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Cyclic vomiting in a young patient with superior mesenteric artery syndrome

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To the Editor,

Superior mesenteric artery syndrome (SMAS) is a rare condition characterized by duodenal compression through aorto-mesenteric clamp, also known as the Wilkie's syndrome. In SMAS passage is blocked in the lower part of the duodenum by vascular compression. Precipitating factors include increased lordoses, loss of abdominal muscle tone, rapid weight loss (2), abdominal surgery, or intrabdominal inflammatory conditions that may compress the mesenteric vessels. Symptoms are usually non specific and intermittent and include postprandial epigastric fullness, bloating, nausea, vomiting, and abdominal pain that are usually relieved by the prone or knee-to-chest position (3). Diagnosis is based on imaging techniques that include X-ray passage series of the upper gastrointestinal tract and angiographical visualization of the superior mesenteric artery and determination of its angle of branching from the aorta. Retrocolic duodenojejunostomy has proven to be the most satisfactory surgical treatment for this lesion (4).

We here report the case of a 16-year-old female patient who was admitted to our GI Endoscopy Unit complaining of meso-gastric pain, vomiting 3-4 hours after meal and weight loss. She underwent surgery for diaphragmatic hernia at the age of 6 months. She had been complaining of chronic intermittent symptoms since the age of 7. No diagnosis of the underlying condition had ever been reached despite barium X-ray of the upper gastrointestinal tract, abdominal US and upper and lower endoscopy. Symptoms were partially relieved by the use of anti-cholinergic drugs. In the last two months, clinical conditions worsened with 3-5 episodes of vomiting each day. Physical examination and biochemistry were normal. Coeliac disease and thyroid dysfunction were excluded. Upper endoscopy was normal; rapid urease test and histology for *H. pylori* infection were negative. Colonoscopy was normal. US and CT scan of the abdomen with intravenous contrast showed mild gastrectasy. A barium study small bowel follow through was performed and showed dilation of the first and second portion of the duodenum with compression of the third part of the duodenum (Fig. 1). Patient underwent MRI angiography of the abdominal aorta which showed narrowing of the aorto-mesenteric angle (Fig. 2), thus leading to the diagnosis of superior mesenteric artery syndrome



Fig. 1. — Small bowel series show an abrupt caliper reduction (arrow) of the last portion of the duodenum when crossing the midline, with mild dilation of the inferior duodenal curvature, due to extrinsic obstruction.

(SMAS). A retrocolic duodenojejunostomy (4) was then performed and patient is now doing well.

In a number of subjects, the angle between the aorta and the SMA is or becomes more acute than normal thus leading to a partial obstruction of the duodenum. Two forms of SMAS are described, an inborn and an acquired one, both of which result from a decrease in the aortomesenterial angle. The clinical course of the disease may

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Fig. 2. — The contrast enhanced magnetic resonance angiography of the abdominal aorta shows significant narrowing of the aorto-mesenteric angle (arrow), responsible of the duodenal caliper reduction.

be acute, due to a complete intestinal occlusion presenting as an ileus, or chronic intermittent with uncharacteristic upper abdominal symptoms. Because the onset of symptoms, in our case, dated back to the age of seven, it is likely that our patient had a congenital abnormality of the aorto-mesenteric angle leading to a chronic intermittent form of SMAS. Alternatively, the surgery for diaphragmatic hernia the patient underwent at the age of 6 months, might have led to a change in the aorto-mesenteric angle that, later, caused the occurrence of upper GI symptoms.

This report points out the elusive nature of this condition, which, in our patient, led to a misdiagnosis of functional disorder of the upper GI tract, despite several hospitalization. In fact, our patient had been treated for several years as having cyclic vomiting with anti-cholinergic, anti-hemetic drugs, or anti-depressants without any long-term benefit. Therefore, we suggest that in young patients with chronic vomiting and intermittent abdominal pain, SMAS should be ruled out by X-ray passage series of the upper GI tract, before diagnosing functional disorders.

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