SUPERIOR MESENTERIC ARTERY SYNDROME FOLLOWING CORRECTIVE SPINAL SURGERY FOR SCOLIOSIS

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ABSTRACT

Superior Mesenteric Artery (SMA) syndrome is a well-known but rare complication following corrective surgery for scoliosis. It can be easily missed if not thought of in patients presenting with abdominal pain and vomiting post-scoliosis surgery. SMA syndrome is most commonly associated with conditions resulting in significant weight loss but can also occur as a result of biomechanical changes following scoliosis surgery. We present a case of a 13-year-old girl who developed SMA syndrome following scoliosis surgery. A high index of suspicion and early surgical referral are paramount for the successful management of SMA syndrome.

Keywords:
Superior Mesenteric Artery Syndrome; Adolescent Idiopathic Scoliosis; Nausea; Vomiting; Abdominal Pain; Complication;

SFP2017; 43(1): 52-56

PATIENT'S REVELATION: WHAT HAPPENED?

SN is a 13-year-old girl who presented with acute abdominal pain with vomiting. Her background included adolescent idiopathic scoliosis (AIS) for which she recently underwent posterior instrumentation and fusion of T2 to L4.

Her abdominal pain had started since post-operative day 24 and had been ongoing for 11 days. It was dull and crampy, particularly over the left iliac fossa (LIF), occurring within minutes after meals, with a maximum pain score of 7/10, lasting for about 15 minutes and then spontaneously resolving. The abdominal pain was absent when she did not eat. This was associated with a history of non-bloody, non-bilious, non-projectile vomiting after meals 2 days prior to her presentation. She also complained of constipation for the past 3 to 4 days. There was no fever, headache, urinary symptoms or gastro-intestinal bleeding. There was no other recent illness, sick contacts or travel history. She was increasingly unable to tolerate any food at home which prompted her parents to bring her to the Children's Emergency (CE).

On examination, SN is a tall, skinny girl with a BMI of 14.2 at less than 1st centile (weight 38.7kg at 20th centile, height 1.65m at 97th centile). Of note, her pre-operative BMI taken 4 months ago was 16.2 at 14th centile (weight 40kg at 35th centile, height 1.57m at 75th centile).

In CE, her vital signs were: temperature 37.2^{0}C, respiratory rate 18, pulse oximetry 100 percent on room air, heart rate 110 and blood pressure 105/74mmHg. She was moderately dehydrated. Cardiorespiratory examination was normal. Her abdomen was scaphoid and soft with no guarding or significant tenderness. Bowel sounds were active. The rest of the examination was normal. Urine pregnancy test was negative and a finger-prick blood sugar level was 5.2mmol/L. Urine dipstick was unremarkable.

Failing a trial of oral rehydration overnight in the CE with sublingual ondansetron, SN was admitted for IV hydration under the provisional diagnosis of viral illness with vomiting with possible constipation colic.

Subsequent blood investigations showed white blood cell count 13.27 x 10^9/L, mild anaemia with haemoglobin of 10.7 g/dL and absolute neutrophil count of 10.04 x 10^9/L (75.8%). The rest of the full blood count was normal. Electrolytes, acid-base studies, and renal function were normal. Abdominal X-rays demonstrated faecal loading in the right iliac fossa and a prominent gastric bubble but no other dilated bowel loops or air-fluid levels indicative of bowel obstruction (Figure 1). She was given lactulose for constipation but with little improvement.

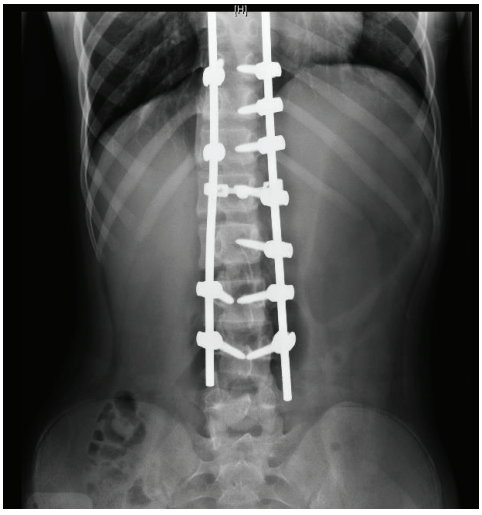

Figure 1: Abdominal X-ray of patient SN demonstrating a prominent gastric bubble.

