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A Rare Case of Late Presentation of Superior Mesenteric Artery Syndrome Following Kyphoscoliosis Surgery

Case Report

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Abstract

Introduction: Obstruction of the third part of the duodenum by the Superior Mesenteric Artery (SMA) can occur following surgical correction of scoliosis. The condition most commonly occurs in significantly underweight patients with severe deformities during the first few days to a week following spinal surgery.

Case presentation: We present the atypical case of an adolescent idiopathic thoracolumbar scoliosis that underwent corrective surgery with instrumentation 3 months back. And developed SMA syndrome several months postoperatively. The condition manifested with recurrent vomiting, abdominal distension, marked dehydration, and severe electrolyte disorder. Prolonged nasogastric decompression, nasojejunal feeding, IV fluids and antibiotics with proper positioning resulted in resolution of the symptoms with no recurrence at follow-up.

Conclusion: SMA syndrome can occur much later than previously reported and with potentially life-threatening symptoms following scoliosis correction. Early recognition of the condition and institution of appropriate conservative measures is critical to prevent the development of severe complications including the risk of death

Keywords: SMA syndrome; Kyphoscoliosis; CT angiography

Introduction

Vascular compression of the third part of the duodenum by the Superior Mesenteric Artery (SMA) results in the development of a rare condition of gastric outlet occlusion known as SMA syndrome [1]. The etiology of the syndrome is connected to the anatomy of the third part of the duodenum in relation to the aortomesenteric angle. Obstruction of the small bowel by the SMA has been previously associated with spinal manipulation in the surgical or conservative management of scoliosis.

In scoliosis, the syndrome occurs most commonly in thin and asthenic patients with a low Body Mass Index (BMI) who undergo spinal manipulation and correction of the curvature by instrumentation, skeletal traction, casting or bracing; these corrective techniques all result in significant lengthening of the vertebral column and an extrinsic compression of the distal duodenum as it passes through the sharp angle formed by the aorta and the spine posteriorly and the SMA anteriorly. Following scoliosis surgery, the condition usually develops during the first postoperative week [2].

We present a patient with an adolescent idiopathic kyphoscoliosis who underwent spinal correction surgery and developed severe SMA syndrome 3 months following surgery. Such late presentation has not been reported in literature till date as per our knowledge.

Case Report

A 15 years old female patient came to the Emergency with

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complain of pain, vomiting and abdominal distension for 15 days and constipation for 2 days. She is a known case of idiopathic thoraco-lumbar kyphoscoliosis. She underwent corrective surgery 3 months back.

X-ray abdomen AP view showed 1-2 air-fluid levels. There is a large air-fluid level in the fundic area. Central abdomen was gas less, however peripheral abdomen showed some gas (Figure 1).

No specific diagnosis could be reached via ultrasonography, as most of the abdomen was obscured by the fluid and solid mixed content within the bowel.

The patient was taken for emergency contrast enhanced CT abdomen & pelvis.

There was gross dilatation of first and second part of duodenum and gross dilatation of stomach with air-fluid level within the stomach (Figure 2 and 3). Abnormal spinal curvature caused decreased aorto-mesenteric distance, measuring 2.5 mm & decreased aorto-mesenteric angle (aorto-SMA angle) which measured 14º (Figure 4 and 5).

It was diagnosed as a case of duodenal obstruction at D3 segment due to decreased aorto-mesenteric distance secondary to abnormal spinal curvature- Secondary Superior Mesenteric Artery (SMA).

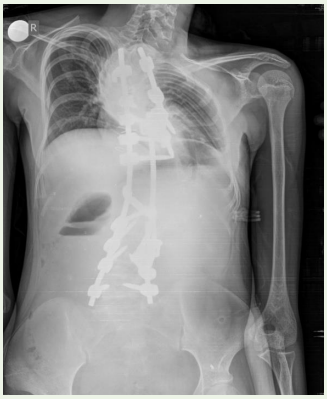

Figure 1: X-ray showing few air-fluid levels with a large air-fluid level in the fundic area.

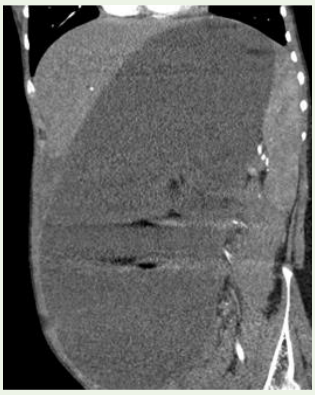

Figure 2: Contrast enhanced CT showing gross dilatation of stomach.

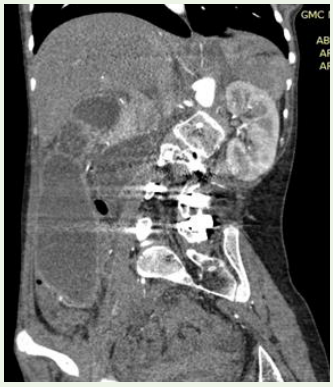

Figure 3: Contrast enhanced CT showing gross dilatation of first and second part of duodenum.

