A 34-YEAR-OLD woman was seen in the emergency department with a 3-day history of abdominal pain associated with nausea and vomiting. The patient reported decreased appetite and weight loss over the previous 6 months. She was afebrile and in no apparent distress. On abdominal examination, a large mass was palpated in the epigastrium and the right upper quadrant. A computed tomographic examination of the abdomen was obtained (Figure).

What Is the Diagnosis?

A. Ménétrier disease
B. Gastrointestinal stromal tumor
C. Rapunzel syndrome
D. Lymphoma

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Answer

Rapunzel Syndrome

Computed tomographic scan of the abdomen showing large bezoar in the stomach.

The Rapunzel syndrome is a rare manifestation of a trichobezoar which was first described by Vaughn et al in 1968. It is named after the heroine of a Brothers Grimm fairy tale who was rescued from a tower by a prince after letting down her long hair. Features include a gastric trichobezoar with a tail and gastrointestinal symptoms. To date, 17 cases have been described in the literature, to our knowledge.

Bezoars are concretions of plant or animal material in the gastrointestinal tract. The term bezoar is derived from the Persian “padzahr” meaning counterpoison or antidote. The classic review of 311 patients was published by DeBakey and Ochsner in 1939. Symptoms range from early satiety, nausea, vomiting, and epigastric pain to obstruction and perforation. Physical findings include the presence of a nontender palpable mass. Computed tomographic examination may show an intraluminal mass without attachment to bowel wall and presence of air within the mass. Treatment depends on the composition of the bezoar. Plant bezoars (phytobezoars) may be dissolved with various enzymes. Trichobezoars (hair) are often resistant to enzymatic dissolution and require endoscopy for removal. Surgical removal is indicated when enzymatic dissolution and endoscopy are unsuccessful.

This patient had a large trichobezoar extending from the stomach into the small intestine causing a complete bowel obstruction. She underwent exploratory laparotomy and removal of the trichobezoar through an anterior gastrostomy. A psychiatric evaluation was requested for trichotillomania and trichophagia (pulling out hair and eating it). The remainder of her hospital course was unremarkable.

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REFERENCES


Submissions

The Editor welcomes contributions to the “Image of the Month.” Send manuscripts to Grace S. Rozycki, MD, Department of Surgery, Emory University School of Medicine, 69 Butler St SE, Atlanta, GA 30303; (404) 616-3553; fax (404) 616-7333 (e-mail: grozyck@emory.edu). Articles and photographs accepted will bear the contributor’s name. Manuscript criteria and information are per the “Instructions for Authors” for Archives of Surgery. No abstract is needed, and the manuscript should be no more than 3 typewritten pages. There should be a brief introduction, 1 multiple-choice question with 4 possible answers, and the main text. No more than 2 photographs should be submitted. There is no charge for reproduction and printing of color illustrations.