On repeat examination the following morning by the general paediatrics team, hyperactive and high-pitched tinkling bowel sounds with positive gastric succussion splash were noted. This aroused the suspicion of bowel obstruction, particularly at a proximal level. The provisional diagnosis was changed from a viral illness to proximal intestinal or gastric

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outlet obstruction — probably SMA syndrome particularly related to her recent scoliosis operation and also in view of her skinny frame and pattern of abdominal pain with vomiting closely related to meals. The duration of symptoms and absence of fever or significant haematological derangements made a viral illness or other infections less likely. Although the pain of SMA syndrome should be more epigastric and not usually in the LIF as in this case, it was still high on the list of differentials given the clinical context. Constipation colic was also thought to have contributed to her clinical picture although the laxatives did not prove to be effective and it could not explain the vomiting.

She was kept nil-by-mouth, placed on drip and underwent an urgent barium meal to look for SMA syndrome. To the team's excitement, barium meal results returned corroborating our provisional diagnosis (Figure 2).

Figure 2: Barium meal and follow-through for patient SN: Stasis of contrast at the third part of the duodenum which was mildly dilated at 4cm in both semi-recumbent and standing positions, with contrast flow seen from the duodenum into the jejunum after patient was put in the left decubitus position, making the diagnosis of SMA syndrome highly likely.

The barium study demonstrated persistent hold-up of contrast both in the semi-recumbent and standing position at the third part of the duodenum (D3). Contrast flow resumed once the patient was put in the left decubitus position, which was highly suggestive of SMA syndrome. The paediatric surgical team was promptly consulted.

Although further investigations could have been done to more accurately assess the angle between the SMA and aorta, they were felt to be unnecessary by the surgical team as SN did not require active surgical intervention because she was improving with conservative medical treatment. She made gradual progress with small, frequent feeds orally and was also advised on changes to feeding position such as turning to the left decubitus position, or sitting forward or knees to chest to facilitate flow of food past the obstruction and relieve symptoms from gastric distension. SN was prescribed a high-calorie liquid diet and was discharged after 4 days when her symptoms improved.

GAINING INSIGHT: WHAT ARE THE ISSUES?

Several issues arose as a result of this case:

- How does SMA syndrome develop and how did it develop in this patient?
- What are the clinical features of SMA syndrome and are there red flags which should alert primary care providers to its diagnosis?
- What is the treatment for patients with SMA syndrome?

STUDY THE MANAGEMENT: HOW DO WE APPLY THE INSIGHTS IN OUR CLINICAL PRACTICE?

How Does SMA Syndrome Develop and How Did It Develop in this Patient?

SMA syndrome results from compression of D3 due to narrowing of the space between the SMA and aorta (Figure 3).

