Wilkie’s syndrome: a rare cause of intestinal obstruction

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DESCRIPTION

A 31-year-old woman was referred to our hospital, with recurrent episodes of profuse vomiting and upper abdominal pain associated with loss of appetite and dyspepsia since 2 months. She had no other comorbidities. She had been treated at another hospital with proton pump inhibitors, analgesics and intravenous fluids. She had a history of chronic anorexia and progressive loss of weight along with recurrent episodes of vomiting and upper abdominal pain. Clinical examination revealed dehydration, asthenicity (body mass index 18.8 kg/m²), abdominal distension, epigastric tenderness and positive suction splash on auscultation. Laboratory investigations showed a total white cell count of 13 500/mm³ and hypokalaemia (serum potassium: 3 mEq/L). Plain radiograph of the abdomen revealed gastric dilation. Ultrasonography was unremarkable. Upper gastrointestinal endoscopy showed dilated stomach and proximal capacious duodenum. Contrast-enhanced CT (CECT) scan revealed grossly distended stomach and duodenum proximal to the third part of the duodenum at the level of the origin of superior mesenteric artery with abrupt narrowing at this level, suggestive of superior mesenteric artery syndrome/Wilkie’s syndrome (figure 1). As conservative treatment failed, the patient underwent laparotomy after 10 days. At laparotomy, the stomach and the first and second parts of the duodenum were found to be greatly dilated and the offending superior mesenteric artery was seen clearly compressing the third part of the duodenum.

Figure 1 CT scan showing dilated stomach along with the first and second parts of the duodenum (D1 and D2). Calcifications can be seen within superior mesenteric artery (black arrow) and the aorta (white arrow).

Figure 2 Operative photographs (A) showing dilated stomach along with the first and second parts of the duodenum (D1 and D2); (B) the superior mesenteric artery (SMA) can be well seen compressing the duodenum; (C) Kocherisation of distended duodenum; (D) completed Roux-en-Y duodenojejunostomy (anastomosis is created between the duodenum and jejunum anterior to the SMA).
of the duodenum (figure 2). A Roux-en-Y duodenojejunostomy was performed. The postoperative period was uneventful, and the patient was discharged on the 10th postoperative day. The patient is on a regular follow-up and is asymptomatic since 3 years.

Learning points

▸ Wilkie’s syndrome is a rare cause of duodenal obstruction (incidence: 0.1–0.3%), which is characterised by compression of the third portion of the duodenum between the superior mesenteric artery and aorta due to narrowing of aortomesenteric angle from 45° (range between 38–56°) to about 6–25° (figure 3).1
▸ Clinical diagnosis requires a high index of suspicion especially in a patient who presents with postprandial abdominal pain, vomiting and a recent history of significant weight loss.
▸ Contrast-enhanced CT scan is often diagnostic with typical findings of duodenal distension along with narrowing of the aortomesenteric angle and assessment of retroperitoneal fat.2
▸ Surgical procedures (laparoscopic/laparotomy) include gastrojejunostomy, loop duodenojejunostomy, Roux-en-Y duodenojejunostomy and Strong’s operation (ligament of Treitz division).3

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REFERENCES