Rapunzel Syndrome: A Rare Cause of Vomiting and Pain Abdomen

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Abstract

Bezoar is an agglomeration of food or foreign material in intestinal tract. Contiguous extension of trichobezoar into the small bowel can lead to the Rapunzel syndrome. We present rare case of Rapunzel syndrome in 16 year old female who presented with recurrent vomiting.

Key Words: Trichobezoar, Rapunzel Syndrome, Small Bowel

Introduction

Bezoar is an agglomeration of food or foreign material in intestinal tract. The term bezoar derives from the Arabic word Bedzhar, which means antidote (1). Bezoar can be classified according to the primary constituent, as trichobezoar (hair) or phytobezoar (plant material), but may fall into miscellaneous category including fungal agglomerations, food boluses, chemical concretions or foreign bodies (2). Contiguous extension of trichobezoar into the small bowel can lead to the Rapunzel syndrome (3). We present rare case of Rapunzel syndrome in 16 year old female who presented with recurrent vomiting.

Case Report

A 16 year old girl presented with complaints of pain abdomen, vomiting and mass felt per abdomen. For last one week, she had daily episodes of postprandial, nonprojectile vomiting (usually twice per day) consisting of recently taken food materials. On per abdominal examination, a hard to firm mobile mass was felt extending about 5 cm from the xiphoid cartilage. Radiograph of abdomen showed an opaque soft tissue mass in distended stomach.

Computed Tomography (CT) abdomen showed an overdistended stomach with mottled, nonenhancing mass (seen distinct from stomach wall) extending from stomach into duodenal loop with few fragmented segments of mass in small bowel loops. Upper gastrointestinal endoscopy showed a big mass of hair occupying whole of stomach extending into fundus, antrum and going across pylorus. Scope could not be negotiated into duodenum. She was taken up for surgery and exploratory laparotomy with gastrotomy was done with removal of hair ball. She had no complications in her postoperative period and was discharged after few days.

Discussion

Trichobezoar is the most common type of bezoar in patients less than 30 years of age. Baudamant reported the first case of a human trichobezoar in 1779 (4,5). Schonborn performed the first surgical removal of a trichobezoar in 1883 (6). Trichobezoar usually occurs in patients with a history of trichotillomania (7). Hair strands too slippery to be propelled are initially retained in the mucosal folds of the stomach and become enmeshed over a period of time (8). Trichobezoar are usually black from denaturation of proteins by acid, glistening from retained mucus and foul smelling from degradation of food residue trapped within it (9).

When trichobezoar extends from the stomach into the duodenum, the proxima small intestine or to the ascending colon it is called Rapunzel syndrome (10, 11, 12). First case of Rapunzel syndrome was published by Vaughn in
1968 (13). The most common manifestations of Rapunzel syndrome are abdominal pain, nausea, vomiting (as in our case) and intestinal obstruction. The complications (14-17) of Rapunzel syndrome include incomplete pyloric obstruction, obstructive jaundice, megaloblastic anemia, protein losing enteropathy, acute pancreatitis, perforation, peritonitis, short bowel syndrome, internal fistulae and rarely death.

Various imaging modalities have been recommended for detection of bezoars (5). Conventional radiograph shows an opaque soft tissue mass in dilated stomach. Sometimes calcified rim may delineate the edge of the bezoar (5,18). Endoscopy is diagnostic in almost all cases while ultrasound has not much to offer as a diagnostic tool. CT best describes the size and configuration of the bezoar and establishes its location. The characteristic appearance on CT is an inhomogenous, nonenhancing mass with mottled appearance (due to minute air pockets trapped within mass) within the lumen of the stomach/bowel. Oral contrast circumscribes the mass and may fill the interstices near the surface.

Surgical removal of the hair ball particularly when it is big, is the usual method of treatment. Treatment consists of removing the mass by a single enterotomy or resection of the bowel, if not feasible (19,20). It is mandatory to perform a thorough exploration of the stomach and small intestine searching for retained bezoars. Endoscopic removal should be resolved for small trichobezoars only (21). Various other methods (5,22) like laparoscopy, extracorporeal shock wave lithotripsy, intragastric
administration of enzymes (pancreatic lipase, cellulose), use of user ignited mini explosive technique and drugs (metoclopramide, acetylcysteine) have been reported in the literature with varying success rate.

**Conclusion**

All patients with trichobezoar should be referred for psychiatric evaluation after surgery to avoid recurrence. Currently in our country there are no reliable data on the frequency of the disease and its recurrence. It is our firm belief that data collection in our environment would help us better in understanding the behaviour of this disorder and help us in analyzing other factors that trigger their cause and behaviour.

**References**