CASE REPORT

Rapunzel Syndrome Presenting with Intussusception and Pancreatitis: A Case Report

Harish V Sutrave[1], Annapoorni Valliappan[2], Sivasankar Jayakumar[3]

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Abstract

Background: Bezoars are ingested foreign materials accumulated most commonly in the stomach as a hard mass. A gastric bezoar composed of hair (Trichobezoar) that has a long tail that extends beyond the pylorus throughout the small bowel and very rarely to the cecum is described as Rapunzel syndrome.

Case presentation: A 14-year-old girl presented with abdominal pain (4 days) with bilious vomiting (1 day). Contrast-enhanced computed tomography (CECT) scan showed a bezoar extending from the stomach to the small bowel within a jejunojejunal intussusception. Serum amylase levels were also elevated. The entire bezoar was extracted laparoscopically after reducing the jejunojejunal intussusception, subsequently, the patient recovered well and has been well after 2 years.

Conclusion: Patients with Rapunzel syndrome can present with intussusception or with pancreatitis, or rarely with both, as seen in our patient.

Keywords: Case report, Children, Ileoileal intussusception, Laparoscopy, Pancreatitis, Rapunzel syndrome, Trichobezoar.

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Background

Trichobezoar was first reported in the 18th century, when Baudamant described a 16-year-old boy with this condition.[1] trichobezoars occur in patients with recognized diagnosis of trichotillomania (compulsive desire to pull out one's hair) and trichophagia (repeated ingestion of hair).[2] About 10% of patients with trichotillomania demonstrate trichophagia.[3] Typically, a history of psychiatric illness and anxious behavior is noted in such patients with trichobezoars.

It can take at least 6 months for a trichobezoar to induce symptoms although different time courses have been reported.[4] The diagnosis is usually delayed in patients with trichobezoars, as generally they do not always give a clear history of hair ingestion. Plain abdominal X-rays often show a nonspecific mass appearance in the left upper quadrant. Upper gastrointestinal contrast studies reveal a filling defect within the stomach[5] but are not diagnostic. Ultrasound scan will demonstrate increased echogenicity secondary to intermixed hair, food, and air in the bezoar, and these are suggestive of bezoars. Computed tomography (CT) scan imaging shows heterogeneous mass with trapped air and is usually diagnostic.[6,7]

Trichobezoars are usually confined to the stomach, however, they can form more distal to the stomach as well and extend into the small intestine as a tail and such cases are described as Rapunzel syndrome.[8] We present a unique case of Rapunzel syndrome who presented with both pancreatitis and jejunojejunal intussusception, which we believe has not been reported so far. Herein, we discuss the presentation as well as the laparoscopy-assisted management of this case.

Case Presentation

A 14-year-old girl presented to our ER with epigastric abdominal pain for 4 days, repeated episodes of bilious vomiting for 1 day, and a prolonged reduced appetite. She had been well previously, and her parents initially did not report of any medical problems in the past. She appeared malnourished, dehydrated, and had tachycardia. She also had tenderness in the epigastric region along with guarding. She was fluid-resuscitated immediately, and her vitals stabilized. Except for slightly elevated serum amylase 134 U/L, lipase levels 89 U/L, and neutrophilia 81%, the rest of the blood tests were within the normal range. An initial ultrasound scan of the abdomen showed epigastric fullness and intraluminal small-bowel echogenic pathology, which was unclear. In view of this and suspected pancreatitis, a contrast-enhanced computed tomography (CECT) abdomen was done, which showed jejunojejunal intussusception along with a long abnormal intraluminal mass acting as a lead point within the intussuscepted bowel loops. The intussuscepted mass was seen originating from a large ovoid structure occupying the entire stomach (Fig. 1). This ovoid structure of mixed density and interspersed air specs within the stomach was suggestive of

[1,2]Department of Pediatrics, Apollo Specialty Hospitals, OMR, Chennai, Tamil Nadu, India

[3]Department of Pediatric Surgery, Apollo Specialty Hospitals, OMR, Chennai, Tamil Nadu, India

Corresponding Author: Sivasankar Jayakumar, Department of Pediatric Surgery, Apollo Specialty Hospitals, OMR, Chennai, Tamil Nadu, India, Phone: +91 9940468383, e-mail: shivajayakumar@gmail.com