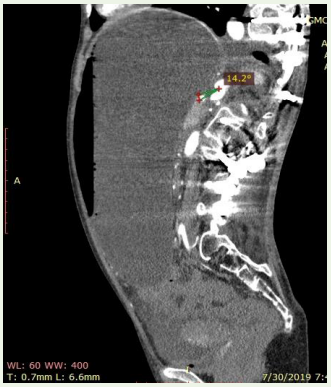

Figure 4: Abnormal spinal curvature is causing decreased aorto-mesenteric distance decreased aorto-mesenteric angle (aorto-SMA angle) which measures 14º.

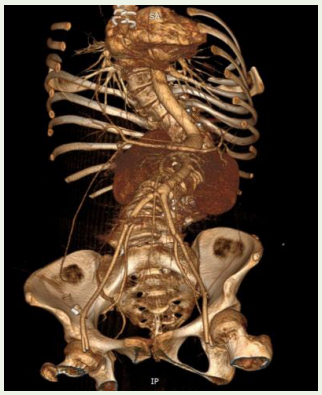

Figure 5: Volumetric CT showing scoliosis with bending of aorta.

Syndrome

Management & followup

The patient was managed conservatively with nasogastric decompression, nasojejunal feeding, IV fluids and antibiotics; and proper positioning was given. She was discharged when the symptoms subsided, was advised for a balanced diet. She is now well with no such complaints at present.

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Discussion

So, here we presented an atypical case of a patient with adolescent idiopathic thoracolumbar scoliosis who underwent corrective surgery with instrumentation 3 months back. She developed SMA syndrome due to progressive weight loss several weeks postoperatively. The condition manifested with recurrent vomiting, abdominal distension, marked dehydration, and severe electrolyte disorder. Prolonged nasogastric decompression, nasojejunal feeding, IV fluids and antibiotics with proper positioning resulted in resolution of the symptoms with no recurrence at follow-up.

Superior Mesenteric Artery (SMA) syndrome [1,2], also known as Wilkie syndrome, is a rare acquired vascular compression disorder in which acute angulation of the Superior Mesenteric Artery (SMA) results in compression of the third part of the duodenum, leading to obstruction. Fat and lymphatic tissues around the SMA provide protection to the duodenum against compression. Under conditions of severe weight loss like anorexia nervosa, malabsorption, hypercatabolic states (burns, major surgery, malignancy), this cushion around the SMA is diminished, causing angulation and reduction in the distance between the aorta and the superior mesenteric artery.

Other conditions may also precipitate this syndrome: increased spinal lordosis, application of a body cast, short ligament of Treitz, multiple attachments of the ligament of Treitz to the duodenum, high fixation of the duodenum by the ligament of Treitz, associated with diabetes mellitus and blunt abdominal trauma, as a Complication of spinal Surgery, etc.

CT and Magnetic Resonance Angiography (CTA/MRA) enable visualization of vascular compression of the duodenum and measurement of aortomesenteric distance [3-6]. Normally, the aortomesenteric angle and aortomesenteric distance are 18-70° and 10-28 mm, respectively. In SMA syndrome, both parameters are reduced, with values of 6° to 15° and 2 to 8 mm respectively.

The incidence of SMA syndrome after surgical procedures to correct spinal deformities has been reported to vary between 0.5 and 4.7% [2,7-11]. This occurs in early post-operative period, within a week or two. Children usually present for surgical correction of an adolescent idiopathic scoliosis during the phase of their most rapid longitudinal growth. The mechanism is that of an acute lengthening of the spinal column, which results in a cephalad displacement of the aorto-SMA junction at the expense of lateral mobility, due to either rapid height gain occurring during adolescence, or following correction of spinal deformities using either conservative (body casts and braces) or surgical methods. This accelerated skeletal growth may alter the relation between the SMA and the spine by decreasing the aortomesenteric angle and, therefore, increase the risk for duodenal compression.

Certain factors has been attributed to delayed onset SMA syndrome in postoperative patients like that mentioned in this case report [2,10,11].

1. Progressive postoperative weight loss

2. The application of the spinal jacket could have caused extrinsic pressure to the abdomen, resulting in further decrease in the aortomesenteric angle and contributing to the onset of the symptoms.

3. In addition, disruption of the autonomic nerve supply to the small intestine, which commonly occurs during the retroperitoneal dissection to approach anteriorly the thoracolumbar spine, can precipitate the development of the condition [7].

Conclusion

We believe that it is essential to identify those patients who are at greater risk of developing duodenal obstruction. Initiate intensive preoperative dietary supplementation in undernourished patients scheduled to undergo spine deformity surgery as a preventative measure. We have described a patient who demonstrates that SMA syndrome can develop late following scoliosis surgery. A high index of suspicion will lead to an early diagnosis of the condition at a stage when conservative measures are more likely to produce a good outcome. If the diagnosis is delayed or missed, SMA syndrome can cause considerable morbidity and may result in potentially life-threatening complications.

Acknowledgments

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