CASE REPORT

WILKIE'S SYNDROME: CASE REPORT

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HOW TO CITE THIS ARTICLE:

Prabhu Dayal Sinwar, Krishna Veer Singh Choudhary, R. K. Kajla, Ramniwas Dhukiya, Rakesh Kumar. "Wilkie's Syndrome: Case Report". Journal of Evolution of Medical and Dental Sciences 2014; Vol. 3, Issue 44, September 15; Page: 10857-10860, DOI: 10.14260/jemds/2014/3416

ABSTRACT: Introduction: Wilkie's Syndrome (also called SMA Syndrome) is a rare cause of small bowel obstruction, characterized by an extrinsic vascular compression of the third portion of the duodenum between the abdominal aorta and overlying superior mesenteric artery, due to loss of cushion of fat. **OBJECTIVES:** To aware, diagnose and manage a rare case of Wilkie's Syndrome Methodology: Case suspected clinically, diagnosed by radiological investigations and managed by duodenojejunostomy **RESULTS:** Clinically, Wilkie's Syndrome presents with postprandial abdominal distension, pain, nausea, vomiting, and weight loss. Fasting, total parenteral nutrition, and gastric decompression constitute usual conservative treatment with a high success rate. Surgery if needed has a low failure rate and consists of creating a gastrojejunostomy or duodenojejunostomy with or without duodenal mobilization (known as the Strong's procedure). **CONCLUSION :** Wilkie's Syndrome is a rare disorder. Only 500 cases reported worldwide till date. Surgical treatment is the treatment of choice in long standing cases otherwise it may result in fatal complications like advanced malnutrition, electrolyte imbalance, gastric rupture, spontaneous upper GI bleeding or sudden cardiovascular collapse.

KEYWORDS : Wilkie's syndrome, SMA, postprandial.

INTRODUCTION: Wilkie's Syndrome is a rare cause of small bowel obstruction, characterized by an extrinsic vascular compression of the third portion of the duodenum between the abdominal aorta and overlying superior mesenteric artery, due to loss of cushion of fat. Clinically, it presents with postprandial abdominal distension, pain, nausea, vomiting, and weight loss.

Fasting, total parenteral nutrition, and gastric decompression constitute usual conservative treatment with a high success rate. Surgery if needed, has a low failure rate and consists of creating a gastrojejunostomy or duodenojejunostomy with or without duodenal mobilization (known as the Strong's procedure).

CASE REPORT: A 37 years old male patient presenting in surgical outdoor referred by gastroenterologist in P.B.M. Hospital Bikaner with chief complaint of recurrent vomiting for 4 months. Vomiting occurs after 3-4 hours of taking meal bilious, non-projectile containing eaten food particles. Vomiting occurs most of the time after taking solid meal. There was no history of constipation. History of weight loss was present in four months (70kg to 55kg). No history of any trauma

Personal History: Opium edict for 10 years, Tobacco Chewing for 10 years, History of alcohol consumption for 7 years.

Drug History: History of NSAID (Diclofenac) intake 2-3 times/day for 4-5 years. Alprazolam 0.5mg intake 2 tablets in morning and 2 tablets in evening for 4-5 years.

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General Physical Examination: Patient was conscious and oriented to time, place and person.

Blood pressure - 110/70mmHg.

Pulse rate – 86/min.

Respiratory rate – 16/min.

Temperature – Afebrile.

Built – Lean thin.

No Icterus, No Cynosis, No Clubbing and No any lymphadenopathy.

Pallor – Present.

Systemic Examination: Abdomen – On inspection - Schaphoid in shape.

On palpation – Soft, not distended, no guarding, no rigidity, no tenderness, no palpable lump.

On percussion – Tympanic note.

On Auscultation – Normal bowel sounds present.

Investigations: Routine Blood Investigations:

Hb – Initially 6.0gm%, than 9.2gm% after 3 blood transfusions.

BT – 1.05 minutes

CT – 2.20 minutes

Blood Urea – 32mg/dl

Serum Creatinine – 0.9mg/dl

Fasting blood sugar – 84mg/dl

Serum Bilirubin

Total – 0.9mg/dl

Direct – 0.3mg/dl

SGOT – 27 IU/l

SGPT – 32 IU/l

Alk PO₄ – 94 IU/l

Serum Electrolyte

Sodium 140mEq/l

Potassium 4.4mEq/l

Radiological Investigations:

X-ray FPA – Distended stomach with air fluid level in the stomach and duodenum.