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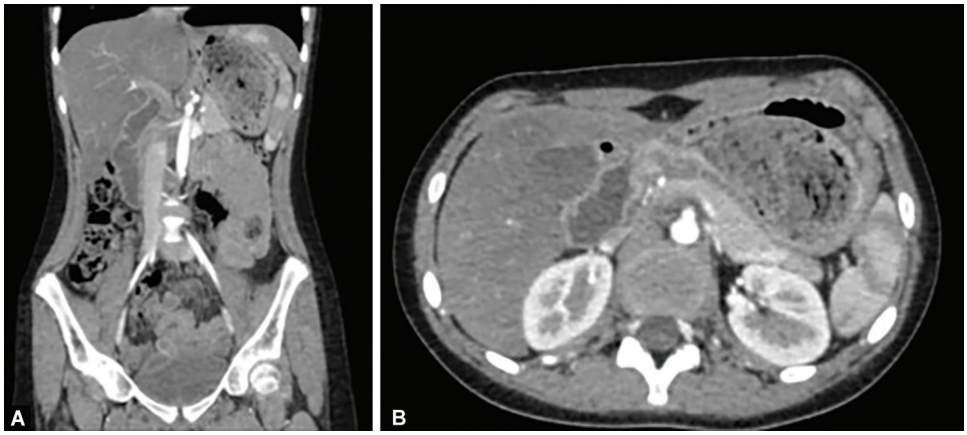

Figs 1A and B: CT images of our patient pre-op showing: (A) A large ovoid structure of mixed density and interspersed air specs occupying the stomach, consistent with bezoar; (B) Long segment of proximal jejunojejunal intussusception with oblong bezoar filling the lumen

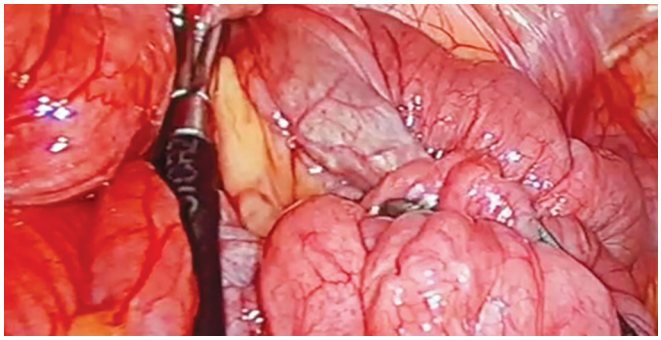

Fig. 2: Intraoperative laparoscopic picture showing jejunojejunal intussusception in our patient

a trichobezoar (ball of hair). On further inquiry, parents reported that their child had been ingesting hair in the past (6 years ago) but not recently.

In view of the CT findings, the child was taken up for emergency laparoscopy, after adequately counseling both parents and the child. Laparoscopic reduction of jejunojejunal intussusception (Fig. 2) was performed, followed by gastrotomy and excision of trichobezoar along with its 4-feet tail (Fig. 3), laparoscopically into an endo bag. The entire bezoar was brought out within the endo bag in toto through a small 4 cm pfannenstiel incision, and 2 drains were left *in situ*. Postoperatively, the child was kept nil oral with continuous nasogastric drainage, and broad-spectrum intravenous antibiotics continued. Serum electrolytes and trace elements were regularly monitored. Serum amylase levels returned to normal within few days. Cultures from the trichobezoar grew *Klebsiella pneumoniae* and *Escherichia coli*. Antibiotics were escalated and rationalized as per the sensitivity results.

However, soon she developed persistent low-grade fever, high bilious NG aspirates, and hypokalemia. Both drains had drained minimal serous fluid and stopped draining on day 3 postoperatively. Abdominal X-rays revealed dilated bowel loops, suggesting paralytic ileus, but her symptoms persisted. In view of this, an ultrasound scan of the abdomen was performed on day 4 following surgery. This showed multiple intra-abdominal collections with likely bowel adhesions and was confirmed by a repeat CT scan. The collections were drained, and adhesiolysis was performed laparoscopically on day 5. Intraoperatively, a small gastric leak was noted and was closed with an omental patch.

