Wilkie’s Syndrome: Effect Or Cause Of Severe Malnutrition - An Unusual Case Report.
N Kumar, B Rehmani

Citation

Abstract
Wilkie’s syndrome (WS) is one of the rarest gastrointestinal disorders. In WS, acute angulation of the superior mesenteric artery (SMA) causes compression of the third part of the duodenum between the SMA and the aorta, leading to duodenal obstruction. We are reporting a very rare case of acute Wilkie’s syndrome after pulmonary tuberculosis causing severe malnutrition in a 30-year-old male. The patient was married and well nourished two years back. He became severely malnourished due to acute Wilkie’s syndrome. The clinical details, difficulty in diagnosis and treatment are discussed.

INTRODUCTION
Wilkie’s syndrome is due to compression of 3rd part of duodenum (D3) between abdominal aorta (AO) and superior mesenteric artery (SMA). Wilkie’s syndrome is also known as superior mesenteric artery syndrome, cast syndrome, mesenteric root syndrome and chronic duodenal ileus. It was first described by Carl Freiherr von Rokitansky in 1861, but this disease was recognized only when Wilkie published the first comprehensive series of 75 patients in 1927. The normal angle between SMA and abdominal aorta is 38-65 degree. The loss of retroperitoneal and visceral fat by any cause decreases the aorto-mesenteric angle. Wilkie’s syndrome occurs when this aorto-mesenteric angle becomes 6 to 15 degree. It presents as early satiety, nausea, vomiting and postprandial abdominal pain. This produces severe malnutrition with spontaneous wasting, which further aggravates the symptoms. So it is effect as well as cause of malnutrition.

CASE REPORT
A 30-year-old male presented with complains of vomiting on and off and weight loss for 2 years, an epigastric lump after feeding for 1.5 years, dysphagia for both solid food and liquids for 3 months, and constipation and fever for 15 days. He had a history of pulmonary tuberculosis 2.5 years back with loss of appetite. He had taken incomplete antituberculous treatment. On admission, he was having good appetite but a feeling of early satiety. He also had peptic ulcer disease 2 years back. There was no history of hematemesis and melena. The patient was cachectic with a body weight of only 25kg (3 years back his weight was 54kg), and having scoliosis. His vital parameters were within normal limits. The abdomen was non distended, soft, and non-tender with positive succession splash, normal bowel sounds and normal digital rectal examination. The patient was admitted with a diagnosis of gastric outlet obstruction. On work-up, laboratory findings were: Hb 12.5gm/dl and TLC 5010/cmm with normal liver and kidney function tests and serum electrolytes. Esophago-gastro-duodenoscopy was suggestive of Grade-D esophagitis with dilated stomach and duodenum. CECT of the abdomen showed grossly distended stomach and duodenum up to D3 without any mass lesion. Barium meal follow-through demonstrated dilated stomach and duodenum up to D3. A Colour Doppler study of the superior mesenteric artery showed an acute aorto-mesenteric angle of 10-20 degree. Diagnosis of Wilkie’s syndrome was made on these findings, the patient was managed with TPN for 4 weeks with no improvement and then he was planned for duodenojejunostomy. Exploratory laparotomy was done and the diagnosis was confirmed. Peroperative findings were dilated stomach and duodenum up to D3 with small passable stricture at the D3-D4 junction. Duodenojejunostomy (side to side) was done between the 3rd part of the duodenum and the jejunum 10cm distal to the duodenojejunal flexure. Barium meal follow-through was done on the 5th postoperative day and showed normal passage of dye in the small intestine without any obstruction. The patient recovered well in the postoperative period with relief of all obstructive symptoms.
Wilkie’s Syndrome: Effect Or Cause Of Severe Malnutrition - An Unusual Case Report.

Figure 1
Fig. 1: X-ray showing pulmonary Koch’s disease in the left chest with scoliosis

Figure 2
Fig. 2: CT scan of the abdomen showing a narrowing at D3 with dilated stomach and proximal duodenum

Figure 3
Fig. 3: CT scan abdomen showing compression at D3 with dilated stomach and proximal duodenum
Wilkie’s Syndrome: Effect Or Cause Of Severe Malnutrition - An Unusual Case Report.

