

A rare variant of rapunzel syndrome-acute small bowel obstruction caused by ball of hairs in distal ileum with its tail extending in caecum and ascending colon

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Abstract

Rapunzel syndrome is an extremely rare variant of Trichobezoar. Trichobezoar commonly occurs in patients with psychiatric disturbances as trichophagia (morbid habit of chewing the hair) and Trichotillomania (habit of hair pulling). Bezoars are commonly found in the stomach. In very rare cases of Rapunzel syndrome, hair extends through the pylorus into the small bowel and very uncommonly in large intestine causing symptoms and signs of partial or complete intestinal obstruction. A case report of a rare variant of Rapunzel syndrome, where ball of hairs in small bowel with its tail extending in caecum and ascending colon causing acute small bowel obstruction, is reported in a 13-year-old girl.

Keywords: Rapunzel syndrome, Trichobezoar, Bezoars.

Introduction

A bezoar is accumulation of undigested material (food or foreign body) in the intestinal tract. They are classified according to their constituents: trichobezoar - hair ball, or phytobezoars -undigested food particles such as cellulose. Others includes mycobezoar, pharmacobezoar and lactobezoar.

Trichobezoar is combination of two words "Trish" & "Bezoar". Trish is a Greek work meaning hair and bezoar means poison antidote in Persian/Arabic. Reason for why the hair collects in the stomach is still a debate. Debakey and Oschner suggested that hair entrapment in the gastric folds is the initiating event.³ Due to its indigestibility, resilience and slippery nature, it becomes entrapped within the mucosal folds where it gets enmeshed, and acquires more hair and thus acquires a larger size.

When the trichobezoar breaks and extends beyond the stomach into the small bowel in the form of a tail, it is called as Rapunzel syndrome,³ after Rapunzel, the heroine of a German fairy tale.

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Case Report

A 13 year old girl was referred to surgical emergency for evaluation of vomiting, abdominal pain, abdominal distention and absolute constipation for one day. In addition, the patient also complained of epigastric pain on and off for last 4 months.

On general physical examination, the patient looked anaemic. Her temperature was 37°C, blood pressure 110/60 mmHg and pulse 90/min. On abdominal examination, abdomen was tense and tender with distention, and bowel sounds were exaggerated.

Her routine laboratory examination showed Hb 9g/dl, White blood cell count of 13,000 mcl (reference range

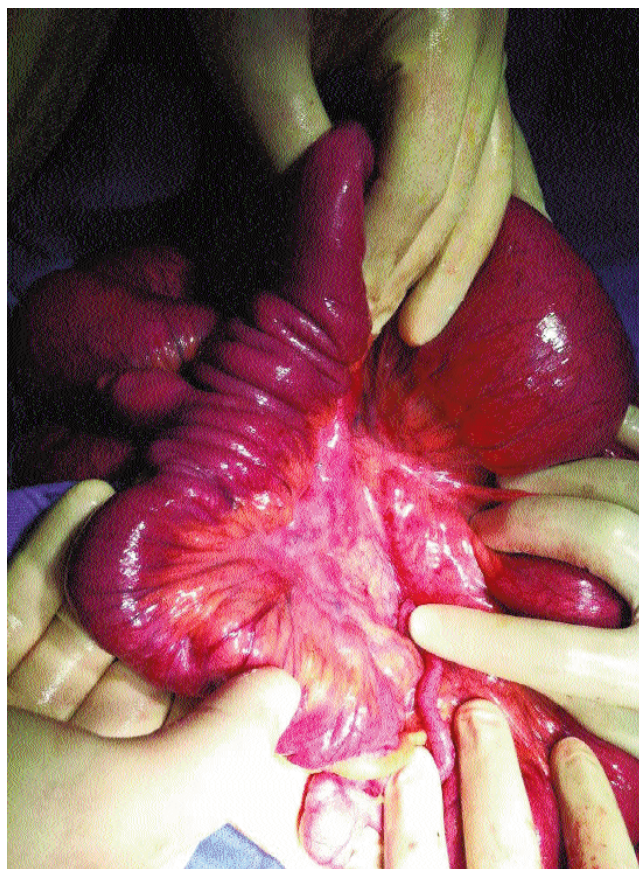


Figure-1: Proximal dilation of ileum by trichobezoar.

