Wilkie's Syndrome as a Rare Cause of Upper Gastrointestinal Obstruction in a Thin Young Lady

Zayıf Bir Genç Bayanda Nadir Bir Üst Gastrointestinal Obstrüksiyon Nedeni Olarak Wilkie Sendromu

Annisa Binti Zainal Mokhtar, Chik Ian, Zamri Zuhdi

National University of Malaysia, Department of General Surgery, Faculty of Medicine, Kuala Lumpur, Malaysia

ABSTRACT

Superior mesenteric artery (SMA) syndrome is a rare cause of upper gastrointestinal (UGI) obstruction. Also known as Wilkie's syndrome, it is caused by compression of third part of duodenum due to narrowing of the angle between SMA and abdominal aorta which results in symptoms of duodenal outflow obstruction. The cause must be sought and treat. Our patient is a thin 21-year-old lady with no known medical illness, who presented with 3 months history of post prandial bilious vomiting associated with dysphagia, loss of weight and loss of appetite. Abdominal examination revealed distended abdomen with positive succusion splash. Blood investigations showed that she was anaemic, had hyponatraemia and hypokalaemia. Abdominal x-ray revealed dilated stomach. Oesophagogastroduodenoscopy (OGDS) revealed dilated stomach filled with food particles, with no evidence of mechanical cause of her dysphagia. Computed Tomography (CT) abdomen done revealed partial obstruction at third part of duodenum with narrowed angle between abdominal aorta and SMA. Diagnosis of SMA syndrome was made. Laparoscopic converted to open gastrojejunostomy was done. Post operatively, patient developed gastroparesis and was treated with metoclopramide. She was discharged well after 10 days of operation. SMA syndrome is diagnostically challenging as there are many disorders which mimic its clinical presentation. Although rare, it should be kept in mind as a cause of UGI obstruction as it is potentially curable.

Keywords: SMA syndrome, upper gastrointestinal obstruction, Wilkie's syndrome

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ÖZET

Superior mezenterik arter (SMA) sendromu, üst gastrointestinal (ÜGİ) obstrüksiyonun nadir bir nedenidir. Wilkie sendromu olarak da bilinir, SMA ile abdominal aorta arasındaki açının daralması nedeniyle duodenumun üçüncü kısmının sıkışması sonucu duodenal çıkış obstrüksiyonu semptomları ile sonuçlanır. Nedeni aranmalı ve tedavi edilmelidir. Hastamız 21 yaşında, bilinen bir tıbbi hastalığı olmayan, 3 aydır yutma güçlüğü, kilo kaybı ve iştahsızlık ile ilişkili yemek sonrası safralı kusma öyküsü olan ince bir bayandır. Karın muayenesinde pozitif sukuzyon sıçraması ile şişkin karın saptandı. Kan tetkikleri anemik olduğunu, hiponatremi ve hipokalemisi olduğunu gösterdi. Karın röntgeni genişlemiş mideyi ortaya çıkardı. Özofagogastroduodenoskopide (OGDS), gıda parçacıklarıyla dolu dilate mideyi ortaya çıkardı ve hastanın disfajisinin mekanik nedenine dair hiçbir kanıt yoktu. Yapılan Bilgisayarlı Tomografi (BT) karında, duodenumun üçüncü kısmında abdominal aort ile SMA arasındaki açıda daralmış kısmi obstrüksiyon saptandı. SMA sendromu tanısı konuldu. Laparoskopik olarak açık gastrojejunostomiye çevrildi. Ameliyat sonrası hastada gastroparezi gelişti ve metoklopramid ile tedavi edildi. 10 günlük operasyonun ardından şifa ile taburcu edildi. SMA sendromu, klinik sunumunu taklit eden birçok bozukluk olduğu için tanısal olarak zordur. Nadir olmasına rağmen, potansiyel olarak tedavi edilebilir olduğu için UGI obstrüksiyonunun bir nedeni olarak akılda tutulmalıdır.

