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A rare instance of Wilkie's syndrome in a young male during the holy month of Ramadan

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ABSTRACT

INTRODUCTION AND IMPORTANCE: Wilkie's syndrome (SMA (Superior Mesenteric Artery) syndrome or Cast syndrome) is a unique and rare presentation which may be included in the differential diagnosis especially when a Gastric outlet obstruction is being investigated and there is a history of an associated element of rapid weight loss. The purpose of presenting this case report is its uniqueness given a history of intermittent fasting rather than the usual eating disorders.

CASE PRESENTATION: A crescendo worsening of symptoms of intractable vomiting, inability to eat, upper abdominal gas bloating and post meal abdominal pain since one week in a patient during a period of fasting for the first time had been observed.

CLINICAL DISCUSSION: After initial resuscitation, an ultrasound abdomen and an abdominal x ray revealed a distended stomach and a nasogastric tube was immediately introduced to avoid aspiration. A CT scan of the abdomen with contrast revealed the compressed duodenum in the aortomesenteric angle and the diagnosis of SMA syndrome was confirmed. An upper gastrointestinal endoscopy was also done to rule out other causes and it was found to be normal.

CONCLUSION: In an acute presentation, the response to conservative management is remarkable and in the absence of any possible future reoccurrences the surgical options may be avoided. High index of suspicion, appropriate radiological diagnosis and stepwise treatment options may guide the troubled patient towards recovery in this rare presentation. A proper history taking is key in diagnosing such a condition and does not have to be limited to eating disorders, typically.

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1. Introduction

The case discusses duodenal obstruction due to Wilkie's syndrome (also known as SMA-Superior Mesenteric Artery syndrome, Cast syndrome) in a young male due to rapid loss of weight during the Holy month of fasting. Presentation to the Emergency department called for an immediate exploration of the abdomen using various methods like that of ultrasound, X-ray of the abdomen and a contrast enhanced CT scan which revealed a diagnosis of SMA syndrome. An initial approach of conservative management involving rehydration, appropriate modification in nutrition alongside intermittent usage of nasogastric tube in the case of episodes of vomiting and gastric distension proved to improve the condition of the patient. Also, the patient remained asymptomatic with no further complaints during subsequent sessions of follow-up. The patient was completely managed in a community hospital through-

out the process of admission and discharge. The patient further continued to follow up at the same institution.

Duodenal obstruction due to SMA (Superior Mesenteric Artery) syndrome, also known as Wilkie's or Cast syndrome is a rare presentation and since there is no absolute criteria for the diagnosis of this condition, the purpose of presenting this case is to help identify high risk individuals while approaching patients in a clinical setting. A history of intermittent fasting alongside the aid of diagnostic tools has proven to rule in for such a condition. The case discussion involves an Emergency patient encounter of a young male brought in with intractable vomiting, bloating and inability to eat with symptoms worsening over a period of 1 week. A history of fasting for the first time during the holy month was provided and the patient was dehydrated as a result. Immediate patient exploration also involved radiological diagnosis to confirm the causes of a gastrointestinal tract obstruction and rule out other possible causes. An integrated approach by taking into account the clinical manifestations, history such as rapid loss of weight [even without necessarily corresponding to a low BMI (Body Mass Index)] and radiological findings together must guide physicians towards considering such a possible diagnosis in high risk patients. This article

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describes the method of treatment and the first line management which is always a conservative approach by modifying nutritional requirements. Surgical management is later considered in those where the condition either fails to respond or recurrent episodes of duodenal obstruction occur. Hence, it is key that conservative approach be held superior to surgical management in such a case and physicians and surgeons work as a team in order to establish best patient outcomes.

2. Patient Information

2.1. De-identified patient demographics

During the Holy month of fasting, a young male patient presented to the emergency department. The patient appeared as a lean individual.

2.2. Complaint and presentation

The patient presented with complaints of intractable vomiting and inability to eat. He was also having post meal abdominal pain and upper abdominal gas bloating. All his symptoms were since 1 week and had progressively increased in severity. He was dehydrated and exhausted secondary to the on going fasting and also due to the repeated vomiting.

