2 kg and the stature was 171 cm with a calculated body mass index (BMI) of 17.95 kg m⁻². However, there was no identifiable precipitating factor for this episode of acute duodenal entrapment.

**Discussion**

The SMA syndrome is an uncommon but well recognised clinical entity which can easily be missed by the busy emergency physician. It is also known as Wilkie’s syndrome, arteriomesenteric duodenal compression, chronic duodenal ileus or body cast syndrome.¹⁻³ It is characterised by vascular compression of D3 against the aorta by the SMA. It was first described in the mid 1800s by von Rokitansky, who proposed that its cause was obstruction of D3 by arteriomesenteric compression.¹⁻⁷

The normal angle between the SMA and the abdominal aorta is between 45 to 60 degrees (Figures 5a & b). In the SMA syndrome, the angle is narrowed down to less than 22 degrees (Figure 4).⁵ Any condition that narrows the arteriomesenteric angle can precipitate entrapment of D3. Factors include thin body build,
exaggerated lumbar lordosis, visceroptosis, abdominal wall laxity, and depletion of mesenteric fat (e.g. rapid severe weight loss, catabolic state such as cancer and burns, severe injury leading to prolonged bed rest, or dietary disorders such as anorexia nervosa or malabsorption). Furthermore, spinal disease, deformity, or trauma and the use of a body cast in the treatment of scoliosis or vertebral fractures; rapid linear growth without compensatory weight gain, particularly during adolescence; and anatomical anomalies such as an abnormally high and fixed position of the ligament of Treitz, or an unusually low origin of the SMA are postulated causes of the SMA syndrome.²⁻⁴,⁶,⁷

Symptoms can be acute or chronic, or with intermittent exacerbation. It usually presents with features of high intestinal obstruction such as upper abdominal pain, nausea and bilious vomiting.⁷ In a review of the SMA syndrome in children, the presenting symptoms included abdominal pain (59%), vomiting (50%), nausea (40%), early satiety (32%) and anorexia (18%).³ Symptoms are typically ameliorated by the prone, knee-elbow or left lateral position and aggravated in the supine posture.¹⁻⁴,⁵,⁷

The precise prevalence is unknown. Some quoted the incidence as between 0.013 to 0.3%.¹ There is no discernible racial difference. There was female preponderance (around two-thirds) in one large series of 75 patients with the SMA syndrome.¹,⁴ It is also more prevalent in older children and adolescents.⁴ One report showed that 75% of cases occurred in patients aged 10-30 years.⁴

Confirmation of the diagnosis of the SMA syndrome is usually by radiological means, such as upper gastrointestinal (GI) series, hypotonic duodenography or contrast CT scanning.³⁷ In upper GI study, characteristic megaduodenum (pronounced dilatation of D1 and D2) and the frequently associated gastric distension are best seen in the supine position. There is a vertical linear compression defect in the transverse portion of the duodenum overlying the spine. There is also an abrupt change in calibre distal to the compression defect with relief of compression by postural change into the prone or knee-elbow position.² Moreover, the left lateral decubitus position and Hayes manoeuvre (i.e. pressure applied below the umbilicus in a cephalad and dorsal direction) which elevates the root of the small-bowel mesentery, may also relieve the obstruction.⁴ CT scanning is useful in the diagnosis of the SMA syndrome. With three-dimensional (3D) reconstruction, there is additional measurement of the aortomesenteric angle and aorta-SMA distance, assessment of intra-abdominal and retroperitoneal fat, and detection of any underlying pathology.⁶,⁷

Figure 5. CT abdomen of a normal patient with sagittal reconstruction showing normal angle between the superior mesenteric artery and abdominal aorta.

(AA: abdominal aorta; CA: celiac artery; D3: third part of duodenum; R: renal vein; S: superior mesenteric artery)
The mainstay of management is conservative.\textsuperscript{1,4,6,7} Precipitating factors should be identified and treated accordingly. Conservative management includes the maintenance of fluid and electrolyte balance, adequate nutrition, bowel decompression and proper positioning of the patient after eating (i.e. by either knee-chest or lateral decubitus position).\textsuperscript{1,4} Surgical intervention including duodenojejunostomy, gastrojejunostomy, or division of the Ligament of Treitz, may be necessary if conservative management failed.\textsuperscript{1,3,4,6,7}

Conclusion

In the absence of diarrhoea, one should be careful in the evaluation of patients presenting with abdominal pain and vomiting. Obstructive causes should be considered. The SMA syndrome is an uncommon cause of abdominal pain and vomiting. Diagnosis starts with the recognition of intestinal obstruction followed by confirmatory radiological imaging (upper GI series and contrast CT scan). Most patients respond to conservative management and only rarely are surgical operations needed.

References