Wilkie's syndrome: an uncommon cause of intestinal obstruction

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Abstract A 10-year-old boy presented with 9 months history of gradually worsening, recurrent postprandial upper abdominal pain, bilious vomiting and loss of weight. On examination the child was undernourished, had epigastric fullness and succussion splash was positive. Ultrasonography of the abdomen suggested a massively distended stomach, while an upper gastrointestinal contrast study showed a hugely dilated stomach along with dilated first and second parts of the duodenum with abrupt cut off at the level of third part of duodenum. Contrast enhanced CT scan of the abdomen revealed dilatation of the second part of the duodenum without any obvious abnormality of the aorta-superior mesenteric artery angle. Upper gastrointestinal endoscopy showed retained fluid and food material within a dilated stomach and second part of the duodenum; scope could not be negotiated into the third part because of an extrinsic compression. The child was diagnosed to be suffering from Wilkie's syndrome. Exploratory laparotomy, performed when conservative management failed, revealed

compression of the third part of duodenum by a shortened ligament of Trietz and dense peritoneal bands near the third part of duodenum. The duodenal obstruction was bypassed by performing duodenojejunostomy. The child had an uneventful postoperative recovery. He gained around 6.8 kilograms within next five months.

Keywords High intestinal obstruction · Superior mesenteric artery (SMA) syndrome · duodenojejunostomy.

Key message Wilkie's syndrome is a rare cause of upper GI obstruction in children and young adults and should be considered when other common causes have been excluded. Surgical intervention, in the event of failure of conservative treatment, gives gratifying results.

Introduction

Wilkie's syndrome is an atypical cause of high intestinal obstruction often posing a diagnostic dilemma. The condition is a rare cause of upper GI obstruction with about 400 cases described since late 1800s. It was first described by Rokitsky in 1861 [1]. Though rare, it is a well recognized clinical entity and usually affects young adults. The syndrome is also known as Superior mesenteric artery (SMA) syndrome, Cast syndrome, chronic duodenal ileus and aorto-mesenteric duodenal compression.

It is commonly caused by compression of the third part of the duodenum at the narrow angle between the superior mesenteric artery and aorta. Compression of the third part of the duodenum by peritoneal bands or shortened ligament of Trietz may also be rarely responsible [2]. Diagnosis is often made by exclusion of other causes of upper gastrointestinal obstruction [3].

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Herein is a report of a very rare case of Wilkie's syndrome in a 10 year old child caused by dual pathology of extrinsic compression of the third part of duodenum due to dense peritoneal adhesions along with a shortened ligament of Trietz.

Case history

A 10-year-old boy reported at the surgical OPD with the history of gradually worsening recurrent pain in upper abdomen for 9 months. Pain was colicky in nature, used to start after feeds, last for 4 to 5 hours, and relieved with vomiting. The vomitus used to contain undigested food admixed with bile. Symptoms had no relationship with the position of the patient. The above symptoms were associated with reduced appetite and loss of weight (to the extent of around 7 kilograms over the period of last 8 months).

His mother gave history of her child suffering from periodic bouts of right upper quadrant fullness in association with flatulence, early satiety, occasional vomiting and constipation dating back to his early childhood days. Failure to thrive was also conspicuous. There was no significant past or family history.

On examination, the thin built child was 120 cm tall, weighed 35 kilos. The abdominal was scaphoid with fullness limited to the epigastrium, mild epigastric tenderness and prominent succussion splash. There was no palpable lump or any organomegaly. Bowel sounds were hurried.

Examination of blood did not show any abnormality except for a low hemoglobin percentage. Plain radiograph of the abdomen showed distended, fluid and gas filled stomach and duodenum. Ultrasonography of the abdomen suggested a massively distended stomach.

Upper gastrointestinal (GI) contrast study with 30% w/v barium sulphate showed a hugely dilated stomach along with dilated first and second parts of the duodenum with abrupt vertical cutoff at the level of third part of duodenum without any intrinsic mucosal abnormality (Figs. 1, 2). There was no apparent relief of obstruction in the left lateral decubitus position or with Hayes maneuver (i.e. infra umbilical pressure in the dorsal and cephalad direction to elevate the root of the mesentery). Gastro duodenal emptying was incomplete even after 6 hours.