The parents of the patient pointed out that this was his first time of religious fasting and he had been compliant to the religious customs. Weight loss had not been a problem suggested by the parents as it is expected during this period. The patient did not have any relevant past history or intake of medications.

2.2.1. Past medical history and outcomes from intervention

The patient did not have any relevant past medical history other than intermittently fasting. There was no history of drug intake since five days before admission. After initial resuscitation, an ultrasound abdomen and an abdominal x ray revealed a distended stomach and a nasogastric tube was immediately introduced to avoid aspiration. A CT scan of the abdomen with contrast revealed the compressed duodenum in the aortomesenteric angle and the diagnosis of SMA syndrome was confirmed. An upper gastrointestinal endoscopy was also done to rule out other causes and it was found to be normal.

A conservative management was initiated with ongoing correction of dehydration and electrolyte imbalances and nasogastric feeding was encouraged. Over a period of 1week the symptoms regressed and the tube was removed. The patient could tolerate diet gradually which was increased stepwise monitoring his improvement. Once stable and able to tolerate diet the patient was discharged for follow up. The patient was followed up for six months post discharge. On follow up, the patient remained asymptomatic with no further complaints.

The patient did not have any history of a recent intake of medications, family history, psychosocial history or habitual smoking history.

3. Clinical Findings

3.1. Timeline

The patient had complained of and experienced the above mentioned symptoms for a period of one week prior to presentation at the emergency. The set of symptoms had only gradually worsened over the same period of the week. The patient was managed conservatively without any surgical intervention and was followed

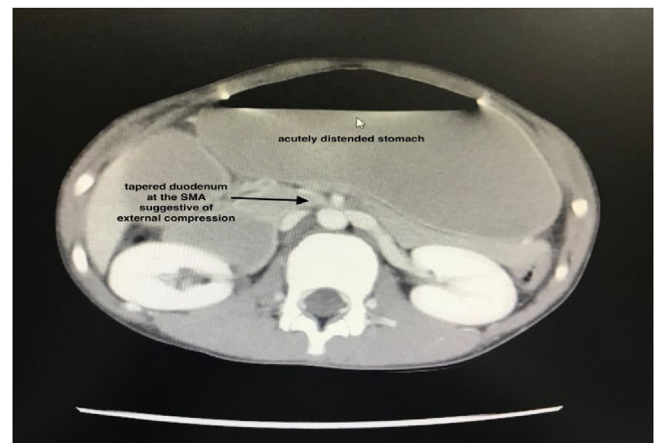


Fig. 1. CT scan of the abdomen on the axial plane demonstrating distension of the stomach and the initial portion of the duodenum thereby causing compression of the third part of the duodenum situated between the Superior Mesenteric Artery (SMA) and the abdominal aorta.

up for a period of six months post his discharge and remained asymptomatic without any further complaints.

4. Diagnostic assessment

4.1. Diagnostic methods

After initial resuscitation, an ultrasound abdomen and an abdominal x ray revealed a distended stomach and a nasogastric tube was immediately introduced to avoid aspiration. A CT scan of the abdomen with contrast revealed the compressed duodenum in the aortomesenteric angle and the diagnosis of SMA syndrome was confirmed (Figs. 1 and 2). An upper gastrointestinal endoscopy was also done to rule out other causes and it was found to be normal. No single investigation of choice exists.

4.2. Diagnostic challenges

There weren't any diagnostic challenges encountered as the patient was co-operative enough to provide relevant history and had agreed to undergo the endoscopic procedure.

4.3. Differential diagnoses and diagnostic reasoning

The following differential diagnoses were considered given the above presentation:

- Gastric outlet obstruction
- Peptic stricture of the stomach
- Neoplasm of the stomach

The aortomesenteric artery angle and aortomesenteric distance have proven to be of great significance in the diagnosis of SMA syndrome. Hence, CT scans were used to rule in for duodenal obstruction due to a reduced aortomesenteric angle in the patient. The normal angle made between the aorta and superior mesenteric artery lie between the range of 25–60 degree and the normal distance lies between 10–28 mm [17].