Anahtar Sözcükler: SMA sendromu, üst gastrointestinal obstrüksiyon, Wilkie sendromu

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ORCID IDs: 0000-0002-7350-8134, 0000-0001-9768-7379, 0000-0003-3634-1854

Address for Correspondence / Yazışma Adresi: Annisa Zainal Mokhtar, MD Department of General Surgery, 8th Floor, Hospital Canselor Tuanku Muhriz UKM, Jalan Yaacob Latif, Bandar Tun Razak, 56000 Cheras, Kuala Lumpur, Malaysia. E-mail: carbonaraXX@gmail.com

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INTRODUCTION

Wilkie's syndrome is a rare cause of upper gastrointestinal (UGI) obstruction. Commonly known as SMA syndrome, patients usually present with symptoms of UGI obstruction and this pose a diagnostic challenge to this syndrome as the causes are vast. We presented a case of thin-built young lady who was diagnosed with SMA syndrome and the discussion on our line of management.

CASE REPORT

Our patient is a 21-year-old lady who is previously well presented with three months history of post-prandial bilious vomiting, associated with dysphagia, loss of appetite and weight loss. She has always been thin with BMI of 16.23. On examination, she was dehydrated. Her abdomen was soft but distended with presence of succussion splash. At this point, our differential diagnoses include peptic ulcer stricture, duodenal lymphoma and adhesion band over duodenum.

Blood investigations revealed anaemia with haemoglobin of 9.9 g/dL. She had mild hyponatraemia and hypokalaemia. Her urea, creatinine and albumin were normal.

Her stomach was dilated in abdominal x-ray [Figure 1]. Oesophagogastroduodenoscopy (OGDS) revealed dilated stomach filled with food particles, with no evidence of mechanical cause of her dysphagia. Computed Tomography (CT) abdomen showed partial obstruction at the third part of duodenum with aortomesenteric angle of 11 degrees and aortomesenteric distance of 2.69 mm [Figure 2a and Figure 2b]. Thus, diagnosis of superior mesenteric artery syndrome was made.

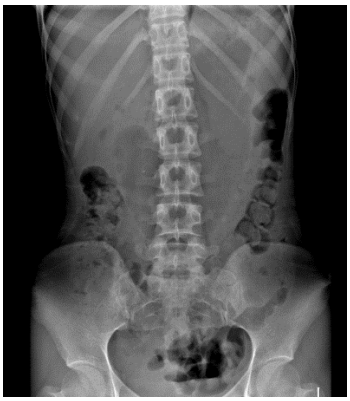
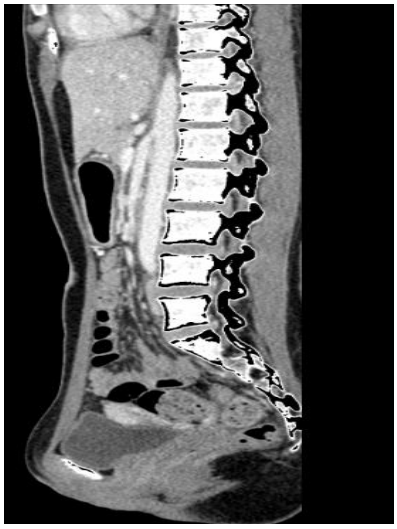


Figure 1 showing abdominal x-ray of this patient with dilated stomach.



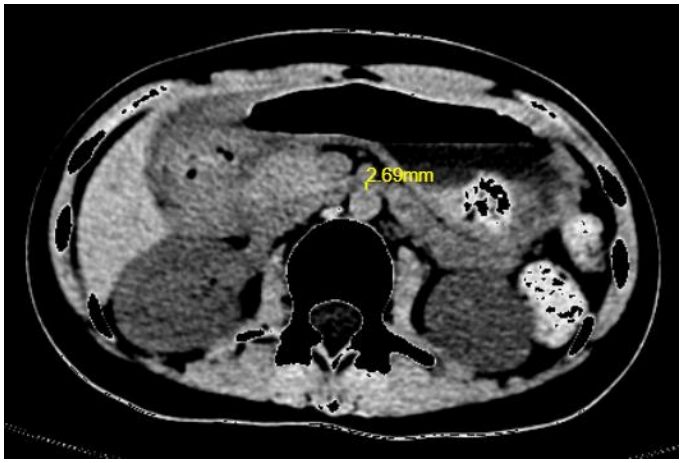


Figure 2a showing aortomesenteric angle of 11 degrees, and **Figure 2b** showing distance of 2.69 cm between and SMA and abdominal aorta. White arrow indicates dilated duodenum proximal to the obstruction.

