

Case Report

Superior Mesenteric Artery Syndrome: An Uncommon Cause of Intestinal Obstruction ; Report of Two Cases and Review of Literature

Naheda H Jawad¹, Abdulla Al-Sanae¹, Wafa'a Al-Qabandi²

¹Department of Pediatrics, Al-Amiri Hospital, Kuwait

²Department of Pediatrics, Faculty of Medicine, Kuwait

Kuwait Medical Journal 2006, 38 (3): 241-244

ABSTRACT

Superior mesenteric artery (SMA) syndrome is caused by the compression of the third part of the duodenum leading to upper intestinal obstruction which is aggravated when the patient is lying in the supine position. Predisposing factors include rapid weight loss, application of a body cast after spinal surgery, prolonged recumbency, and abnormal position of the ligament of

Treitz. Diagnosis may be difficult but can be confirmed by upper gastrointestinal contrast studies. Treatment is mainly conservative and if failed, surgical intervention is warranted. We report two children who were diagnosed with SMA syndrome and discuss the clinical picture, ways of diagnosis and methods of treatment.

KEYWORDS: aortomesenteric angle, duodenojejunostomy, intestinal obstruction, nasojejun tube, superior mesenteric artery

INTRODUCTION

Superior mesenteric artery (SMA) syndrome is an uncommon but well recognized clinical entity. It is caused by the compression of the third part of the duodenum between the superior mesenteric artery and the aorta causing upper gastrointestinal tract obstruction.

In the following, we report two children who were diagnosed with SMA syndrome and discuss the predisposing factors, diagnosis and modalities of treatment.

Case No. 1:

An 11-year-old girl was diagnosed 18 months earlier with multiple sclerosis of the relapsing-progressive type. She had suffered from frequent relapses, nearly once per month, which resulted in significant physical disability. She presented with history of abdominal distension, nausea and voluminous greenish vomiting. On examination: she was completely bed-ridden and unable to sit or stand without support. She also had right facial palsy and bulbar palsy, which manifested by drooling, inability to swallow and slurred nasal speech. She was noticed to have a weight loss of seven kilograms within a four-week period. In addition, she had squint, intention tremors indicating cerebellar involvement, and was incontinent to both urine and stool. Examination of

the abdomen revealed distension in the epigastric area with succussion splash. A nasogastric tube was inserted and 1700 ml of greenish gastric aspirate was removed. A plain abdominal X-ray showed aeric distension of stomach and proximal duodenum with paucity of distal bowel gas (Fig. 1). An upper gastrointestinal contrast series showed a hugely dilated stomach, dilatation of the first and second parts of the duodenum, with an abrupt vertical cutoff of the barium flow at the level of the third part of the duodenum with normal mucosal folds (Fig. 2). These findings were highly suggestive of SMA syndrome. The diagnosis was confirmed by performing a spiral-computed tomography with angiography (CTA), which showed a sharp narrow angle of 15° between the aorta and the SMA (Fig. 3). A nasojejun tube to feed the patient was inserted beyond the site of the obstruction under fluoroscopic control; unfortunately the patient pulled it out twice. Therefore, a Hickman central line was placed through which she received total parenteral nutrition (TPN). After six months from starting her on TPN, her weight increased by 10 kg, from 23 to 33 kg and she was able to tolerate small amounts of nasogastric tube feeds which were gradually increased and included high caloric nutritional supplement Pediasure (Abbott Laboratories). Since she was still unable to chew and swallow properly, a

Address correspondence to:

Dr. Naheda H. Jawad, PO Box 28358, Safat13144, Kuwait. Tel: (965) 2464728, Pager: 9174332, Res: 2517480, E-mail: naheda@consultant.com



Fig. 1: A plain abdominal X-ray showing distension of stomach and proximal duodenum with paucity of distal bowel gas.

gastrostomy tube was fixed to prevent recurrence. She was discharged home after staying in hospital for seven months. The gastrostomy tube was removed one year later when her facial and bulbar palsy improved and she continued to maintain her weight within the normal range, with no recurrence of symptoms.

Case No. 2:

A 12-year-old girl was admitted complaining of epigastric pain of one month duration. The pain was marked after food intake, lasting for 4-6 hours, relieved by vomiting and usually it contained undigested food. This was associated with decreased appetite and a loss of weight of about four kilograms since the onset of her symptoms. On examination: the girl was tall and thin (her height was on the 90th percentile while her weight was just below the 25th percentile) with a body mass index (BMI) of 13.6. Examination of the abdomen showed no significant abnormality. Barium meal and follow through showed a markedly elongated stomach, moderate dilatation of the duodenal loop with an abrupt cutoff of the flow of barium at the site of the third part of the duodenum. This obstruction improved in the prone position. There was significant delayed-emptying of the stomach; after



Fig. 2: An upper gastrointestinal contrast study showing a hugely dilated stomach, dilatation of the first and second parts of duodenum with an abrupt vertical cutoff of barium flow at the level of third part of duodenum.

five hours at least 50% of the barium was still in the stomach. After 20 hours, barium was located in the cecum, sigmoid colon, and rectum. An endoscopy showed retained fluid in the stomach, which was dilated with an extrinsic pressure at the third part of the duodenum precluding the passage of the scope. A biopsy was taken from that part of the duodenum and it showed normal histology. She was diagnosed as having SMA syndrome and was treated conservatively with cisapride (0.2 mg/kg), which was prescribed as a prokinetic agent. She was advised to lie on her left side after meals to enhance emptying of the stomach. She improved slowly with relief of symptoms allowing her to be discharged from the hospital. She remained symptom free when she was seen during follow-up two years from the onset of her symptoms.

