

Rapunzel Syndrome: A Rare Presentation with Intestinal Intussusception

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Abstract

A bezoar is a collection of foreign material in the gastro-intestinal tract. A trichobezoar is a bezoar formed by the ingestion of hair. When the bezoar extends into the small intestine it is called as Rapunzel syndrome. Rapunzel syndrome is an extremely rare intestinal condition in humans. Trichobezoar with Rapunzel syndrome is an uncommon diagnosis in children is predominantly found in emotionally disturbed or mentally retarded youngsters can even present with complications like intestinal obstruction, perforation and peritonitis. Here we present a rare case of Rapunzel syndrome with intestinal intussusception in a 6 year old girl.

Keywords: Gastro-intestinal tract; Bezoar; Hypochondrium

Introduction

A bezoar is a collection of foreign material in the gastrointestinal tract. A trichobezoar is a bezoar formed by the ingestion of hair. When the bezoar extends into the small intestine it is called as Rapunzel syndrome.

Rapunzel syndrome is an extremely rare intestinal condition in humans. Trichobezoar with Rapunzel syndrome is an uncommon diagnosis in children It is predominantly found in emotionally disturbed or mentally retarded youngsters it can even present with complications like intestinal obstruction, perforation and peritonitis.

Here we present a rare case of Rapunzel syndrome with intestinal intussusception in a 6 year old girl thrombosis.

Case Presentation

A 6 year old girl was referred to surgical emergency for evaluation of vomiting, abdominal pain and absolute constipation for one day.

On examination, the patient was conscious, cooperative, unussualfacies, pale and haemodynamically stable.

Abdominal examination revealed a firm epigastric mass measur-

ing 3 cm*, 2 cm.

Laboratory investigations revealed hemoglobin of 10 g/dL, a mildly elevated total leucocyte count of 12000.

The plain abdominal x ray showed grossly distended stomach [1].

Ultrasound examination of the abdomen was revealed appearance of intestinal intussusceptions at the level of the left hypochondrium with a strand of intussusceptions of 24 mm of thickness. She was taken up for elective laparotomy. On exploration, the stomach was distended and therewere an intussusception in the jejunum. The tail of bezoar was palpable throughout the jejunum [2]. Intussusception was manually reduced and gastrotomy along the body of the stomach parallel to greater curvature for a length of 5 cm was carried out.

The trichobezoar made of bile stained foul smelling hair and cotton thread was removed along with the tail part Gastrotomy was closed in two layers. Post-operative period was un-eventful and she was discharged with psychiatric counseling (**Figure 1**).



Figure 1 Ultrasound examination of the abdomen

Discussion

The term “bezoar” is derived from the Persian word “Padzahr” or from the Arabic word “bedzehr” meaning “protecting against a poison. The first reference to a bezoar in a human was in 1779 during an autopsy of a patient who died from gastric perforation and peritonitis There are various types of bezoar including trichobezoar (hair), phytobezoar (vegetable material),

lactobezoar (milk products), pharmaco bezoar and bezoars of honey comb, and cotton fibers. Debakey and Oschner suggested that the slippery nature of hair and its entrapment within gastric folds could be the reason for bezoar formation [3].

Over a period of time, hair gets matted with each other and with other indigestible materials like cotton fibers and vegetable matter to assume the shape of the stomach. Sometimes its tail may extend into the small intestine. Rapunzel syndrome is an extremely rare intestinal condition in humans resulting from ingesting hair (trichophagia). The syndrome is named after the long haired girl Rapunzel in the fairy tale by the Brothers Grimm. The use of the Rapunzel syndrome first appeared in the literature in 1968.

Characteristics of the syndrome include: The body of a trichobezoar (hairball) located in the stomach and its tail (hence the reference to Rapunzel in the syndrome's name) in the small bowel and/or in the right colon Small or large bowel obstruction. Occurring in psychiatric patients Trichobezoars form when hair strands, escaping peristaltic propulsion because of their slippery surface, are retained in the folds of the gastric mucosa. As more hair accumulates, peristalsis causes it to be enmeshed into a ball. As this ball gets too large to leave the stomach, gastric atony may result. The ball of hair becomes even more matted together and assumes the shape of the stomach, usually as a single solid mass. Bezoars can present with mass in the abdomen, abdominal pain, nausea, vomiting, weakness, weight loss, constipation, diarrhea and malnutrition. In the present case, the patient had mass per abdomen associated with pain and vomiting.

Rarely may it present with complications like intestinal obstruction, bleeding, perforation and obstructive jaundice, pancreatitis and appendicitis. Intestinal obstruction can be the direct effect of the bezoar or secondary to intussusception. The clinician must consider the possibility of Rapunzel syndrome when patient with trichobezoar presents with features of intestinal obstruction. The diagnosis can be made out by ultrasound, computerized tomography, barium meal and endoscopy. Endoscopic retrieval of gastric trichobezoar is less invasive and cost effective than

surgical removal. But it is successful in only few patients and there is instances of respiratory arrest due to airway obstruction while attempting to remove trichobezoar. When Rapunzel syndrome is suspected endoscopic retrieval should not be tried.

Large gastric bezoars can be removed with laparoscopic approach safely but at the rate of prolonged operative time. There are instances where Rapunzel syndrome has been managed laparoscopically In the present case, although the bezoar was extending into the small bowel as a lead point causing intussusceptions. In cases with the bezoar extending into the intestine, usually enterotomy is advised to remove the bezoar. In the present case the complete trichobezoar could be delivered out through a gastrotomy after reducing the intussusceptions. It is a potentially life threatening condition where early diagnosis, surgical intervention has its role. Unless psychiatric counseling and follow up is done, this condition can recur.

Conclusion

The key message is that although surgery is the initial treatment, a comprehensive and long term postoperative psychiatric follow up is needed in patients with Rapunzel Syndrome as a late relapse is possible.

Multidisciplinary health care teams headed by a psychiatrist as well as family support play a key role in the prevention of recurrence.

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