The patient was kept nil by mouth, given fluid resuscitation with correction of electrolytes abnormality. Nasogastric tube was inserted to decompress the stomach. She was started on total parenteral nutrition (TPN). Laparoscopic gastrojejunostomy was planned initially. Upon entering the abdomen, there was enlarged stomach with transverse colon pushed downwards, reaching the pelvis. Dilated duodenum was seen proximal to SMA [Figure 3]. Jejunum was transected at 40 cm from duodenojejunal junction. While performing antecolic gastrojejunostomy, there was difficulty in doing intracorporeal suturing. Thus, the operation was converted to open. The rest of the operation was completed uneventfully.

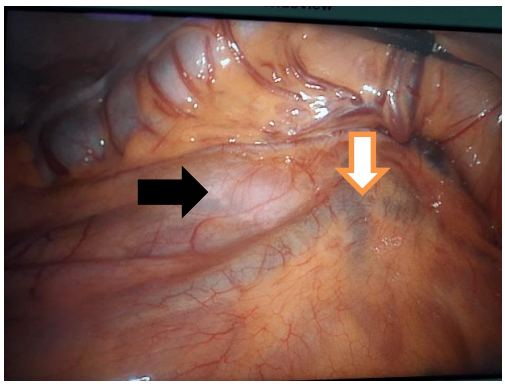


Figure 3 showing SMA (white arrow) with dilated duodenum proximal to it (black arrow).

Postoperatively, TPN recommenced. Patient was allowed initially clear fluid, followed by nourishing fluid and soft diet orally from day 2 till day 5 in stages. However, at day 6 post surgery, patient vomited multiple times and was unable to tolerate feed. Gastrograffin study revealed slow transit of contrast from stomach through the gastrojejunostomy with dilated stomach. There was no stricture or leak over the anastomosis. Treating as gastroparesis, she was given regular metoclopramide and subsequently was able to tolerate orally with no more vomiting. She was discharged well at day 10 post surgery.

DISCUSSION

SMA syndrome is a rare entity with estimated incidence of 0.1 to 0.3%. Majority occurred in adolescents and young adults with female to male ratio of 3:1. It is defined as compression of third part of duodenum due to narrowing of angle between abdominal aorta and superior mesenteric artery causing obstruction of duodenum. It was first described by Austrian professor Carl von Rokistansky in 1842 and Wilkie in 1927 published the first comprehensive series of patients with this syndrome(1). It is postulated that loss of intervening fat pad surrounding SMA causing reduction of aortomesenteric angle causing compression of the third part of duodenum is the main cause of SMA syndrome. Our patient has always been had low BMI. Although she had no prior symptoms suggestive of SMA syndrome, low fat tissue in our patient could be the aetiology of her developing SMA syndrome.

Clinical examination of our patient revealed UGI obstruction. CT abdomen showed SMA syndrome as the cause. Normal aortomesenteric angle is 28 to 65 degrees while normal aortomesenteric distance is 10 to 34mm(2,3). In our patient, these measurements are greatly reduced.

Initial conservative management include stomach decompression, correction of dehydration and electrolytes imbalances and nutritional support. There is no time limit for conservative management, but symptom relief has been reported between two to twelve days. Due to advancement of enteral and parenteral nutrition in the recent years, more patients has been treated successfully with conservative management(4). However, surgery was planned early for our patient as she has always been thin with minimal peri-SMA fat pad and will normally took small amount of food. Although weight gain can be promoted through hyperalimentation, once patient commences her previous eating habit with stoppage of nutritional support, there is high chance of recurrence.

Although duodenojejunostomy is widely accepted as the procedure of choice, in our case, gastrojejunostomy was performed due to surgeon's familiarity with the procedure. Gastrojejunostomy in this patient also has the advantage of avoiding potential higher risk of anastomotic leak if the anastomosis is done at dilated region of duodenum near the area of compression.

CONCLUSION

SMA syndrome is diagnostically challenging as there are many disorders which mimic its clinical presentation. Although rare, it should be kept in mind as a cause of UGI obstruction as it is potentially curable and once diagnosed, the cause should be sought and corrected.

Conflict of interest
No conflict of interest was declared by the authors.

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