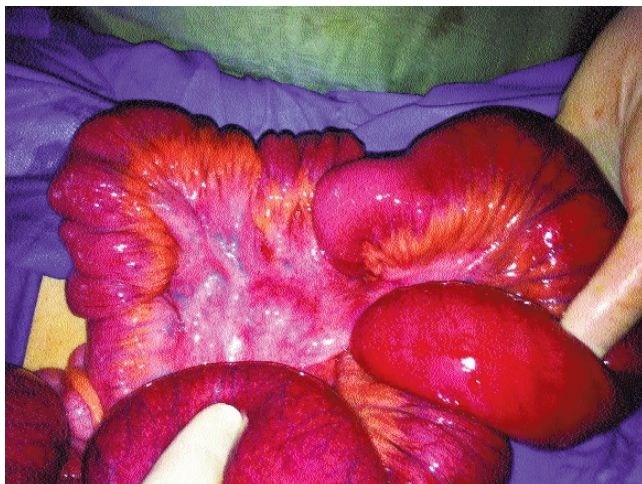


Figure-2: Proximal dilation of ileum by trichobezoar.



Figure-4: Enterostomy with trichobezoar retrieval.

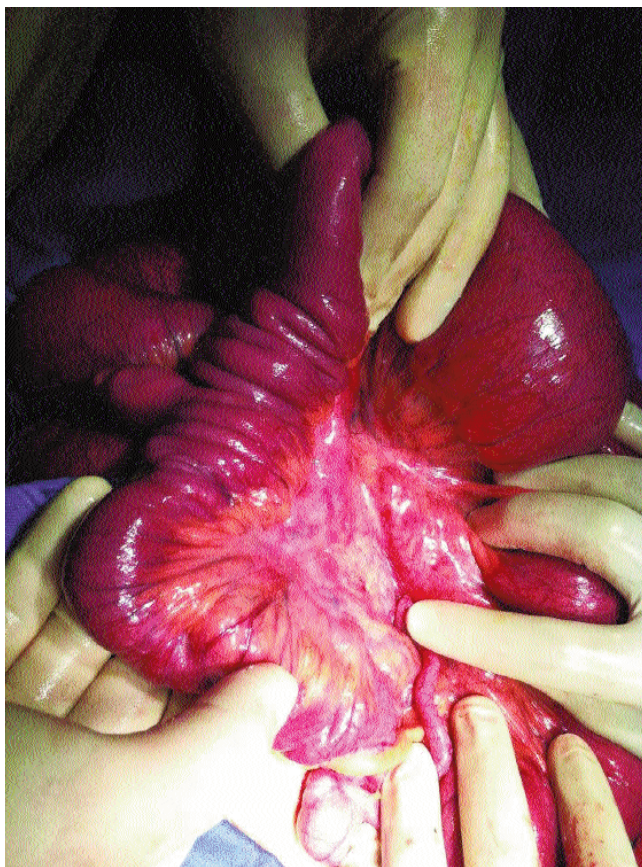


Figure-3: Proximal dilation of ileum by trichobezoar.

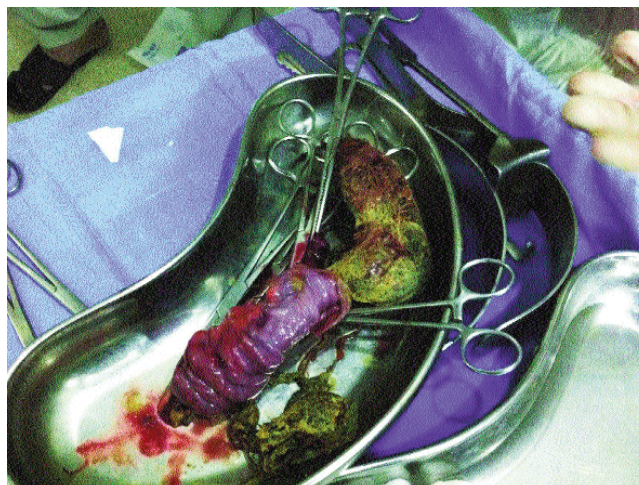


Figure-5: Resected terminal ileum with trichobezoar.

4000-11,000 mL), and normal platelet count $2,45,000 \times 10^9$ (reference range $150,000-4,50,000 \times 10^9$). X-ray plain abdomen was done in erect posture and was reported as normal. Ultrasound abdomen was also done which was

reported as normal. CT scan was not performed, as the facility is not available in emergency department. A preliminary diagnosis of acute small bowel obstruction was made. Appendicular perforation was assumed as most likely diagnosis with atypical presentation and other differentials included obstruction secondary to a congenital band, for instance, congenital vitello intestinal band. After fluid resuscitation, surgery was planned immediately.

On examination of abdomen after anaesthesia, a mass was palpable in right iliac fossa, which was hard to firm in consistency. Patient presentation and physical examination all favoured acute small bowel obstruction, so exploratory laparotomy was planned and carried out from midline incision despite of probable appendicular



Figure-6: Resected terminal ileum with trichobezoar.

