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(CASE REPORT)

Rapunzel syndrome and small bowel Intussusceptions in a girl: A case report

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Abstract

Trichobezoars are indigestible masses of ingested hair commonly found in the stomach, often presenting with symptoms related to gastric outlet obstruction and severity related to the mass's size and location. Rapunzel syndrome is caused by gastric trichobezoar with extended tail and small bowel obstruction. Patients with gastric trichobezoar can be asymptomatic until the bezoar increases in size. We report a case of a 8 years old girl who visited the emergency department with abdominal pain. She was finally diagnosed with Rapunzel syndrome that causes multiple small bowel intussusceptions associated with trichophagia. Surgery was needed to reduce the multiple intussusceptions, and to remove the large trichobezoar. This case highlights to consider the possibility of Rapunzel syndrome when diagnosing the main cause of intussusceptions.

Keywords: Rapunzel syndrome; Trichobezoar; Intussusceptions; Abdominal mass

1. Introduction

Small bowel intussusception is unusual, representing 1%-10% of intussusceptions [1]. It can be caused by infection, Meckel's diverticulum, polyp, duplication cyst, tumor, hematoma or vascular malformation. Multiple, simultaneous small bowel intussusceptions are rare.

Bezoars are accumulations of undigested materials in the gastrointestinal tract. In a trichobezoar, the main component is a hair bundle, and gastric bezoar can lead to small bowel obstruction. Rapunzel syndrome is defined as gastric trichobezoar with extended tail into the small bowel causing its obstruction [2].

Patients may be asymptomatic or present with a spectrum of gastrointestinal symptoms, ranging from mild symptoms such as pain, nausea, and/or anorexia to more severe symptoms such as perforation or obstruction from mass effect or intussusception [2- 8]. Early diagnosis and treatment of bezoars are therefore vital, with the management approach largely dependent on the type of material ingested. Amongst bezoars at large, trichobezoars are unique; 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 [1,9-12]. We describe a child that presented with progressive obstruction from a trichobezoar with associated Rapunzel syndrome.

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2. Observation

We present a case of a 8-year-old child female with no significant past medical or psychiatric history presented with sharp, crampy abdominal pain of two weeks duration associated with nausea and non-bloody, non-bilious emesis with complaints of weight loss and loss of appetite .

On physical examination, a hard, nontender mass was palpated in the epigastrium raining the clinical suspicion of a trichobezoar. Blood tests were normal. Initial abdominal ultrasound performed at our institution showed the presence of an echogenic curvilinear mobile shadow in the stomach with a dense posterior acoustic shadow and multiple target lesions in the small bowel (Figure 1).