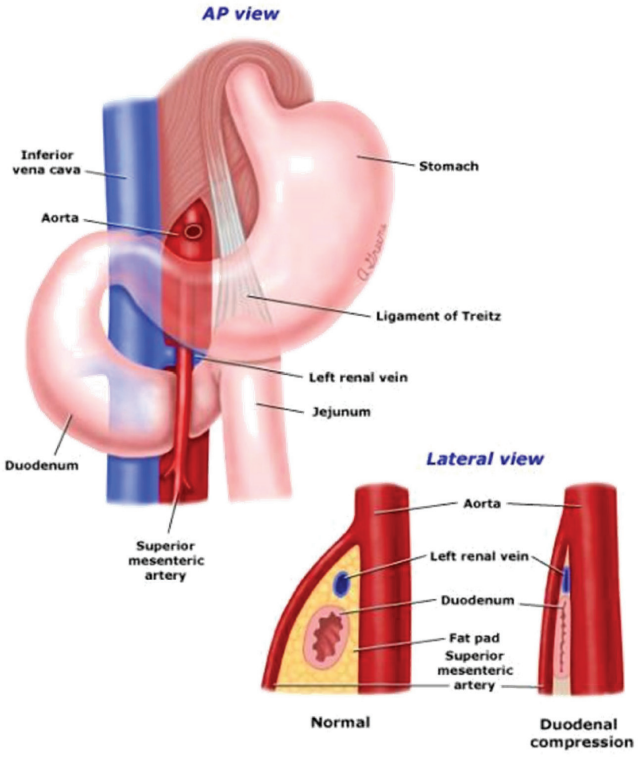

Figure 3: Superior Mesenteric Artery (SMA) syndrome.[4] The third part of the duodenum passes between the aorta and the SMA at the L3 vertebral level suspended by the ligament of Treitz. The aorto-mesenteric distance is normally 10 to 28 mm and the normal angle between the SMA and the aorta is between 38° and 65° and correlates with body mass index.[1,2,3] In SMA syndrome, due to the loss of the intervening mesenteric fat pad, the angle can be narrowed to as low as 6° with the aorto-mesenteric distance as low as 2 mm, potentially leading to duodenal compression.[4]

SMA syndrome is most commonly associated with conditions

causing significant weight loss such as malignancy, malabsorption syndromes, AIDS,[5] trauma,[6] burns,[7] bariatric surgery,[8] spinal cord injury, paraplegia,[9] drug abuse,[10] prolonged bed rest, and anorexia nervosa and other eating disorders.[11]

However, weight loss may not always be present. Interestingly in younger patients as described in our case, SMA syndrome is known to most commonly occur following corrective surgery for scoliosis, with an incidence of 0.5 percent to 4.7 percent.[3,12-18] Tall, thin, and asthenic adolescents with low BMI who undergo spinal manipulation and curvature correction by instrumentation, traction, casting or bracing are at highest risk.[17,19] This is because these patients with AIS usually present during the time of their most rapid longitudinal skeletal growth which can decrease the aorto-mesenteric angle and therefore increase the risk for duodenal compression.[20] Additionally, these corrective spinal procedures result in significant acute lengthening of the vertebral column which results in vertical tension on the SMA and a cephalad displacement of the aorto-SMA junction at the expense of lateral mobility, causing extrinsic compression of the distal duodenum as it passes through the sharp aorto-mesenteric angle.[15,20] Moreover, majority of patients with AIS have associated thoracic hypokyphosis, which causes a more extended spine and a reduced aorto-mesenteric angle.[18,21]

Our patient was first referred to the Orthopaedic Department from School Health Services in March 2015, having reported lower back pain after sitting for long periods of time. She had a family history of scoliosis in her father who was conservatively managed. X-rays showed moderate to severe scoliosis with Cobb angles of left thoracic T1–8 at 32º, right thoracolumbar T8–L3 at 52º, and a Risser grade of 1–2. She was a candidate for surgical intervention in view of her age and risk of spinal curve progression based on aforementioned radiological features.[22] Other than weight loss and a pre-op BMI of less than 25th centile which put SN at increased risk of SMA syndrome post-operatively, other known risk factors for SMA syndrome after scoliosis correction include a staged procedure, lumbar modifiers of B or C as opposed to A, increased stiffness of the thoracic curve, sagittal kyphosis, height percentile less than 50 percent, and heavy and quick halo-femoral traction after spinal anterior release.[13,17]

Clinical Features of SMA Syndrome

SMA syndrome usually presents within days to weeks following scoliosis surgery although late presentations have also been reported.[13,18,20,21,23] Typical features are vomiting (92.9%), abdominal pain (57.1%), distension (42.9%), bilious vomiting (35.7%), and hypoactive bowel sounds (28.6%).[18-20] Those with mild obstruction may manifest only with postprandial epigastric pain and early satiety, while those with more advanced obstruction may have severe nausea, bilious vomiting, and weight loss. Reflux may also be present.

Notably, as seen in our patient, symptoms of SMA syndrome may be relieved when the patient is lying prone, in the left lateral decubitus, or in a knee-chest position. These positions remove tension from the mesentery and SMA by increasing the aorto-mesenteric distance.[24] Although a specific relieving position was not reported by SN, postural improvement was clearly demonstrated during the barium study when she was placed in a lateral decubitus position which facilitated contrast flow and alleviation of symptoms.

Although symptoms of abdominal pain with distension and vomiting are not specific for SMA syndrome, the presence of other unusual features such as relief with positional changes in the lateral decubitus or knee-chest position as in SN's case should be regarded extra cautiously as these are not normal findings in other less serious causes of abdominal pain, nausea, or vomiting, such as viral gastroenteritis, even if the diagnosis of SMA is not considered.