DISCUSSION

Superior mesenteric artery syndrome was first described in 1861 by Von Rokitansky^[1] and since then more than 400 cases have been reported worldwide making it an uncommon but well recognized clinical entity. Although there is no precise incidence, there are reports that from 0.013-



Fig. 3: A spiral-computed tomography with angiography showing a sharp narrow angle (arrow) between the aorta and the superior mesenteric artery.

0.3% of barium studies of the upper gastrointestinal tract have shown changes to support the diagnosis^[2]. It usually affects young females from 10-39 years of age^[3] and in one study in children the mean age was 13 years (10-16 years)^[4]. The pathophysiology of this disorder relates to the angle between the SMA and the abdominal aorta at the level of the first lumbar vertebra. It is caused by trapping of the third part of the duodenum as it crosses between the SMA anteriorly and the aorta and vertebral column posteriorly. The normal angle between the SMA and the abdominal aorta is approximately 45° (range 38-56°) in standing position. Any factor that sharply narrows this angle to between 6-16° results in a vascular, extrinsic compression of the duodenum as it passes between the SMA and the aorta^[5].

Although the exact aetiology is not known, certain predisposing conditions are well recognized:

(1) Diseases associated with rapid severe weight loss with decrease in the retroperitoneal and mesenteric fat such as anorexia nervosa^[6], malabsorption syndrome, increased catabolic states as in cancer and burns.

(2) Neurological disorders causing dystonic denervation of the abdominal wall and spinal

muscles such as acute trauma to the spine^[7], patients who had spinal surgery, the use of a body cast in the treatment of scoliosis or vertebral fractures, traumatic brain injury, spastic quadriplegia, and prolonged recumbency^[8,9].

(3) Anatomical anomalies such as abnormally high and fixed position of the ligament of Treitz, or an unusually low origin of the superior mesenteric artery^[10].

(4) Constitutional factors such as tall thin body build which is reported in 80% of patients^[11], exaggerated lumbar lordosis, visceroptosis, and rapid linear growth without compensatory weight gain, particularly during adolescence.

Clinical symptoms can be acute, or chronic with intermittent exacerbations. The patient usually presents with symptoms of gastrointestinal obstruction, which includes postprandial nausea, epigastric pain, bilious vomiting or vomiting of partially digested food. The symptoms are typically relieved when the patient is lying prone, in the left lateral decubitus, or in the knee-chest position; and they are aggravated when the patient is lying in the supine position.

Diagnosis of SMA syndrome is difficult and requires confirmation by radiographic studies. Upper gastrointestinal contrast series will show the characteristic criteria described by Hines *et al*^[12]:

1) Dilatation of the first and second parts of the duodenum with or without gastric dilatation

2) Abrupt, partial or complete, vertical obstruction of barium flow at the site where SMA crosses the third part of the duodenum (duodenal clamp)

3) Antiperistaltic waves proximal to the obstruction producing a "to-and-fro" movement.

4) Relief of the obstruction when the patient is placed in the left lateral, prone, or knee-chest position.

However, these findings may not be apparent in the chronic intermittent form of SMA syndrome; therefore, their absence does not exclude the disease and if they are negative, the procedure should be repeated during an acute attack.

Recently, the use of contrast-enhanced spiral-computed tomography can confirm the diagnosis by evaluating the aortomesenteric angle. It can also assess the amount of retroperitoneal and mesenteric fat^[13].

Many therapeutic options have been proposed to manage SMA syndrome and these vary from conservative non-operative to operative procedures. Conservative treatment is recommended in all patients with SMA syndrome and it aims at identifying and correcting reversible predisposing factors and restoring retroperitoneal fat to relieve the obstruction. It involves decompression of the stomach and correction of dehydration and

electrolyte imbalance by intravenous fluids. Then feeding starts with small meals of liquids and proper positioning of the patient after meals either prone or in the left lateral decubitus position. Furthermore, metoclopramide administration may be used to enhance emptying of the stomach. Patients with severe rapid weight loss may benefit from TPN or enteral feeding by the insertion of a nasojejun tube, which is passed distal to the site of obstruction^[14]. Medical treatment has a high success rate in cases with an acute presentation of SMA syndrome but most chronic cases require surgery.