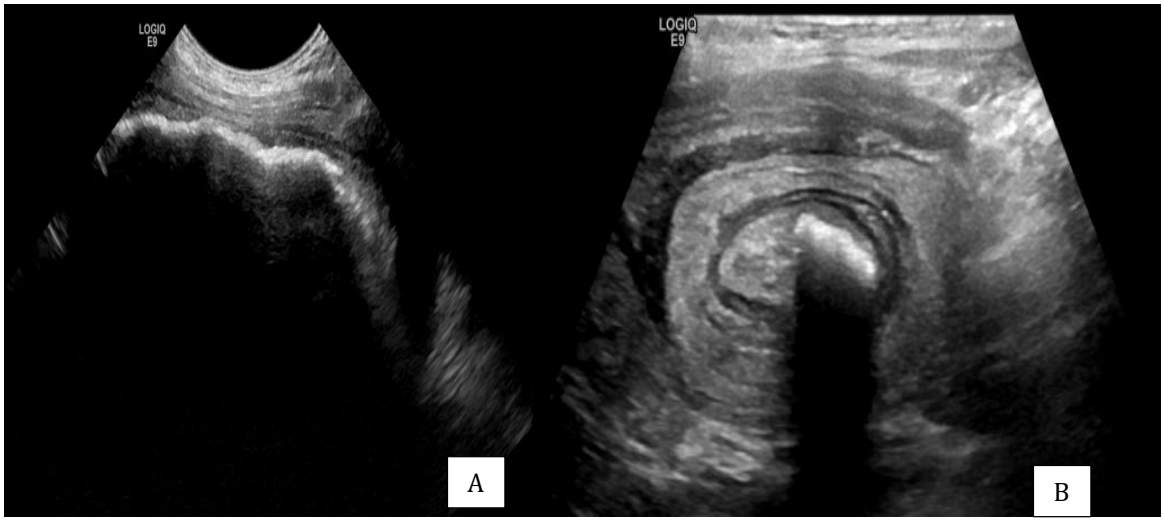

Figure 1 1-A :Abdominal ultrasound showing a curvilinear echogenic moving shadow in the stomach with dense posterior acoustic shadow . 1-B : Multiple target lesions are detected in the small bowel

Computerized tomography revealed a large gastric bezoar distending the stomach, likely resulting in partial gastric outlet obstruction. Multiple prominent short segments small bowel-small bowel intussusceptions were also noted (Figures 2A, 2B). The decision was made to take her to the OR for exploration and removal of the bezoar.

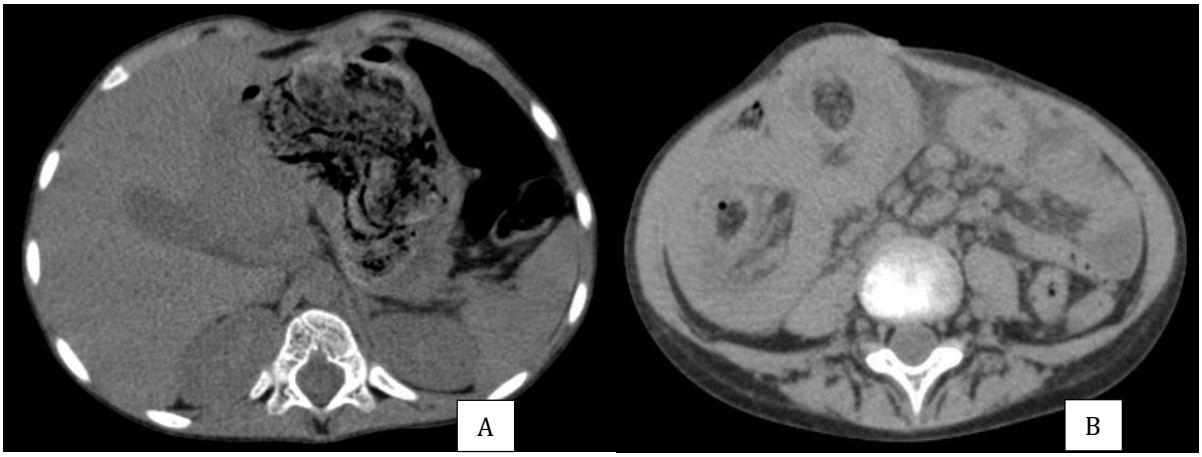

Figure 2 Axial computed tomography. (A) view showing heterogeneous mass in the stomach consistent with a bezoar. (B) view with evidence of small bowel to small bowel intussusception.

An upper midline laparotomy with an oblique gastrotomy was performed. Intraoperatively, demonstrated a large, organized, intraluminal bezoar with a mottled appearance due to entrapped air, extending into the duodenum , jejunum and proximal ilium. The trichobezoar was removed, and the gastrotomy site was closed . The intestines were then examined sequentially, which led to the discovery of multiple proximal intussusceptions that were each manually

reduced with an appreciation of viable tissue along the corresponding intestinal tract. No complications occurred, with minimal estimated blood loss.

