Research Paper

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A Rare Case of Wilkie's Syndrome

Medical Science

KEYWORDS : Superior mesenteric artery syndrome; duodenal obstruction; weight loss

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ABSTRACT

Wilkie's syndrome is a rare syndrome of intestinal obstruction occurring due to compression of third part of duodenum by the superior mesenteric artery.It usually occurs due to reduction of aorto-mesenteric angle caused by a variety of causes. It needs strong suspicion and keen evaluation to diagnose the condition.We present a case of 18 year old boy who presented with post prandial vomiting of 6 months duration along with loss of weight. This case highlights the fact that even rarest of causes should be kept in mind while evaluating a patient with intestinal obstruction.

INTRODUCTION

Superior mesenteric artery (SMA) syndrome, also called Wilkie's syndrome or cast syndrome, is a rare disorder in which acute angulation of the SMA causes compression of the third part of the duodenum between the SMA and the aorta, leading to obstruction. SMA syndrome is an atypical cause of proximal intestinal obstruction, most frequently occurring in young patients who have had an important weight loss due to various reasons. Loss of retroperitoneal fatty tissue as a result of this variety of conditions is believed to be the etiologic factor causing the acute angulation. Symptoms vary from postprandial nausea and bilious vomiting to abdominal pain as well as weight loss and can occur acutely or chronically[1]. The severity of the symptoms largely depends on the degree of the compression as reflected by the aorto-mesenteric angle. Advances in imaging, such as computed tomography (CT) and magnetic resonance imaging, have tremendously helped with clear visualization of the angle between the aorta and the SMA and thus improved the diagnostic rate [2]. In adults, clinical SMA syndrome manifestations appear if the angle drops below 20°, and it is believed that values of this angle may be lower for pediatric patients[3]. Thus, in the appropriate clinical context, detailed history as well as imaging findings should highly raise the clinical suspicion for the diagnosis of SMA syndrome. A delay in this diagnosis can potentially lead to many complications, such as electrolyte imbalance, catabolic wasting, gastric perforation and peritonitis.

Conservative therapy mainly consists of weight gain achieved orally or parenterally, with the aim of restituting the mesenteric fat pad and increasing the aorto-mesenteric angle[4]. If this non-invasive approach fails, surgical therapy may be the next approach, with duodenojejunostomy being the currently preferred treatment[5].

In this paper, we describe a case of SMA syndrome in a young male with characteristic symptoms of duodenal obstruction, which was diagnosed after 6 months. He had poor quality of life until the diagnosis was made and surgical treatment performed. Identification of this underestimated syndrome can be a diagnostic dilemma and is frequently delayed. We present this case to heighten the awareness of this syndrome and the need for early management to prevent delay in diagnosis and serious complications.

CASE REPORT

A 21 year young male patient presented to the surgical OPD with complaints of pain in upper abdomen and vomiting of 6 month duration. Pain was colicky not radiating and mostly localized to upper abdomen. It used to increase after food intake and was reduced after vomiting. The vomitus was bilious and contained partially digested food material. He had observed reduction in weight but was not able to quantify it. On examination he had a BMI of 14 and was dehydrated at presentation. Visible gastric peristalsis was observed, auscultopercussion test showed dilated stomach till hypogastric area. His lab investigations were within normal limits. He was evaluated with a working diagnosis of proximal small bowel obstruction. Barium meal showed dilated stomach and duodenum. Upper gastrointestinal Endoscopy showed esophagitis, dilated stomach and duodenum dilated till second part. Computed tomography of abdomen revealed partial obstruction distal to second part of duodenum. Abdominal angiography showed reduced aortomesenteric angle(20^0) (Picture 2) and reduction of aortomesenteric distance(5mm).A diagnosis of Superior mesenteric artery syndrome was established.

Over the next few days he was started on intravenous fluids, stomach wash was given and was prepared for elective surgery. Intra operatively gross dilatation of stomach, first and second part of duodenum was seen. Arterial compression over third part of duodenum was appreciated. Duodenojejunostomy was performed(Picture 3).

Post-operative recovery was smooth. He was started on oral liquids on second post-operative day and soft solids on the third day. Barium follow through was done on the eighth day which showed smooth flow of barium to small intestine. He was discharged in a stable condition on ninth post-operative day. On follow up after one month his appetite had improved and was tolerating large quantity of food and had gained two kilograms.

DISCUSSION

Superior mesenteric artery syndrome results from compression of third part of duodenum by the SMA. It was first observed by Rokitansky [6] and was published by Wilkie[3] as a case series, hence it was named as Wilkie's syndrome. It is also known by a variety of names such as arteriomesenteric duodenal compression, chronic duodenal ileus or cast syndrome.

Wilkie's syndrome is a rare cause of duodenal obstruction (incidence: 0.1–0.3%), which is characterized by compression of the third portion of the duodenum between the superior mesenteric artery and aorta due to narrowing of aorto-mesenteric angle from 45° (range between 38–56°) to about 6–25° [3].

Acute loss of the retroperitoneal fat pad between the SMA and

the aorta due to significant weight loss seen in patients with severe wasting conditions such as burns, severe trauma, cancers, eating disorders, or drug abuse results in narrowing of the aorto-mesenteric angle. Conditions that lead to prolonged bed rest including severe head trauma, cerebral palsy, paraplegia and application of a body cast may press the SMA against the duodenum or compress the duodenum against the hyperextended lumbar spine. Rapid growth spurts that exceed compensatory weight gain in adolescents, iatrogenic postoperative obstruction, adhesions, and an enlarged abdominal aortic aneurysm may also lead to vascular compression of the duodenum[7].