DISCUSSION

Wilkie’s syndrome is characterised by compression of D3 between aorta (AO) and overlying superior mesenteric artery (SMA). There is retroperitoneal and lymphatic tissue between AO and SMA, which provides a cushion for the duodenum. This cushion protects the duodenum from compression by the SMA. Any condition which leads to loss of this retroperitoneal fat and lymphatic tissue decreases the aorto-mesenteric angle and causes compression of D3 between AO and SMA.

Wilkie’s syndrome may be of two types: chronic variety and acute variety.

The chronic variety presents as early satiety, nausea and vomiting, postprandial abdominal pain (due to both duodenal compression and compensatory reverse peristalsis) and severe malnutrition with spontaneous wasting, which further aggravates the symptoms and vice versa. The risk factors for chronic Wilkie’s syndrome are asthenic body, high insertion of the duodenum at the ligament of Treitz, low origin of SMA and intestinal malrotation around an axis formed by the SMA.

Acute Wilkie’s syndrome presents as high intestinal obstruction with severe malnutrition or exacerbation of chronic symptoms. It occurs in patients with spinal cord injury, prolonged supine bed rest (after application of body cast), scoliosis and left nephrectomy. These conditions forcibly hyperextend the SMA across the duodenum. Predisposing factors for Wilkie’s syndrome are poor motility of digestive tract, retroperitoneal tumors, loss of appetite, malabsorption, kachexia, exaggerated lumbar lordosis,
abdominal trauma, rapid weight loss and starvation; 75% of cases occur between 10-30 years of age. The comorbid conditions associated with Wilkie’s syndrome are; hyperchlorhydria 50%, peptic ulcer disease 25-45%, pancreatitis and scoliosis\(^1\).

Diagnosis is difficult and made by exclusion\(^9\). Esophago-gastro-duodenoscopy and contrast-enhanced computed tomography (both IV and oral contrast) of abdomen and pelvis should be done. Esophago-gastro-duodenoscopy enables visualization of gastric and duodenal dilatation without any intraluminal cause. CECT of abdomen and pelvis may visualize a dilated proximal duodenum with compression of the distal duodenum between AO and SMA.

An acute case is usually managed by medical treatment and the chronic case requires surgical intervention. Medical management consists of removal or reversal of precipitating factors and providing proper nutrition and correction of fluid and electrolyte balance. Nutrition can be given by either feeding jejunostomy or by TPN (total parenteral nutrition) through central catheter. Symptoms improve after gaining weight, except reverse peristalsis, or when fat accumulation does not occur at the aorto-mesenteric angle\(^8\). When conservative treatment fails, or in severe cases of chronic Wilkie’s syndrome, surgery should be done\(^7\). The most commonly performed surgery is open or laparoscopic duodenojejunostomy\(^10,11\). It was first proposed in 1907 by Bloodgood\(^6\). In duodenojejunostomy, side-to-side anastomosis is done between D3 and jejunum. Thus, the compression of the duodenum is bypassed\(^11\). Some less commonly performed surgeries are Roux-en-Y duodenojejunostomy, gastrojejunostomy, anterior transposition of D3, and intestinal derotation and division of the ligament of Treitz. If there is persistence of symptoms after surgical bypass, prominence of reverse peristalsis can be traced. A duodenal circular drainage procedure (DCDP) can be done for correction of reverse peristalsis\(^8\). DCDP was originally implemented and performed in China. Our patient had scoliosis and pulmonary tuberculosis with loss of appetite as predisposing factors, with peptic ulcer disease. These factors lead to severe weight loss and acute-onset Wilkie’s syndrome in adulthood, the very rare gastrointestinal disorder which further aggravates the malnutrition. So, Wilkie’s syndrome is effect as well as cause of severe malnutrition.

References
Author Information

Navin Kumar, MS
Assistant Professor, Himalayan Institute of Medical Sciences

Babar Rehmani, MS
Associate Professor, Himalayan Institute of Medical Sciences