

SECTION EDITOR: BEVERLY P. WOOD, MD

## Radiological Case of the Month

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**A** 14-YEAR-OLD African American adolescent girl presented with a 6-month history of intermittent upper abdominal pain and postprandial vomiting. These symptoms had become progressively worse during the 2 months prior to hospital admission. The medical history was unremarkable except for dysmenorrhea. The physical examination revealed a thin adolescent girl

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with a weight of 35 kg (10th percentile) and height of 150 cm (25th percentile). Her vital signs were normal. The abdomen was slightly distended with mild tenderness in the epigastric region and normal bowel sounds on auscultation. No hepatosplenomegaly or ascites was detected. Results of a complete blood cell count and erythrocyte sedimentation rate were normal. A guaiac test of the stool was negative for occult blood. A radiograph of the abdomen revealed gastric distention. Abdominal and pelvic ultrasound was normal. An upper gastrointestinal tract contrast-medium study was obtained (**Figure**).



Figure.

# Denouement and Discussion

## Superior Mesenteric Artery Syndrome

Upper gastrointestinal tract contrast-medium study showing duodenal dilatation of the first and second parts of the duodenum (arrow) establishes a diagnosis of superior mesenteric artery syndrome.

Superior mesenteric artery (SMA) syndrome is an unusual but well-recognized clinical entity characterized by compression of the third, or transverse, portion of the duodenum against the aorta by the SMA or leaf of the mesentery, and results in chronic, intermittent, or acute duodenal obstruction.<sup>1-4</sup> Superior mesenteric artery syndrome was first described in 1861 by Von Rokitansky,<sup>5</sup> who proposed that its cause was obstruction of the third part of the duodenum, by arteriomesenteric compression. Despite 400 cases reported in the English literature,<sup>1,2,6,7</sup> the existence of SMA syndrome has been doubted; indeed, some investigators have suggested that the SMA syndrome is overdiagnosed because it is confused with other causes of megaduodenum.<sup>2,8</sup>

The SMA normally forms an angle of approximately 45° (range, 38°–56°) with the abdominal aorta, while the third portion of the duodenum crosses caudally to the origin of the SMA, running between the SMA and the aorta.<sup>1,2</sup> Any factor that narrows the aortomesenteric angle (between 6° and 16°) can produce entrapment of the third portion of the duodenum as it passes between the vessels.<sup>9</sup> Important causative factors precipitating narrowing of the aortomesenteric angle are thin body build, exaggerated lumbar lordosis, visceroptosis, abdominal wall laxity, and depletion of the mesenteric fat by rapid severe weight loss, and due to catabolic states (eg, cancer and burns), severe injuries (eg, head trauma leading to prolonged bed rest), and dietary disorders (including anorexia nervosa or malabsorption). Also, spinal disease, deformity, or trauma, and use of a body cast in the treatment of scoliosis or vertebral fractures, rapid linear growth without compensatory weight gain, particularly during adolescence, and anatomical anomalies such as an abnormally high and fixed position of the ligament of Treitz, or an unusually low origin of the SMA are postulated causes of the SMA syndrome.<sup>1,2,9</sup>

This syndrome usually occurs in older children and adolescents and has a female preponderance.<sup>6,10</sup> Symptoms can be acute<sup>11,12</sup> or chronic, with intermittent exacerbations.<sup>3,9,10,13</sup> The patient often presents with chronic upper abdominal symptoms such as epigastric pain, bilious vomiting, postprandial discomfort, or subacute small-bowel obstruction. The symptoms are often relieved when the patient is in the left lateral decubitus, prone, or knee-chest position<sup>1,2</sup> and aggravated in the supine position. Delay in the diagnosis of SMA syndrome can result in malnutrition, dehydration, electrolyte abnormalities, and death.<sup>14</sup>

Confirmation of the diagnosis usually requires radiographic demonstration such as an upper gastrointestinal tract series, hypotonic duodenography, or computed tomography. Upper gastrointestinal tract study reveals dilatation of the first and second part of the duodenum with an abrupt vertical or linear cutoff at the third part. Mucosal folds are normal.<sup>2</sup> Fluoroscopy demonstrates to-and-fro peristalsis of the barium in the dilated portions of the duodenum. Hy-

potonic duodenography may depict the site of obstruction and a dilated proximal duodenum with antiperistaltic waves within the dilated portion.<sup>1</sup> Computed tomography is useful in the diagnosis of SMA syndrome and can provide diagnostic information including aorta-SMA distance, duodenal distention, and amount of intra-abdominal and retroperitoneal fat.<sup>7,15,16</sup>

The differential diagnosis of the SMA includes anorexia nervosa and bulimia. Superior mesenteric artery syndrome should be differentiated from other causes of megaduodenum, such as diabetes mellitus, collagen vascular conditions, or chronic idiopathic intestinal pseudo-obstruction.<sup>17,18</sup>

Reversing or removing the precipitating factor is usually successful in patients with acute SMA syndrome. Conservative treatment is recommended initially for all symptomatic patients.<sup>1,2</sup> Enteral feeding can be an effective adjunct in the treatment of patients with rapid severe weight loss.<sup>2</sup> Surgical intervention is indicated when conservative measures fail, particularly for patients with a history of chronic symptoms and pronounced duodenal dilation.<sup>1</sup> Duodenojejunostomy is the most frequently used procedure and is successful in 90% of cases.<sup>13</sup>

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