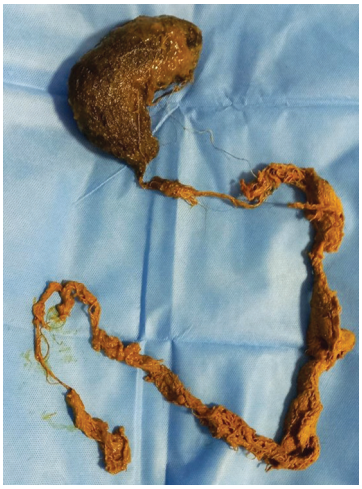

Fig. 3: Trichobezoar with its 4-feet long tail extracted via endo bag laparoscopically in our patient

Total parenteral nutrition (TPN) via a central venous line was commenced postoperatively. Subsequently, the fever subsided and the bilious NG aspirates reduced considerably. Oral feeds were introduced slowly to prevent refeeding syndrome. Serum electrolytes and trace elements were monitored regularly and were normal. She tolerated feeds very well, regaining her appetite, and walked home as a happy child with a total length of 10 days in the hospital. Both the patient and her parents were counseled adequately by our psychiatrist to address her trichophagia. After 2 years, the child, now an adult, is eating, drinking well, and had gained weight without any signs of recurrence of pancreatitis.

Discussion

Patients with Rapunzel syndrome can present with symptoms ranging from a painless mass per abdomen with early satiety, to bowel obstruction and frank gastrointestinal perforation.[9,10] Many associated complications with Rapunzel syndrome have been reported such as small-bowel intussusceptions, cholangitis, pancreatitis, hypoalbuminemia and biliary obstruction.[11] It has been postulated that the tail of the bezoar and the intraluminal strands interfere with small-bowel peristalsis, resulting in bowel dysmotility and acting as a lead point, predisposing such patients

to intussusception.[12] Another theory explained by Naik et al. is a purse-string effect due to shorter length of the bezoar tail compared with the length of the small intestine, leading to small bowel intussusception.[13] These small-bowel intussusceptions in Rapunzel syndrome have also been reported in patients with small-bowel obstruction[14] as well as in patients who were completely asymptomatic.[15] Our patient presented with bilious vomiting, which could have been secondary to either multiple high jejunojejunal intussusceptions causing bowel obstruction or pancreatitis, as both were seen in our patient.

Rapunzel syndrome is a very rare cause of pancreatitis but should be considered as one of the differential diagnoses. Pancreatitis due to obstruction of ampulla of vater by the bezoar has been reported.[16] Schreiber and Filston first described that the irritation by the bezoar tail extending into the duodenum causes edema and obstruction to the drainage of bile,[17] which explains the transient nature of the pancreatitis that resolves after removal of the bezoar as seen in our patient. Derangement of liver enzymes, acute cholangitis, and cholestasis are also some of the rare reported biliary complications caused by the bezoar.[17] Although 10 cases of pancreatitis associated with Rapunzel syndrome have been reported so far,[11] none of them report on both pancreatitis and jejunojejunal intussusception seen in Rapunzel syndrome, as seen in our patient.

Upper gastrointestinal endoscopy can establish the diagnosis of a gastric bezoar; however, endoscopic retrieval is difficult and rarely the definitive treatment.[18] The initial treatment of trichobezoars and Rapunzel syndrome is almost exclusively surgical. Most of these cases reported laparotomy for removal of the bezoar. However, laparoscopy-assisted removal of large and long bezoars is feasible but more challenging,[19–21] resulting in less scarring for the already mentally traumatized patients. In our patient, the entire 4-feet-long bezoar was removed in an endo bag laparoscopically assisted with a small pfannenstiel incision, and we recommend laparoscopic approach in all such cases.

Psychiatric evaluation is an important component of the treatment and should be done in every such patient to prevent relapse. Psychiatric intervention is effective in treating trichophagia and should be considered as a standard component of treatment.[22] Recurrence requiring reoperation has been reported in very few patients and is mostly due to lack of psychiatric follow-up.

Conclusion

Trichobezoars are usually confined to the stomach; however, they can extend as a tail into the small intestine, and such cases are described as Rapunzel syndrome. Patients with Rapunzel syndrome can be asymptomatic or present with bowel obstruction secondary to intussusception or with pancreatitis, or rarely with both, as seen in our patient.

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