Other causes of SN's recurrent nausea, vomiting, and abdominal pain associated with meals were also considered. In particular, her pain medications on discharge from the recent operation were reviewed as side effects from opioids which are commonly prescribed for post-operative pain could have contributed to her symptoms. A random point-of-care-test blood sugar level was also checked to ensure this was not a first presentation of diabetic ketoacidosis in view of her symptoms and dehydrated state on arrival. Causes for an acute surgical abdomen such as intussusception and appendicitis were also considered although the pattern of pain, lack of fever and physical signs, as well as a low Alvarado score made the latter less likely. Constipation colic and gastritis were also considered initially, but the acute history in the absence of any previous abnormalities in bowel or dietary habits and dyspepsia made them less likely. Moreover, SN did not respond to the initial trial of laxatives and symptomatic medications for gastritis which made a search for another underlying cause imperative. Psychogenic causes such as adolescent rumination syndrome and bulimia nervosa were distant possibilities that were also entertained although the lack of psychological overlay and the presence of significant nausea and forceful vomiting pointed much more strongly to an organic cause.

The diagnosis of SMA syndrome is usually delayed due to the lack of symptom specificity and the long list of differential diagnoses. Potentially life-threatening complications such as electrolyte abnormalities, gastric perforation, gastric pneumatosis and portal venous gas, and the formation of an obstructing duodenal bezoar can occur, with most of the reported deaths involving patients in whom the diagnosis was markedly delayed or was completely missed.[25,26] Fortunately, SN was spared life-threatening complications due to prompt recognition, expedited investigations followed by an early surgical referral. This serves as a stark reminder for primary care physicians to have a high index of suspicion for SMA syndrome especially when its presentation could so easily have led one to label it conveniently as "early gastroenteritis" or "viral illness with vomiting" and to dismiss it simply with symptomatic medications such as anti-emetics, anti-spasmodics, or acid suppression therapy.

Treatment of SMA Syndrome

The treatment of SMA syndrome is aimed at rehydration, correction of electrolyte imbalances, gastric decompression, and nutritional support. Early surgical referral should be

considered especially for high-risk patients (BMI less than 5th centile, sagittal kyphosis) in order to reduce the risk of complications and need for surgery.[20] Psychiatric evaluation may be needed to help manage an eating disorder. Enteral nutrition is preferred and a naso-jejunal tube placed distal to the obstruction may be necessary initially until patients are able to increase their oral intake.[27,28] Changes in feeding position, as aforementioned, may also be useful in relieving the obstruction. High-calorie feeds are often prescribed in order to achieve weight gain, with the goal of increasing the mesenteric fat pad and thus enlarging the aorto-mesenteric angle to relieve duodenal compression.

Conservative medical treatment is usually attempted for at least 6 weeks, failing which surgical interventions such as open or laparoscopic duodenojejunostomy, division of the ligament of Treitz (Strong's procedure), and open derotation of the duodenum may be needed.[24,29]

Fortunately, although a potentially life-threatening condition, majority of cases usually respond to appropriate conservative treatment.[16] This was thankfully seen in our patient, in whom conservative treatment alone with physical manoeuvres for symptom relief and gradual introduction of diet over 4 days was successful.

CONCLUSION

SMA syndrome is a rare but well-known complication of scoliosis surgery and can occur in conditions that cause significant weight loss. Key lessons for family physicians include the need for a high index of suspicion to facilitate early identification of this potentially life-threatening condition especially in patients with risk factors and to provide timely surgical referral. Not every case of vomiting and abdominal pain in children is simply due to a viral illness, gastritis or gastroenteritis. It is vital for clinicians to reconsider the differential diagnoses carefully where there is any doubt.

ACKNOWLEDGEMENTS

I would like to thank Dr Mas Suhaila, Consultant, Division of Paediatric Infectious Disease, Khoo Teck Puat-National University Children's Medical Institute, National University Hospital, as well as Dr Tham Tat Yean, Associate Programme Director, NUHS Family Medicine Residency Programme for their invaluable support in the writing of this paper. Patient consent was also obtained and documented in the electronic medical record (CPSS CDOCs) prior to the writing of this paper.

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