A contrast enhanced CT scan demonstrated distended fluid and air filled stomach and duodenum without any obvious abnormality of the aorto-superior mesenteric artery angle. There was holdup of oral contrast at the third part of duodenum.

Endoscopic examination of the upper GI tract showed mild esophagitis, retained fluid and food material within a dilated stomach and second part of the duodenum, and narrowing of the distal duodenum due to an extrinsic compression through which the scope could not be negotiated.

The clinical signs and symptoms in conjunction with the investigative findings suggested the diagnosis upper GI obstruction possibly due to Wilkie's syndrome.

Initially a conservative approach was tried. The child was told to take frequent small meals of nutritious liquid, advised to lie on left side, prone or in the knee to chest position following meals. Metoclopramide was also advised. However, there was no symptomatic improvement and the child continued to suffer from epigastric pain, post-prandial abdominal discomfort and continued to lose weight.

A decision to abandon conservative management was taken after one month. The child underwent an exploratory laparotomy through a transverse upper abdominal incision. Intraoperative findings confirmed the extrinsic obstruction of the third part of duodenum by a short ligament of Treitz along with dense peritoneal bands near it. The stomach and

Fig. 1 Upper GI barium study shows hugely dilated stomach along with dilated first and second parts of the duodenum

Fig. 2 Upper GI barium study shows abrupt vertical cutoff at the level of third part of duodenum without any intrinsic mucosal abnormality.

proximal duodenum was markedly distended. The site of obstruction was further confirmed by nasogastric air insufflation.

The dense peritoneal bands precluded any attempt of lysis and mobilization of the duodenum. A duodenojejunostomy was constructed to bypass the obstruction. The child had an uneventful post operative recovery, tolerated oral fluids within 72 hours of operation without any vomiting and discharged from hospital after 10 days when he could tolerate solid foods. He gained around 6.8 kilograms within next five months and was completely symptom free.

Discussion

Though the SMA syndrome was first described way back in 1861 by Rokitsansky, it was in 1921 that Wilkie provided a patho-physiological description of the condition and named it “chronic duodenal ileus” [4]. Wilkie later published one of the largest series, totaling 75 cases [5].

The incidence of this condition varies from 0.013–0.3% of the barium series of the upper GI tract [6]. But many have suggested that the condition is actually over diagnosed because of its confusion with other causes of megaduodenum.

The most common cause is thought to be a sharp decrease of the aorto-mesenteric angle from the normal 45° to the range of around 6–16° resulting in the extrinsic compression on the duodenum as it passes between the aorta and SMA [6]. Any condition which leads to severe and rapid weight loss causes diminution of the fatty cushion around the SMA (which normally protects the duodenum from compression) leading to decrease of the aorto-mesenteric angle and compression of the duodenum. Rapid linear growth without compensatory weight gain in adolescence is an interesting cause as well [7].

Many other causes have also been implicated including high insertion of the duodenum at the ligament of Treitz, a congenitally low origin of the SMA, a short ligament of Treitz, and compression of the third part of duodenum by peritoneal adhesion as in this reported case [2, 8].

Patients generally present with chronic vague abdominal symptoms, recurrent episodes of pain in abdomen with vomiting or with occasional acute exacerbations of chronic symptoms. Typically the symptoms are relieved by lying prone, left lateral decubitus or in knee-chest position. The condition is difficult to diagnose and often made by a pro-

cess of exclusion. Non invasive procedures like contrast enhanced spiral CT scan and magnetic resonance angiography can help in confirming the diagnosis. The exact cause of this syndrome can sometimes be discerned only after exploration, as in this reported case, where it was found that a short ligament of Treitz along with extensive dense peritoneal bands near the third part of the duodenum was responsible.

If all age groups are considered, the differential diagnosis includes anorexia nervosa, bulimia, diabetes mellitus, collagen vascular disorders and chronic intestinal pseudo-obstruction.

The classical SMA syndrome because of narrow aorto-mesenteric angle compressing the duodenum responds to conservative management in the form of adequate nutrition by enteral (double lumen naso-jejunal tube)/parenteral feeding and proper positioning of the patient after feeds. Surgery is resorted to when conservative measures are ineffective or in patients with long history of progressive weight loss or pronounced duodenal dilatation with stasis and complications like peptic ulcer. In this reported case an initial conservative approach failed because of the intrinsic nature of the pathology. Duodenojejunostomy, the procedure performed in this case, has been reported to have a success rate of 90% [6].

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