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SUPERIOR MESENTERIC ARTERY SYNDROME*

A Case Report

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A case of superior mesenteric artery syndrome is reported. A 16-year-old boy with bilious vomiting for six months and weight loss within the previous year is presented. In this symptomatic period, he was fed many dietary formulations. This conservative management failed. Physical examination revealed that he was malnourished. The diagnosis was made by means of an upper gastrointestinal barium study performed through a tube. Gastrojejunostomy was preferred over duodenal mobilization because he had a markedly dilated stomach and enlarged pylorus. The postoperative course was satisfactory. *Key words: superior mesenteric artery syndrome, vascular compression, duodenal obstruction.*

Superior mesenteric artery syndrome (SMAS) results from extrinsic compression of the duodenum by the superior mesenteric artery (SMA) due to a narrow aortomesenteric angle and distance[1]. The site of obstruction is usually the third part of the duodenum. It may present either acutely or chronically with mild abdominal pain, anorexia, nausea and voluminous but infrequent bilious vomiting[2]. Since SMAS is an uncommon condition, the diagnosis can only be made if a high index of suspicion exists.

We recently encountered a 16-year-old boy referred because of bilious vomiting, abdominal fullness, weight loss and early satiety for the previous six months. The diagnosis of SMAS was made on the basis of an upper gastrointestinal contrast study (by inserting a large cannula into the duodenum) that showed almost total obstruction in the third part of the duodenum.

Case Report

A 16-year-old boy, weighing 38 kg (< 5th percentile) and measuring 1.67 m in height (10th - 25th percentile). presented with bilious vomiting and a six-month history of abdominal fullness. The patient had lost a remarkable amount of weight

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within a year prior to the start of his complaints. He was found be malnourished (body weight 60 percent of standard according to weight-for-height criteria) on physical examination. There was no abdominal distension or mass.

His hematological indices and results of biochemical and malabsorption tests were all within normal limits. A plain x-ray of the abdomen showed a dilated stomach and proximal duodenum. An upper gastrointestinal barium study displayed an excessively enlarged stomach and distension of the proximal duodenum with an external obstruction in its third part and antiperistaltic waves from the duodenum towards the stomach (Fig. 1). On the basis of these radiological findings, compression of the duodenum by the SMA was suspected. The stomach was decompressed by a nasogastric tube, and the patient was given total parenteral nutrition. A preoperative gastroduodenoscopy revealed no intrinsic duodenal obstruction but a continuously enlarged, open pylorus. There was no histopathological abnormality on the duodenal biopsy specimen except for giardiasis. At laparotomy, a markedly enlarged stomach and an extrinsically compressed third part of the duodenum by the SMA was found. Gastrojejunostomy was performed and a gastrostomy tube was placed. Following surgery, all symptoms disappeared and the patient began to gain weight. His gastrostomy tube was later removed, and he was discharged home when symptom-free.

Fig. 1: Upper gastrointestinal contrast radiography of patient. Note almost total obstruction in the third part of the duodenum.

Discussion

In the absence of an intestinal mass or anomalies such as malrotation, webs and atresias, vascular compression should be considered as a cause of obstruction. There have been previous case reports of duodenal obstruction due to SMA compression in the literature[2-7].

SMAS is also called Wilkie's syndrome, cast syndrome, chronic duonenal ileus, and arterio-mesenteric duodenal compression syndrome. It was first described by Von Rokitansky in 1861[8] and then reported as a distinct clinical entity by Wilkie in 1921[9]. It may be either congenital or acquired. The acquired form of the syndrome may result from a) accelerated linear growth with no associated weight gain, b) rapid weight loss due to chronic illness or bed rest, or c) hyperextension of the vertebral column in a cast or spica[10].

The third part of the duodenum crosses the spinal column and abdominal aorta at the level of the second lumbar vertebra. This position is caudal to the origin of the SMA, which normally forms an angle of approximately 45° with the aorta. The SMA is normally embedded in mesenteric fat and lymphatic tissue and does not compress the duodenum. If a patient loses weight rapidly, grows rapidly in height without associated weight gain or his vertebral column hyperextends in a cast, the tension in the root of the mesentery increases and/or the fatty cushion around the SMA diminishes. All these conditions result in a decreased aorta-superior mesenteric artery angle to approximately 15° and thus compression of the third part of the duodenum, leading to SMAS[2]. Among the other predisposing factors are peritoneal adhesions, incomplete duodenal rotation and abnormally high position of the ligament of Trietz with an upward displacement of the duodenum, rendering the aortomesenteric angle narrow. Our patient's body weight was not appropriate for his height; thus rapid weight loss within one year prior to his admission to the hospital may be the predisposing factor leading to SMAS.

Patients with SMAS may present either acutely or with a chronic history. The main symptoms are bilious vomiting, epigastric fullness and pain. The chronic cases presenting with intermittent vomiting, early satiety and anorexia may remain undiagnosed for many years or be misdagnosed as having anorexia nervosa. The presented case had a chronic syndrome characterized by bilious vomiting, abdominal fullness, weight loss and early satiety.

The diagnosis of SMAS can be confirmed radiologically by duodenography and/or angiography[4]. To demonstrate compression of the duodenum in our case, we used an upper gastrointestinal barium study performed through a tube. In one study of approximately 6,000 upper gastrointestinal contrast studies over a 10-year period, Anderson et al.[3] reported 12 cases (0.2%) with SMAS, indicating that the syndrome is uncommon.

Conservative therapeutic measures include alteration in diet and feeding habits, such as giving frequent feedings of pureed or blenderized foods and nursing in the prone position or while the patient is lying on his left side. Alimentation of the patient may result in weight gain, deposition of fat within the mesentery and around the SMA and, finally, relief of the duodenal compression. In the acute presentation of the syndrome or if conservative management fails, surgical intervention is necessary. During surgery may be necessary to insufflate the duodenum with air to confirm the diagnosis using the maneuver advocated by Wilkie and popularized by Jones[11], as was done in this case. In children, the most effective method is the one described by Louw et al.,[12] Burrington[13], and Wayne et al.[14] This method consists of mobilizing the entire duodenum and lysing the Treitz ligament through a small, transverse right-upper-quadrant incision. Since there was a markedly dilated stomach with an enlarged pylorus in our patient, gastrojejunostomy along with placement of a gastrostomy tube was performed, resulting in complete relief of symptoms and weight gain.

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