The symptoms are pain typical of small bowel obstruction-abdominal pain, anorexia, nausea, bilious vomiting, early satiety and post prandial fullness. Persistence of symptoms for prolonged periods leads to malnourishment, gross weight loss and deterioration of quality of life.

Diagnosing this condition is a challenge. An upper gastrointestinal (UGI) tract series reveals dilation of the stomach and the first and second portion of the duodenum, retention of barium within the duodenum, and a characteristic vertical or linear cut-off extrinsic defect in the third portion of the duodenum. The use of hypotonic duodenography can increase diagnostic accuracy to 90%. Aortic and SMA angiography in conjunction with hypotonic duodenography can delineate the aorto-mesenteric angle and crossing of the SMA over the duodenum at the site of obstruction and is considered the gold standard. Computed tomography scan with oral and IV contrast can potentially diagnose the condition and also the other causes of duodenal obstruction[7,2]. Upper GI Endoscopy also reveals dilated stomach and obstruction at the level of third part of duodenum. It can be used as an adjunct in the diagnosis.

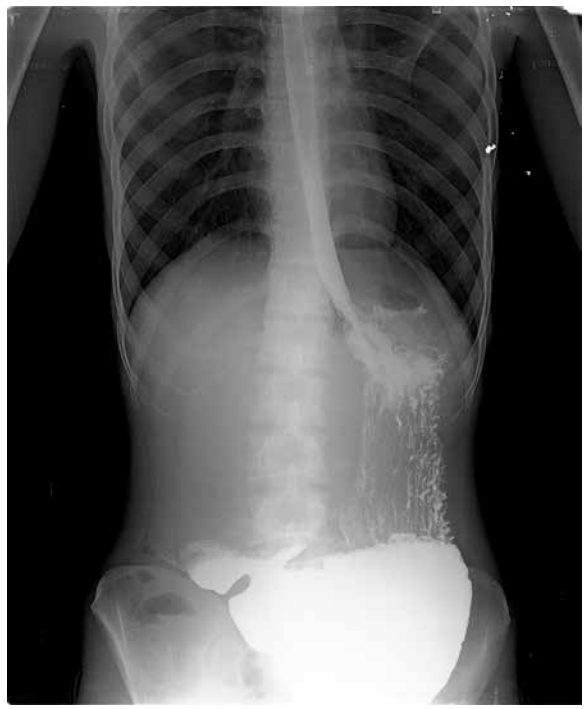
Management has been a controversy wherein a few studies have preferred only conservative management. But it may be effective in acute cases and might give temporary relief but recurrence is almost certain.

Surgical management is the definitive therapy. Wide range of operations,open and laparoscopic have been described but the standard of care at present is Laparoscopic duodenojejunostomy. Laparoscopic lysis of the ligament of Treitz with mobilization of the duodenum is another minimally invasive approach (Strong's procedure for congenital bands)[8].

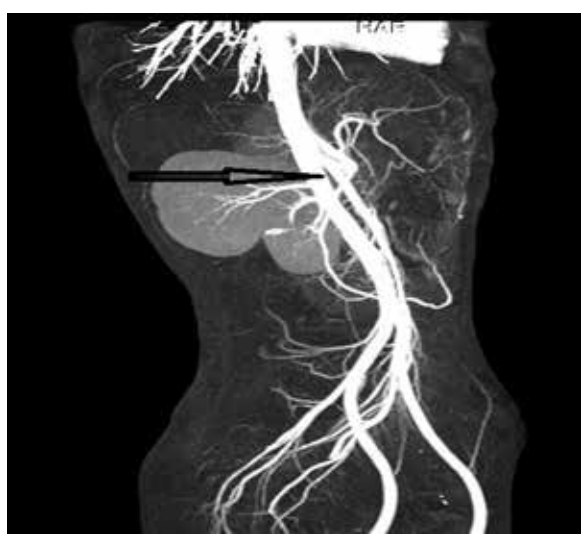
CONCLUSION

Wilkie's syndrome is a rare cause of duodenal obstruction due to compression of the third portion of the duodenum between the superior mesenteric artery and aorta due to narrowing of aorto-mesenteric angle. 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 reduction in retroperitoneal fat. Surgical procedures (laparoscopic/laparotomy) include gastrojejunostomy, loop duodenojejunostomy, Roux-en-Y duodenojejunostomy and Strong's operation (ligament of Treitz division). Early detection and treatment is necessary to prevent metabolic and nutritional complications.

Picture 1.
Barium swallow showing massive dilatation of stomach and duodenum



Picture 2.
CT angiogram with arrow depicting narrowed angle between Superior mesenteric artery and aorta.



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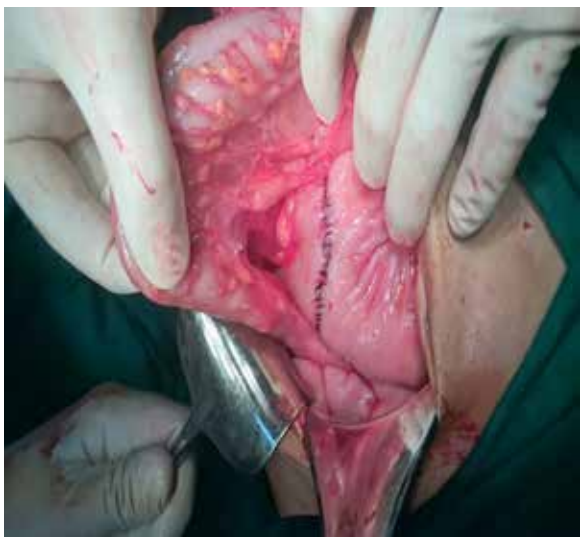
Picture 3.
Duodenojejunostomy completed anastomosis



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