Laparoscopic management of a complicated case of Wilkie’s syndrome: A case report

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ABSTRACT

INTRODUCTION: Superior mesenteric artery (SMA) syndrome also known as Wilkie’s syndrome is a rare condition caused by the entrapment of the third part of the duodenum between the aorta and the SMA. The incidence of Wilkie’s syndrome range between 0.013% and 0.3%. The normal angle between the aorta and SMA has been described to range between 38° and 65°, whereas in Wilkie’s syndrome this angle is reduced to less than 20° causing gastric outlet obstruction.

CASE PRESENTATION: We report a case of a previously diagnosed 43 year-old male with SMA syndrome, whom had been conservatively managed for 5-years for recurrent admissions with symptoms of gastric outlet obstruction. During his last admission, CT abdomen demonstrated gastric pnumatosus and portal venous gas requiring urgent surgical intervention. Duodenojejunostomy was successfully performed using laparoscopic technique.

DISCUSSION: SMA syndrome is thought to occur secondary to the rapid and excessive weight loss leading to the reduction of the mesenteric fat around the aorta and SMA, thereby reducing the normal angle between the two arteries.Conservative medical management is usually the first line of treatment in uncomplicated cases. Surgical management is usually reserved only after failed conservative management or complicated cases, at which time either an open or laparoscopic surgical approach is undertaken.

CONCLUSION: Surgical intervention is the mainstay in complicated cases of SMA syndrome and in refractory cases to conservative management. Advantages of laparoscopic approach over open surgery include rapid recovery time, reduced post-operative pain and shorter hospital stay.

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1. Introduction
Superior mesenteric artery (SMA) syndrome also known as Wilkie’s syndrome is a rare condition caused by the entrapment of the third part of the duodenum between the aorta and the SMA. The incidence range between 0.013% and 0.3% [1]. The normal angle between the aorta and SMA has been described to range between 38° and 65°, whereas in Wilkie’s syndrome the angle is reduced to less than 20° leading to gastric outlet obstruction [2]. Loss of retroperitoneal fat due to rapid weight loss, surgeries for spinal deformities and high insertion of ligament of Treitz are among the common etiological factors for SMA syndrome [3]. Treatment options vary from conservative medical management for uncomplicated cases to surgical treatment, which can be either through open approach or laparoscopic surgery [4]. The work has been reported in line with the SCARE criteria [5].

2. Case presentation
A previously diagnosed 43 years-old gentleman with SMA syndrome was re-admitted 2 weeks following discharge for a similar presentation of gastric outlet obstruction with multiple episodes of vomiting. He had a total of 12 admissions since being first diagnosed 6 years ago, all of which were medically managed. On his last admission, he was referred to our surgical team-care. On examination he was vitally stable, but appeared moderately dehydrated. Abdominal exam revealed a soft mildly distented abdomen with moderate tenderness in the epigastrium & periumbilical region; there was no rebound tenderness, and no abdominal guarding elicited. Bowel sounds were audible and hernia orifices were intact. Digital rectal exam revealed soft stools with no palpable masses. Hematological investigation demonstrated significantly raised lactate level at 6.1 mEq/L, but otherwise all other hematological tests were within the normal range. CT scan of the abdomen and pelvis reported evidence of a complicated gastric outlet obstruction with gastro-duodenal pneumatosus, as well as right and left intrahepatic portal venous pneumatosus (Fig. 1). Initial medical management included nil by mouth, insertion of nasogastric (NG) tube to decompress the
stomach, fluid and electrolyte replacement and optimizing the general condition of the patient pre-op using a multidisciplinary team approach.

Laparoscopic duodenojejunostomy was discussed with the patient and consent was obtained (Fig. 2). Approximately 40 cm of the jejunum from the duodenojejunal junction was brought up for anastomosis with the distended third part of the duodenum. The post operative period was uneventful and the patient started soft diet by day-2 after methylene blue injection through the nasogastric tube confirmed the integrity of the anastomosis. The drain was removed on day-3 post-op. The patient was discharged to home the following day with a follow up appointment in outpatient clinic.

3. Discussion

SMA syndrome is caused by the compression of the third part of the duodenum when the angle formed by the aorta and SMA decreases to less than its normal range. It is thought to be due to the rapid and excessive weight loss that leads to the loss of the mesenteric fat around the artery [1,2]. Other causes include high insertion of the ligament of Treitz and post surgical surgeries for the correction of spinal deformities [3].

Uncomplicated cases of SMA syndrome are generally conservatively managed in the first instance with the aim of treating the symptoms of small bowel obstruction. These include gastric decompression by insertion of NG tube, rehydration & electrolyte replacement. Moreover, dietician input is essential to prevent inappropriate weight loss leading to symptom exacerbation and restoration of appropriate weight to conserve the normal angle between the two arteries involved to avoid recurrence [4]. Surgical management is usually only considered once conservative management fails or complications arise. Several surgical options are available with duodenojejunalstomy being the commonest procedure with a good success rate (over 90%). A less common surgical approach is the gastrojejunostomy due to the blind loop syndrome and the persistence of symptoms due to the non-decompression of the duodenum. Another surgical method is the division of the ligament of Treitz, which is less invasive but its main disadvantage is the recurrence rate that may be up to 25% [6]. All of these surgical procedures can be performed either through open surgery or laparoscopic technique. In our case we opted for a laparoscopic duodenojejunalstomy after optimizing the general condition of the patient as it has a faster recovery time, reduction in post-op pain and earlier return to daily activities.

An important note to make is that given the potential life threatening complications that can arise from SMA syndrome, surgical approach should not be deferred until the very late stage of the disease. Clinical decision must be undertaken early on when conservative medical management fails in order to treat the condition without the onset of complications thereby avoiding emergency surgery.

4. Conclusion

Conservative management is the standard treatment for uncomplicated cases of Wilkie’s syndrome. Due to the potential life-threatening complications surgery becomes unavoidable and the only therapeutic option. Taking in consideration the faster recovery, shorter hospital stay and earlier return to daily life activities, laparoscopic surgical approach outweigh the open surgery for treating Wilkie’s syndrome.

Conflicts of interest
Nothing to declare.

Funding
Nothing to declare.

Ethical approval
None required.

Fig. 2. Laparoscopic images of duodenojejunostomy procedure.
A- Final repair of the duodenojejunostomy.
B- Testing the integrity of the anastomosis by injecting methylene blue under pressure into the nasogastric tube.
Consent
Written informed consent was obtained from the patient for the publication of this case report and accompanying images and video. A copy of the written consent is available for review by the editor-in-chief of this journal on request.

Authors contribution
Dr. Wisam Al-Ramli: Surgical Intern. Responsible for literature review, writing and manuscript preparation.
Dr. Yahya Khodear: Surgical Registrar. Assisted in the laparoscopic surgery. Responsible for literature review, writing and manuscript review.
Mr Zsolt Bodnar: Consultant Surgeon. Performed the laparoscopic surgery. Responsible for manuscript review.

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Guarantor
Dr Wisam Al-Ramli.
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