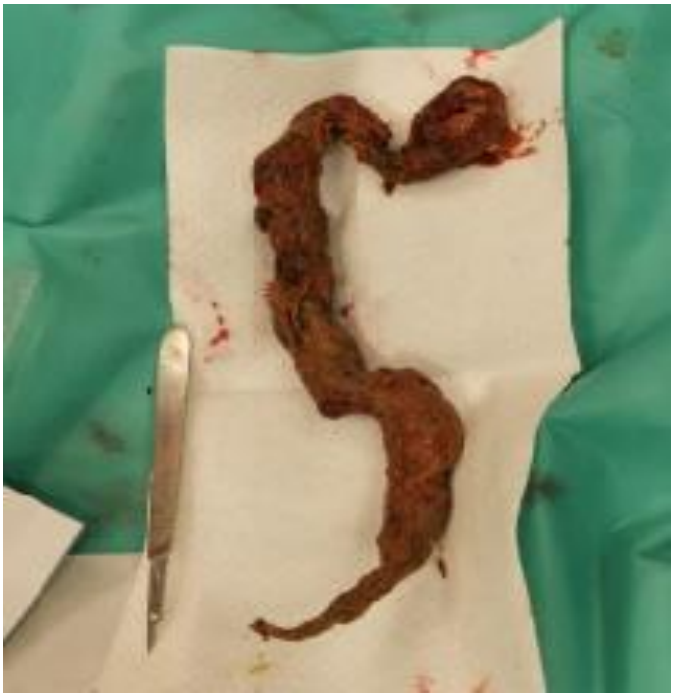

Figure 3 Picture of Trichobezoar after extraction

3. Discussion

Bezoars in and of themselves are not dangerous, yet the potential for obstruction of the gastrointestinal tract warrants urgent diagnosis and treatment. We present a case of a trichobezoar with Rapunzel syndrome leading to a proximal bowel intussusceptions. Trichobezoars are distinct from other bezoars in

that hair can generate highly dense masses that are resistant to enzymatic digestion. Furthermore, the smooth surface is thought to prevent hair from passing with peristaltic waves and to be retained within the gastric folds [3,13,14]. The extensive length of the bezoar inevitably served as a lead point for the development of intussusceptions further along the small intestine. While others have noted that trichobezoars can separate into smaller parts and cause obstruction at several levels of the gastrointestinal tract, this case highlights the potential for ischemia and necrosis at various levels due to concurrent intussusceptions from a single, contiguous object causing multiple lead points along its length [15-17]. The diagnosis of trichobezoars is often delayed, as they are not often recognized on initial presentation due to non-specific abdominal symptoms [13]. The initial use of plain radiographs and sonography may hinder the diagnosis further. Reports have shown that 20%-60% of bezoars can be identified by plain film and ultrasound [18]. The density of hair makes identification difficult, yet gastric distension and obstruction should be readily visible using these two methods. Computerized tomography is able to identify 97% of bezoars in addition to determining the location, cause, and degree of obstruction and other relevant sequelae. Our case further highlights the value of computerized tomography and demonstrates that bezoars should be included on the differential in patients with recurrent, nonspecific abdominal symptoms and atypical eating habits. Rapunzel Syndrome was first described by Vaughan et al. in 1968 and has since been reported with varying features [19]. A review of these cases identified three unifying criteria: (1) a trichobezoar with a tail, (2) extension of the tail at least to the jejunum, and (3) symptoms of obstruction [10]. We support these criteria and believe they appropriately represent the pathophysiologic processes found in our case. Prior reviews have suggested that it is a rare sequela of trichobezoars, but a recent case series diagnosed Rapunzel syndrome in 71% of its trichobezoar patients [11]. The treatment of bezoars largely falls into three categories: medical dissolution, endoscopy, and surgery [1]. Trichobezoars are typically resistant to medical dissolution [3]. Endoscopic removal has been used in several isolated cases but is often not successful, particularly for those cases with larger masses. A review by Gorter et al. identified 40 cases in which endoscopic removal was attempted with only two proving successful (5%) [13]. Attempts at endoscopic removal are often prolonged, delaying effective treatment. Up-front laparotomy should be considered the gold standard for large trichobezoars, especially if there is preoperative evidence for Rapunzel syndrome. The decision to pursue enterotomy and/or gastrotomy varies on a case-bycase 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 [20].

4. Conclusion

Rapunzel syndrome, a large gastric trichobezoar causing transient intussusception, is a very rare phenomenon. The diagnosis should be considered in patients with history of trichophagia. The treatment of choice is surgical removal of the mass and psychiatric treatment accompanied by behavior therapy to prevent recurrence.

Compliance with ethical standards

Acknowledgments

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Disclosure of conflict of interest

No conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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