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A rare case of Rapunzel syndrome with small bowel intussusception and bowel obstruction

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Abstract

Rapunzel syndrome is a rare disease that is characterized by a gastric trichobezoar with a long tail extending from the stomach to the small bowel. Patients can be asymptomatic for a long period, and symptoms develop later when the bezoar enlarges in size. The most common clinical presentations include chronic abdominal pain, malabsorption, gastrointestinal tract obstruction, gastrointestinal bleeding, and intussusceptions. We report a case of a 17-year-old girl with Rapunzel syndrome with intussusception presenting as small bowel obstruction. preoperative CT was only reported as intussusception and small bowel obstruction. A definite diagnosis was made only during laparotomy + enterotomy over the obstructed bowel site. it was a unique case of small bowel rapunzel syndrome.

Keywords: Rapunzel syndrome; Intussusception; Trichobezoar.

Introduction

Trichobezoar (hairballs) are unusual and are virtually exclusively found in young psychiatric patients. It is caused by the ingestion of hair, which remains undigested in the stomach. The hairball can lead to ulceration and gastrointestinal bleeding, perforation, or obstruction. Treatment consists of removal of the bezoar, which may require surgical treatment. In trichobezoar - hair is resistant to enzymatic digestion, resulting in impaction within the gastric body with the potential for extension through the pylorus and into the small bowel, termed Rapunzel syndrome [2]. We report a case of a 17-year-old girl with Rapunzel syndrome, but in this case, hairball was primarily noted in the small bowel causing bowel obstruction and intussusception.

Case report

A 17-year-old girl presented with severe abdominal pain, fever, and vomiting for 3 days. On physical examination palpable mass in the right side of abdomen with guarding and rigidity. Abdominal ultrasound revealed dilated small bowel loops with to and fro movements suggesting subacute bowel obstruction. Abdominal computerized tomography revealed significant dilatation of the small bowel with feces within - feces sign (Figure 1).

Telescoping of small bowel into small bowel - target sign is seen in right iliac fossa. In view of the acute presentation of the patient, we went for an urgent laparotomy, where we found largely dilated, congested small bowel loops. With ileoileal intussusception. It was reduced easily. After that when we tried to empty the contents within the small bowel into the caecum, content was not reduced manually, so we did enterotomy over most distended unhealthy site of the bowel, surprisingly we noted large ball of hair primarily in the small bowel and tail extended till the ileocaecal valve. Even though the head primarily noted in the small bowel the head of bezoar shape looks like stomach, it may be a gastric bezoar passed out into small bowel causing obstruction. The total length of the trichobezoar was 130 cm. This is the longest trichobezoar (Figure 3) not yet reported in any literature. Unhealthy looking small bowel of about 30 cm of bowel is resected and primary Anastomosis was done. The gastrointestinal tract was completely examined by palpa-

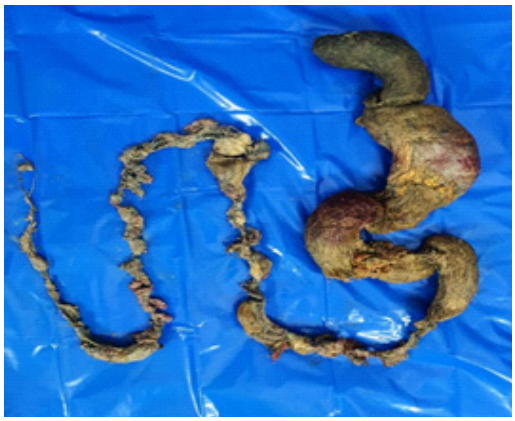

Figure 1: Extracted trichobezoar from our patient head of the bezoar shape like stomach

tion for other residual bezoars.pt made smooth post operative recovery, subsequent questioning of the patient gave no history of hair pulling or ingestion.

Discussion

Rapunzel syndrome is a rare condition, first described by Vaughan et al. as large, gastric trichobezoar that extends from the stomach through the duodenum and jejunum, and potentially to the ileocaecal junction. The term, "Rapunzel syndrome," derives from a fairy tale, written in 1812 by the Brothers Grimm [3]. Bezoars themselves are not dangerous, yet the potential for obstruction of the gastrointestinal tract and warrant urgent diagnosis and treatment [2]. We present a case of a trichobezoar with Rapunzel syndrome, longest trichobezoar, passed out the head of bezoar from the stomach into the small bowel. Trichobezoars are distinct from other bezoars in that hair can generate highly dense masses that are resistant to enzymatic digestion [2]. Furthermore, the smooth surface is thought to prevent hair from passing with peristaltic waves and to be retained within bowel. The extensive length of the bezoar inevitably served as a lead point for the Development of intussusceptions further along the small intestine [4]. Also trichobezoars can separate into smaller parts and cause obstruction at several levels of the gastrointestinal tract, The diagnosis of trichobezoars is often delayed, as they are not often recognized on initial presentation due to non-specific abdominal symptoms [2]. The initial use of plain radiography and sonography may hinder the diagnosis further. Reports have shown that 20%-60% of bezoars can be identified by plain film and Ultrasound. Computerized tomography can identify 97% of Bezoars in addition to determining the location, cause, and degree of obstruction [7]. Our case further highlights that even with CT radiologists not think about trichobezoar because primary bezoar in the small bowel with obstructive presentation. It is rare presentation of trichobezoar. The treatment of bezoars largely falls into three categories: medical dissolution, endoscopic removal and surgery [5]. Trichobezoars are typically resistant to medical dissolution. Endoscopic removal has been used in several cases but is often not successful, especially in large bezoar. Laparotomy should be considered the gold standard for large trichobezoars, especially if there is preoperative evidence for Rapunzel syndrome [2]. The decision to made enterotomy and/or gastrotomy varies on a case-by case basis and is largely based on the location, length, overall size, and intestinal viability. Post-operative psychiatric consultation is vital to prevent recurrence in those patients with trichophagia [6]. In our case Since it's a mobilized trichobezoar from the stomach we did UGI scope 4 weeks after surgery to rule out any residual trichobezoar.

Conclusion

We report a case of 17 year old girl with Rapunzel syndrome having the longest trichobezoar tail reported first time and also the extremely rare presentation of mobilized head of trichobezoar from stomach into the small bowel that primarily causing small bowel obstruction and intussusception. The diagnosis should be considered in patients with history of trichophagia. The treatment of choice is the surgical removal of mass and psychiatric treatment accompanied by behaviour therapy to prevent a recurrence.

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