Food for Thought: Complications of Acute Gastric Distension in Prader–Willi Syndrome

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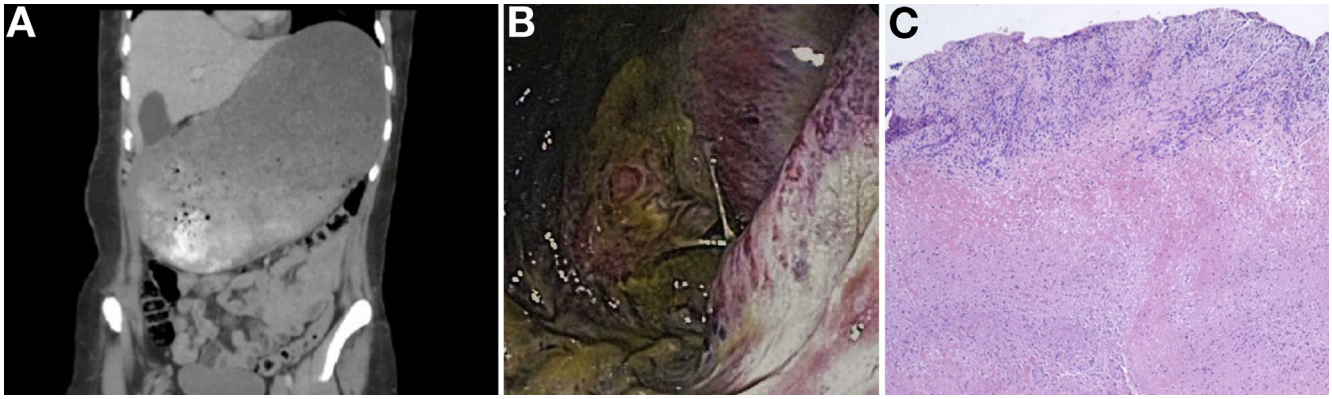

A 24-year-old man with Prader–Willi syndrome (PWS) presented with abdominal pain, nausea, and emesis 3 days after ingesting 8 pounds of raisins and 48 ounces of peanut butter during a food binge. On physical examination, he was afebrile and found to have a diffusely tender abdomen without peritoneal signs. Laboratory test results were significant for a normal white blood cell count, amylase level of 180 U/L, lipase level of 114 IU/L, and a lactate level of 3.2 mEq/L. A computed tomography of the abdomen showed massive distention of the stomach, duodenal bulb, and second portion of the duodenum (Figure *A*). After oral gastric lavage, an upper endoscopy on hospital day 3 showed diffuse erythema, congestion, and dusky-appearing mucosa from the gastric cardia to the second portion of the duodenum (Figure *B*). The patient initially was managed conservatively and appeared to improve clinically with bowel rest and total parenteral nutrition. However, on hospital day 20 he developed hematemesis, fever, tachycardia, and peritoneal signs. A repeat endoscopy showed diffusely necrotic-appearing gastric mucosa with possible perforation of the distal body. The patient was taken to the operating room for an exploratory laparotomy and underwent a total gastrectomy, splenectomy, distal pancreatectomy, and distal esophagectomy with creation of a split fistula and jejunostomy. Pathology specimens later confirmed transmural necrosis of the gastric wall (Figure *C*). After 6 additional surgeries, the patient was discharged on hospital day 55 to a rehabilitation facility. At 2 years after surgery he has not had any major nutritional or clinical consequences as of yet aside from significant weight loss.

Discussion

Total gastric necrosis is a rare diagnosis because the stomach has a rich blood supply that protects it from ischemic damage. However, several case reports have described acute gastric distension and necrosis in PWS patients.[1] PWS is a multisystem genetic disorder characterized by mental retardation, hypogonadism, obesity, and hyperphagia. One study found gastric necrosis to be the cause of mortality in 3% to 6% of patients with PWS.[1] Although the pathophysiology of gastric necrosis in acute gastric distention is poorly understood, increased intragastric pressure has been shown to cause ischemia of the mucosa and submucosa.[2] In our patient, the food binge and resulting fluid influx acutely increased intragastric pressures beyond vascular perfusion pressures, leading to transmural gastric ischemia. The increased intragastric pressures then persisted because of delayed gastric emptying of the massive food

bolus, leading to an inability to reperfuse the ischemic tissue and eventual gastric tissue necrosis with perforation.

The diagnosis of gastric necrosis or perforation is challenging in the setting of PWS because these patients are known to have an increased tolerance to pain and a decreased ability to vomit.[3] Therefore, abdominal pain or vomiting after a recent food binge should heighten the clinician's suspicion for gastric necrosis. Our case highlights the importance of close clinical monitoring after acute gastric distension in PWS because necrosis and gastric perforation are possible devastating complications that can be treated successfully with early identification.

References

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Conflicts of interest
The authors disclose no conflicts.

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