Post initial resuscitation, an ultrasound of the abdomen and an abdominal X-ray revealed a distended stomach and a nasogastric tube was introduced alongside to prevent aspiration. An upper GI endoscopy was also performed to rule out other causes like annular pancreas, diabetic gastroparesis, internal hernia or duodenal bezoar. It is important to rule out all other conditions that closely



Fig. 2. CT sagittal section demonstrating the angle between the Superior Mesenteric Artery(SMA) and the aorta(the aortomesenteric angle)to be 8-9 degree and the aortomesenteric distance to be 2-3 mm.

mimic SMA syndrome ranging from scleroderma to other collagen and vascular disorders, or other causes of duodenal obstruction like peptic ulcer disease and duodenal tumors [18,19]. A high level of suspicion for SMA syndrome arises in the patient due to a recent history of fasting for the first time.

4.4. Prognostic characteristics

The patient's condition responded by conservative management which included correcting the dehydration and electrolyte imbalances, gastric decompression with nasogastric tube was also beneficial. Since the patient had episodes of vomiting between normal periods of being able to eat, a nasogastric tube was used intermittently. Thus, a conservative approach was successful given that the symptoms appeared only a week ago and as a result a sooner body weight accommodation was possible [10,23].

5. Therapeutic Intervention

5.1. Pre-intervention considerations

The patient did not undergo any surgical intervention and instead was asked to regain the lost body weight by making changes to the diet. Thus the patient was checked with a conservative man-

agement which proved to be of use and hence did not require surgical correction by any of the discussed methods.

5.2. Types of intervention

NA. No intervention

5.3. Peri-intervention considerations

Dietary management and regaining body weight was the first line therapy and intervention by surgical means was not required.

5.4. Who performed the procedure

No procedure was performed.

5.5. Any changes in intervention

NA

5.6. Post intervention consideration

NA

6. Follow up and outcomes

6.1. Assessment and outcomes

The patient was discharged once rehydrated and the correction of electrolyte imbalances was made during the period of admission over one week. The patient did not have any complaints and remained asymptomatic which helped initiate discharge.

6.2. Follow up and measures

In order to prevent any episodes of relapse, the patient was followed up for a period of six months post his discharge from the hospital.

6.3. Intervention adherence and tolerability

No intervention performed

6.4. Complications and adverse effects

The patient did not have any adverse effects since no surgical intervention was performed. The patient tolerated the medical management by correcting the calorie intake and making appropriate dietary modifications to prevent any relapse.

7. Discussion

7.1. Limitations, strengths and weaknesses of the case report

As the patient was managed conservatively and no surgical interventions were performed, this writing cannot hold any surgical method superior to another.

This case report confirms that medical management is first line given a case of gastric outlet obstruction due to rapid weight loss and this approach must be tried before any surgical correction.

7.2. Relevant literature

The SMA syndrome is a rare condition and the prevalence is estimated to be 0.013% to 0.3% in the general population [1]. The

duodenum makes an acute angle with the aorta and is found between the aorta and the superior mesenteric artery. A pad of mesenteric and retroperitoneal fat envelopes this region. In the case of rapid weight loss, there is a loss of fat in the same region. The distance along with the angle between these anatomical structures decreases. The third part of the duodenum that passes between the arterial vessels is compressed eventually and this results in some of the clinical manifestations of this condition like duodenal obstruction, abdominal pain, vomiting, bloating and inability to eat as a result [2,3].

BMI does not accurately indicate a measurement of the visceral fat tissue volume and hence the Gold standards for measuring it are CT and MRI [22]. Compression of the third part of the duodenum may be visible upon endoscopy of the upper gastrointestinal tract [4]. The usual aortomesenteric angle usually lies between 25–60 degrees and the aortomesenteric distance is normally around 10–28 mm [2,5,6].