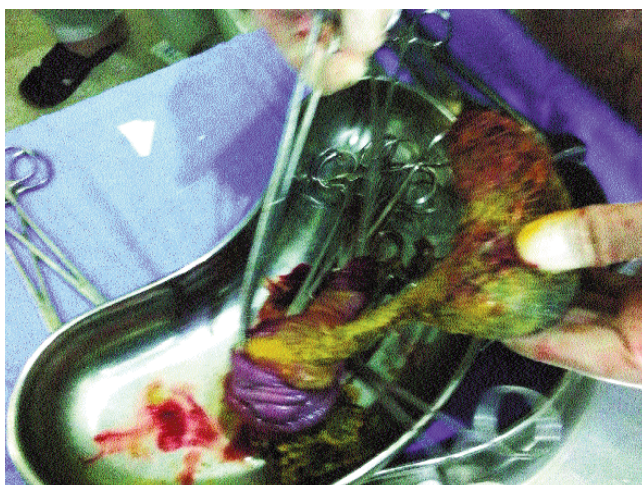


Figure-7: Retrieval of trichobezoar from terminal ileum resected specimen.

mass on palpation under general anaesthesia which could be treated conservatively.

The bowel on examination revealed, the distal ileum having a typical pattern of intussusception extending in the caecum and ascending colon (Figure-1,2,3). A mass was also found 2 feet proximal from ileocaecal junction. On palpation of the small bowel, strands of threads were felt in the ileum. First it was thought to be worms causing the bowel obstruction, and milking maneuver was tried to push the worms into the caecum and large bowel but the maneuver failed. While performing the milking maneuver one of the surgeon came up with a diagnosis of trichobezoar and history of hair pulling and eating was taken from patient's parents, which was positive and then enterostomy was made to remove the trichobezoar. A

huge mass of hair was found and the strands were extending up to ascending colon. The mass completely filled the small bowel lumen, such that it could not be retrieved. Resection and primary anastomosis of the one foot ileum containing the main bulk of hairball was done and strands retrieved from the terminal ileum and ascending colon (Figure-5,6,7). Stomach and whole of small and large gut was examined for other trichobezoar and was found normal. Patient had an uneventful recovery. Psychiatric evaluation was advised, which was refused by parents and the patient was lost to follow up after two outpatient visits.

Discussion

Baudamant first reported Trichobezoar in 1779¹ and Vaughan and colleagues in 1968 reported the Rapunzel syndrome.²

Trichobezoar are the commonest type of bezoars accounting for 55% of all bezoars and majority of the patients are females.³ The main pathology are the long hairs of varying length getting matted together in the form of a hair ball in the stomach. The acidic media of the stomach contents denature proteins in hair and turn them black irrespective of the color of the hair. Undigested dietary fat and bacterial colonization in the hair mesh give it a putrid smell.³ Usually the surface of a bezoar is glistening; due to the mucous covering.

Rapunzel syndrome is an extremely rare entity and a literature review published in 2015, reported fewer than 40 cases.^{3,4} The condition presents with a wide variety of symptoms such as abdominal mass, abdominal pain, nausea and vomiting, weight loss, constipation or diarrhoea and haematemesis.³ Personal history, psychiatric disorders in family, previous bezoar as well physical examination of palpable mass, halitosis and patchy hair loss aids in diagnosis. Presentation of trichobezoar is late, due to low index of suspicion by the clinician.

The complications of the Rapunzel syndrome ranges from attacks of incomplete pyloric obstruction to complete obstruction of the bowel, perforation of bowel leading to peritonitis and mortality.^{3,7} Trichobezoars with small bowel extensions may produce other complications, namely bleeding, perforation, protein losing enteropathies, steatorrhoea, pancreatitis, appendicitis, and intussusceptions.^{3,6,7}

Imaging modalities of various types help in the diagnosis of bezoars, Ultrasonography has diagnostic efficacy of 88%.⁹ Computed tomography had 97% sensitive diagnostic modality in this case.⁹ GI endoscopy remains

the gold standard procedure for diagnosis.^{5,9} MRI has also been recently recommended for intestinal diseases.

Treatment for bezoars includes endoscopic removal, chemical dissolution, mechanical fragmentation, endoscopic biopsy devices and laser ignited mini explosive techniques which are used for small bezoar with variable success.^{7,12} Laparoscopic techniques are also being tried and results are promising.¹⁰ Laparotomy remains the sole treatment for large bezoars (Gastrostomy or enterostomy), especially in Rapunzel syndrome, where it has an extension in small bowel, whereas endoscopic can be used for removal of small bezoars.^{3,8,11,12}

Psychological support and psychiatric treatment is the corner stone for preventing recurrence.^{3,4}

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