Several surgical solutions have been suggested including gastrojejunostomy, dissection and cutting of the ligament of Treitz with mobilization of the duodenum^[15], and duodenojejunostomy. The latter is agreed upon by most surgeons as the mainstay of treatment to prevent recurrence with a success rate of 90%^[16]. Recently, successful laparoscopic duodenojejunostomy was reported as a feasible alternative to open surgery with the advantages of a much lower operative time and rapid postoperative recovery^[17].

Indications for surgery:

- (1) Failure of conservative medical therapy
- (2) A long history of indigestion, progressive weight loss, and pronounced dilatation of the duodenum with stasis
- (3) Complicated peptic ulcer disease^[18]

In our patients, each had a different predisposing factor; the first patient had a neurological disorder resulting in facial and bulbar palsy with inability to chew and swallow food which led to rapid weight loss. In addition, she had significant physical handicap and prolonged recumbency. The second patient had a disproportionate increase in height over the weight with the onset of the growth spurt at puberty. Each of these two patients required a different approach to treatment, which was tailored according to their pre-existing medical conditions, predisposing factors and the progress observed after initiating treatment.

CONCLUSION

In cases of high intestinal obstruction with the presence of a dilated stomach and absent gas in the small intestine, a high index of suspicion for SMA syndrome should be entertained. An abrupt cutoff at the third part of the duodenum after an oral

contrast and a decreased angle (6-16%) between SMA and the aorta on CT-angiogram will confirm the diagnosis.

REFERENCES

- 1) Rokitansky CV. Lehrbuch der Patologische Anatomie, 3rd ed, vol 3. Vienna: Braumuller,1861:187.
- 2) Baltazar U, Dunn J, Floresguerra C, Schmidt L, Browder W. Superior Mesenteric Artery Syndrome: an uncommon cause of intestinal obstruction. *South Med J* 2000; 93:606-608.
- 3) Kaushik R, Attri AK. Acute superior mesenteric artery syndrome. *Indian Pediatr* 2003; 40:1014-1015.
- 4) Philip PA. Superior mesenteric artery syndrome: an unusual cause of intestinal obstruction in brain-injured children. *Brain Inj* 1992; 6:351-358.
- 5) Shetty AK, Schmidt-Sommerfeld E, Hayman ML, Udall JN. Radiological case of the month. *Arch Pediatr Adolesc Med* 1999; 153:303-304.
- 6) Adson DE, Mitchell JE, Trenkner SW. The superior mesenteric artery syndrome and acute gastric dilatation in eating disorders: a report of two cases and a review of the literature. *Int J Eat Disord* 1997; 21:103-114.
- 7) Laffont I, Bensmail D, Rech C, Prigent G, Loubert G, Dizien O. Late superior mesenteric syndrome in paraplegia: case report and review. *Spinal Cord* 2002; 40:88-91.
- 8) Delgado X, Belpaire-Dethiou MC, Chantrain C, et al. Arteriomesenteric syndrome as a cause of duodenal obstruction in children with cerebral palsy. *J Pediatr Surg* 1997; 32:1721-1723.
- 9) Kishi T, Kawahara H, Tanaka T, et al. Superior mesenteric artery syndrome complicating mitochondrial encephalopathy. *J Pediatr Gastroenterol Nutr* 1998; 26:464-467.
- 10) Wilson-Storey D, MacKinlay GA. The Superior mesenteric artery syndrome. *J R Coll Surg Edinb* 1986; 31:175-178.
- 11) Barnes JB, Lee M. Superior mesenteric artery syndrome in an intravenous abuser after rapid weight loss. *South Med J* 1996; 89:331-334.
- 12) Hines JR, Gore RM, Ballantyne GH. Superior mesenteric artery syndrome: diagnostic criteria and therapeutic approaches. *Am J Surg* 1984; 148:630-632.
- 13) Lippel F, Hanning C, Weiss W, Allescher HD, Classen M, Kurjak M. Superior mesenteric artery syndrome: diagnosis and treatment from the gastroenterologist's view. *J Gastroenterol* 2002; 37:640-643.
- 14) Roth EJ, Fenton LL, Gaebor-Spira DJ, Frost FS, Yarkony GM. Superior mesenteric artery syndrome in acute traumatic quadriplegia: case reports and literature review. *Arch Phys Med Rehabil* 1991; 72:417-420.
- 15) Murthi GV, Raine PA. Superior mesenteric artery syndrome in children. *Scott Med J* 2001; 46:153-154.
- 16) Ylinen P, Kinnunen J, Hockerstedt K. Superior mesenteric artery syndrome: A follow-up study of 16 operated patients. *J Clin gastroenterol* 1989; 11:386-391.
- 17) Kim IY, Cho NC, Kim DS, Rhoe BS. Laparoscopic duodenojejunostomy for management of superior mesenteric artery syndrome: two cases report and a review of the literature. *Yonsei Med J* 2003; 44:526-529.
- 18) Fromm S, Cash J. Superior mesenteric artery syndrome: an approach to the diagnosis and management of upper gastrointestinal obstruction of unclear etiology. *S D J Med* 1990; 43:5-10.