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Wilkie's Syndrome in Children: A Case Report and Review of Literature

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Abstract: ***Wilkie's syndrome is a rare vascular cause of small bowel obstruction. It is more common in female and the etiology is multifactorial. It may be congenital or acquired. It occurs due to loss of perivascular fat between abdominal aorta and the superior mesenteric artery. This leads to decrease in aorto–mesenteric angle and cause compression of duodenum. The diagnosis can be made by upper gastrointestinal contrast study; Doppler ultrasonography and contrast enhance computed tomography. Management options are conservative or surgical. We are going to present a case of Wilkie's syndrome in an 11 years male child which was managed conservatively.***

Keywords: Aorto-mesenteric angle; children; malrotation, superior mesenteric artery; vomiting

1. Introduction

Wilkie's syndrome (WS) or superior mesenteric artery (SMA) syndrome is a rare vascular disease in which the third portion of the duodenum is compressed between the SMA (anterior) and aorto-vertebral plane (posterior). It is more common in women with reported incidence of 0.2-0.78% [1, 2]. This syndrome may be congenital or acquired. The congenital type is less common and symptomatic from childhood. Acquired type is due to loss of perivascular fat between abdominal aorta and the superior mesenteric artery which lead to decrease in aorto–mesenteric angle. Reduced aorto–mesenteric angle compresses the structures that are situated between aorta and superior mesenteric artery, such as third part of duodenum and the left renal vein. Patients present with painful crises, postprandial vomiting and left side varicocele [3, 4]. The diagnosis can be made by upper gastrointestinal contrast study and measuring the aorto– mesenteric angle with Doppler ultrasonography and contrast enhance computed tomography (CECT) [5]. After diagnosis, management options are conservative or surgical. We are going to present a case of Wilkie's syndrome in an 11 years male child which was managed conservatively.

2. Case Report

An 11 years male child presented to our outpatient department with history of intermittent pain abdomen and nonbilious vomiting for last two years. He had history of anorexia, weight loss and early satiety. There was no history of previous surgery, trauma, contact with tuberculosis and prolong immobilization. On examination, he was active, alert, conscious and afebrile. On per abdominal examination, upper abdominal fullness was present with no organomegaly. Clinically external genital was normal. Restsystemic examinations were normal. As per history and clinical examination, we were suspecting malrotation of gut and advised for ultrasonography. Ultrasound report suggested that dilatation of stomach, first and second part of duodenum. The angle between SMA and aorta was 18 degree and distance 2.5mm. We were advised CECT abdomen for confirmation and findings were dilated stomach and 1st and 2nd part of duodenum (Figure 1A) with luminal narrowing of third part of duodenum (Figure 1B). Aorto-mesenteric angle was 11 degree and aorto-mesenteric distance was 2.5mm (Figure 1C).

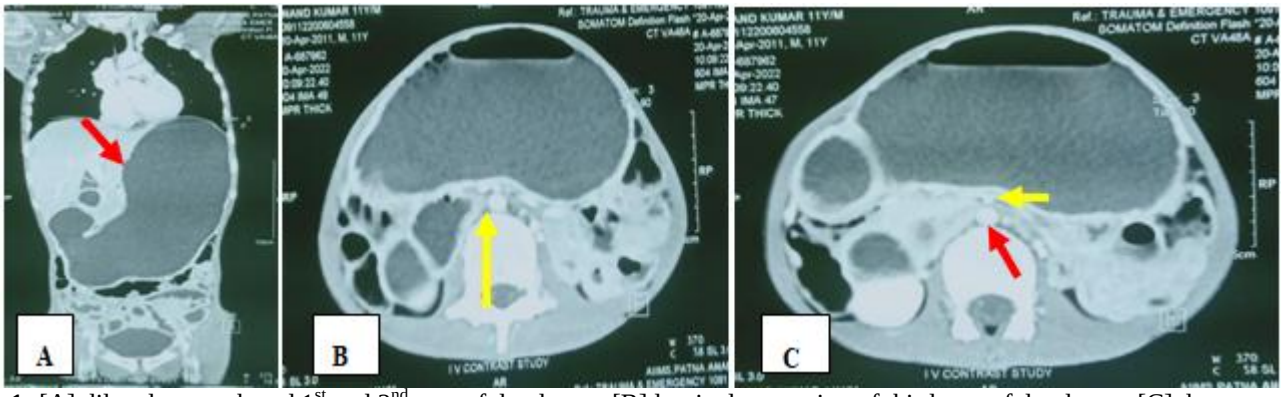

Figure 1: [A] dilated stomach and 1st and 2nd part of duodenum [B] luminal narrowing of third part of duodenum [C] decrease Aorto-mesenteric angle

After diagnosis, naso-jejunal tube was placed under fluoroscopy guidance and high calories diet started along with medium chain triglyceride. Gradually emetic symptoms were improved and patient gained weight. Follow-up ultrasound suggested that aorto– mesenteric angle was increased to 20 degree and adipose tissue's thickness was increased to 4mm. We kept patient on regular follow-up and planned for ultrasonography to see further increase in aorto-mesenteric angle and adipose tissue thickness.

3. Discussion

Wilkie's syndrome is a rare vascular cause of small bowel obstruction. It is more common in female (2:1) [2]. The etiology is multifactorial such as weight loss, prolong immobilization, spine injury and spinal surgery [6, 7, 8]. Laffont et al reported that Wilkie's syndrome occurred in paraplegic patients after injury. They hypothesized that, an imbalance in autonomic nervous system that results in overactive parasympathetic activity which may correspond to the development of Wilkie's syndrome [9]. Previous surgical interventions which reduce the width of the aorto-mesenteric angle have been associated with Wilkie's syndrome such as bariatric surgery, scoliosis surgery, ileo-anal pouch anastomosis, and aortic aneurysm repair [10]. Patients present with symptoms like bowel obstruction such as intermittent vomiting, nausea, postprandial fullness, early satiety, anorexia and abdominal pain [11]. Wilkie's syndrome can be diagnosed with Upper gastrointestinal contrast study; Doppler ultrasonography and contrast enhance computed tomography (CECT) [5]. The ultrasound is a very sensitive method and accurately measure decrease in aorto–mesenteric angle. Normally this angle is between 36–56 degrees with distance of 10–20 mm [12, 13]. In our index case, aorto–mesenteric angle was 11 degree and distance was 2.5mm. Management options for Wilkie's syndrome are conservative or surgical. Goals of conservative management are mainly focused on bowel decompression, correction of fluid and electrolyte imbalance, nutritional support, rehabilitation and removal of the body cast during acute condition [14]. After stabilization, small frequent high caloric oral diet with changes in feeding position such as knees to chest or left lateral position may be helpful in bypassing the obstruction. Our case was managed conservatively. There are no such guidelines for total duration of the conservative treatment [15]. Surgery is indicated when symptoms are not improve on conservative management and options are duodeno-jejunostomy or gastrojejunostomy. There are no such guidelines for total duration of the conservative treatment [15]. We concluded that, Wilkie's syndrome can be managed conservatively and surgical intervention is reserved for that patient which is refractory to medical management.

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