Symptoms progress as the weight loss increases to extrinsic partial or complete compression of the duodenum (D3) resulting in chronic food intolerance, bloating, nausea, eructation, abdominal discomfort, and voluminous bilious vomiting [7,8].

As an additional point to note the Nutcracker Syndrome may be coexistent, where in the left renal vein gets compressed in between the aorta and the SMA along with the duodenum. The patient may remain asymptomatic or present with renal symptoms and even occasionally a left sided varicocele [9]. Progressive radiographic investigations include abdominal X-rays, barium studies, and CT scan of the abdomen. The definitive diagnosis can be made only by CT or MRI angiography wherein the anatomic detail outlining the aorto-mesenteric angle can be best appreciated. An angle of ≤ 25 degrees associated with an aorto-mesenteric distance of ≤ 8 mm can be diagnostic of the Wilkie's syndrome. A duodenal obstruction with an abrupt cut off in the third portion and active peristalsis is essential to establish the resultant Gastric outlet obstruction. Along with this, the finding of a high fixation of the duodenum by the ligament of Trietz, abnormally low origin of the SMA or anomalies of the SMA may also be seen [10–14].

The risk factors for SMA (Superior Mesenteric Artery) syndrome range from eating disorders like anorexia to even burns or rapid weight loss (even if the weight loss is relatively small which may lead to a loss of mesenteric or retroperitoneal fat, chronic or malignant conditions. A high insertion point of the ligament of Treitz, the use of spinal casts, excessive lumbar lordosis, trauma and even substance abuse [15,16].

Although both conservative treatment and surgical management have proven to be useful in the case of duodenal obstruction from SMA syndrome, we proceeded with the first line treatment of conservative approach through regaining some amount of weight in the patient. Thus, conservative management alongside positional maneuvers like lying in the left lateral decubitus position and knee chest position have proven to reduce the symptoms.

Only when the patient relapses to SMA syndrome further leading to recurrent episodes of duodenal obstruction or often when the individual fails to regain the required amount of weight, several surgical methods of intervention are considered like laparoscopic duodenojejunostomy, gastrojejunostomy and Strong's procedure [20]. Of these methods, the one proven to have a high success rate (90 %) and considered superior to all other forms is duodenojejunostomy [21].

7.3. Conclusion

The purpose of reporting this case is to emphasize the importance of holding a high index of suspicion in diagnosing for SMA syndrome causing duodenal obstruction by considering multiple factors ranging from clinical manifestations to confirming the radi-

ological diagnosis with a given history of fasting and dehydration. This case report is being published to highlight that such a condition may exist given a history of intermittent fasting alone and not inclusive of any eating disorder.

7.4. Take away lessons

Although there remains no exact range of values for the aortomesenteric angle and distance between the aorta and superior mesenteric artery or even specific low BMI (Body Mass Index) range as criteria to confirm such a diagnosis, one must consider a differential diagnosis of SMA syndrome as a possible cause for the duodenal obstruction. Given such a presentation of post meal abdominal pain, intractable vomiting, bloating of the abdomen, inability to eat alongside a history of intentional fasting leading to weight loss must hint a physician of such a possible diagnosis. Most importantly, the efficiency of the methods of management (either conservative or surgical) must be monitored with follow up sessions as recurrence of Wilkie's syndrome (SMA or Cast syndrome) is usually due to unsuccessful outcomes of the condition with worsening symptoms. In cases of recurrent episodes even post surgery, corrections have been made by converting a previous duodenojejunostomy to a Roux-en-Y-duodenojejunostomy [21].

This manuscript is reported in line with the SCARE 2020 criteria [24].

Patient perspective

NIL.

Declaration of competing interest

None.

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Ethical approval

Approval given.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

First author-Contribution to this case report involves manuscript writing, reviewing and organizing the final draft.

Corresponding author-Specialist general surgeon involved in the manuscript and treatment of this case.

Registration of research studies

Not applicable.

Guarantor

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