USG Abdomen-On 5.5.2013, normal study was reported.

On 5.7.2013, Dilated stomach, 1st, 2nd and proximal part of 3rd part of duodenum narrowing seen at distal part of third part of duodenum.

Barium Meal and follow through-On 17.6.13 no significant abnormality seen in BMFT study.

CECT abdomen (with oral and IV contrast)-Superior mesenteric vessels compressing the 3rd part of duodenum with proximal dilatation – SMA Syndrome

Upper GI Endoscopy-Stomach – Fundus, body, antrum and pylorus massively dilated.

Duodenum – 1st and 2nd part dilated.

Treatment: Preoperative Treatment: NBM/CRTS, TPN, Electrolyte imbalance corrected, Three Unit whole blood transfusion.

Operative management: Duodeno-jejunostomy (Side to side anastomosis of 2nd part of duodenum to jejunum).

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Intraoperative findings: Stomach, 1st and 2nd part of duodenum was grossly dilated while rest part of duodenum and jejunum was collapsed.

Outcome: Patient was discharged on 7th postoperative day with no complaint of vomiting; Patient was taking orally semi solids

Follow up: At follow up, patient had no complaint of vomiting and his general condition was good.

DISCUSSION: Wilkie's Syndrome is a very rare, life-threatening gastro-vascular disorder characterized by a compression of the third portion of the duodenum by the abdominal aorta (AA) and the overlying superior mesenteric artery. The syndrome is typically caused by an angle of 6°-25° between the AA and the SMA, in comparison to the normal range of 38°-56°, due to a lack of retroperitoneal and visceral fat. In addition, the aortomesenteric distance is 2-8 millimeters, as opposed to the typical 10-20.¹

SMA syndrome can present in acute, acquired form (e.g. abruptly emerging within an inpatient stay following scoliosis surgery) as well as chronic form (i.e. developing throughout the course of a lifetime and advancing due to environmental triggers, life changes, or other illnesses). Acute cases usually respond to medical management, while chronic cases require surgical intervention.²

In acute or mild cases, conservative treatment should be attempted first. Nasogastric tube placement for duodenal and gastric decompression and mobilization into the prone or left lateral decubitus position often is effective in the acute setting.³

Acute superior mesenteric artery syndrome involving the reversal or removal of the precipitating factor with proper nutrition and replacement of fluid and electrolytes, either by surgically inserted jejunal feeding tube, nasogastric intubation or peripherally inserted central catheter (PICC line) administering total parenteral nutrition (TPN). Pro-motility agents such as metoclopramide may also be beneficial.²

If conservative treatment fails, or if the case is severe or chronic, surgical intervention is required. The most common operation for SMA syndrome, duodenojejunostomy. Bypassing the compression caused by the AA and the SMA.¹

Less common surgical treatments for SMA syndrome include Roux-en-Y duodenojejunostomy, gastro-jejunostomy, and anterior transposition of the third portion of the duodenum, intestinal de rotation, and division of the ligament to Treitz (Strong's operation).

The possible persistence of symptoms after surgical bypass can be traced to the remaining prominence of reversed peristalsis in contrast to direct peristalsis, although the precipitating factor (the duodenal compression) has been bypassed or relieved. Reversed peristalsis has been shown to respond to duodenal circular drainage—a complex and invasive open surgical procedure originally implemented and performed in China.⁴

In our case, we had done gastro-jejunostomy after which patient having no complaint of vomiting in regular follow up.

CONCLUSION: Wilkie's Syndrome is a rare disorder. Only 500 cases reported worldwide till date. Surgical treatment is the treatment of choice in long standing cases otherwise it may result in fatal complications like advanced malnutrition, electrolyte imbalance, gastric rupture, spontaneous upper GI bleeding or sudden cardiovascular collapse.

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Fig. 1: Intra-operative dilated duodenum

Fig. 2: Intra-operative duodeno-jejunosomybypass anastomosis done

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Date of Submission: 22/08/2014.
Date of Peer Review: 23/08/2014.
Date of Acceptance: 30/08/2014.
Date of Publishing: 13/09/2014.