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Fungal Peritonitis with Fungus Balls, a Complication of Trichobezoars and Rapunzel Syndrome

Authors' Contribution:
Study Design A
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Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
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Patient: Female, 7
Final Diagnosis: Rapunzel syndrome
Symptoms: Abdominal pain
Medication: —
Clinical Procedure: Laparotomy
Specialty: Surgery

Objective: Unusual clinical course
Background: Rapunzel syndrome is a rare condition involving the extension of bezoars from the stomach to the distal gastrointestinal tract. Laparotomy remains the gold standard treatment for this condition because of the size of the bezoars. Although bacterial peritonitis is a known complication of laparotomy in Rapunzel syndrome, very few cases of post-surgical fungal peritonitis have been reported in these patients.

Case Report: In this case report, we present a case of Rapunzel syndrome complicated by post-surgical fungal peritonitis and formation of fungus balls. To our knowledge, fungal peritonitis with fungus balls has never been reported in the English literature as a Rapunzel syndrome complication.

Conclusions: It is important to cover *Candida* and other fungi with an antifungal regimen in pediatric patients with Rapunzel syndrome pre- and post-surgery. In addition, prolonged fever and septic symptoms post-surgery warrant a search for peritoneal fungus balls that are not simply responsive to anti-fungal therapy and may necessitate repeat laparotomy.

MeSH Keywords: Bezoars • Fungi • Peritonitis

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Background

Bezoars are residual foreign materials in the gastrointestinal (GI) tract, mainly in the stomach. Given the type of the residual material, bezoars are called by different names including phytobezoars, pharmaco-bezoars, trichobezoars, lactobezoars, and foreign body bezoars [1]. Trichobezoars are more common in psychiatric patients with trichotillomania and trichophagia. Rapunzel syndrome is a rare condition involving the extension of bezoars from the stomach to the distal GI tract. There is still controversy about the exact definition of Rapunzel syndrome. Extension of the bezoar tail up to the ileocecal junction, extension of bezoar tail to jejunum, and a bezoar of any size causing intestinal obstruction has been described as a definition of Rapunzel syndrome by different authors [2,3].

The Rapunzel syndrome can be asymptomatic for a long period of time. The clinical presentation is mainly because of gradual enlargement of bezoar causing various degrees of obstruction. The most common symptoms are abdominal pain, nausea and vomiting, bowel obstruction, and peritonitis. Other less common presentations include weight loss, anorexia, hematemesis, intussusception protein-losing enteropathy, iron deficiency, and megaloblastic anemia. Gastric ulcer, obstructive jaundice, acute pancreatitis, duodenojejunal fissuration, and gastric emphysema have been reported in Rapunzel syndrome secondary to large bezoars [3]. Despite recent advances in minimally

invasive surgery, laparotomic surgery remains the gold standard treatment for this condition because of the size of the bezoars. Post-surgical complications including peritonitis are not uncommon in these patients. Although fungal peritonitis can happen secondary to perforated peptic ulcers unrelated to bezoars, very few cases of post-operative fungal peritonitis have been reported as complications of Rapunzel syndrome. In this manuscript we described a case of Rapunzel syndrome which became complicated by fungal peritonitis after laparotomy.

Case Report

The patient was a 7-year-old malnourished female with a 1-year history of endoscopy-proven trichobezoars presenting with abdominal pain. A prior endoscopic attempt to remove the bezoar had been unsuccessful. Despite medical advice, the parents decided not to proceed with surgery after the endoscopic attempt. The patient was first evaluated in our Emergency Department because of abdominal pain and distention, and nausea and vomiting. At the time of presentation, the patient was anemic with a hemoglobin of 6 g/dL. Abdominal computed tomography (CT) scan was consistent with a large bezoar in the stomach with extending tail toward the jejunum in association with evidence of small bowel obstruction, as seen in Rapunzel syndrome (Figure 1A).



Figure 1. Coronal abdominal computed tomography shows distended stomach with a large bezoar (A). The removed trichobezoar from the stomach (B).

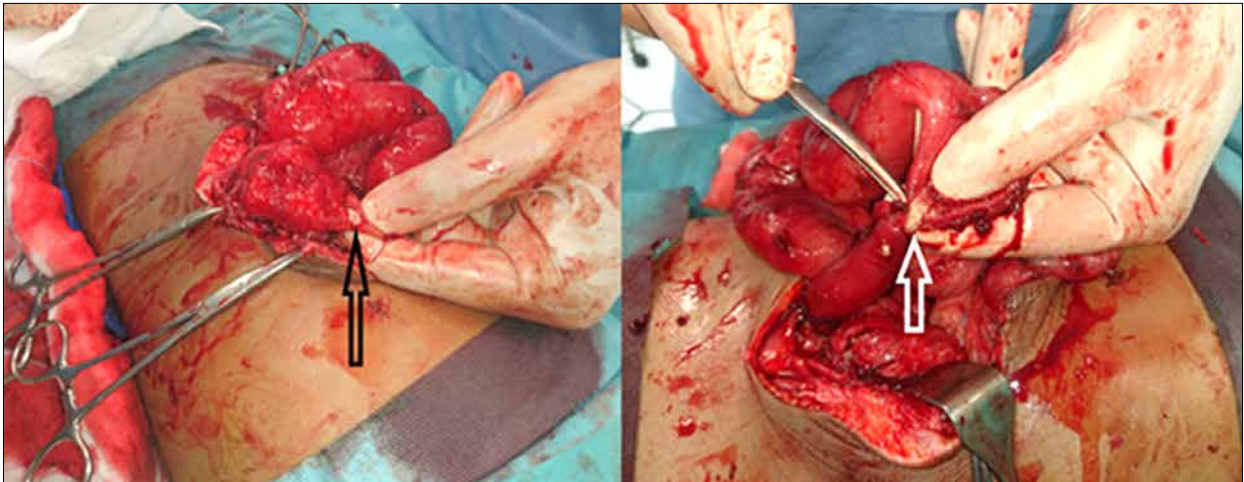


Figure 2. Images during the second laparotomy demonstrate white nodules over the anti-mesenteric surface of small bowel (arrows) which were proven to be fungus balls on subsequent fungal smear.

It was deemed necessary to treat the patient with laparotomy given the prior unsuccessful endoscopic attempt and evidence of small bowel obstruction. At the time of surgery, a large bezoar was noted within the stomach (Figure 1B) extending to the small bowels consistent with Rapunzel syndrome. The stomach and proximal small bowel were distended but without evidence of ischemia, ulceration, or perforation. The large trichobezoar was removed from the stomach by single gastrotomy incision without definite visible peritoneal contamination. Several fragments of the trichobezoars were removed from duodenum and proximal jejunum by milking, and the peritoneal cavity was irrigated. No enterotomy was performed. The patient received prophylactic antibiotics including cefazolin and metronidazole.

Three days after surgery, the patient became febrile and on physical examination generalized tenderness were noted in her abdomen. To treat the presumed peritonitis, cefazolin was discontinued and vancomycin and meropenem were initiated. Four days post-surgery, ultrasound was performed because of abdominal pain which demonstrated evidence of free abdominal fluid without evidence of bowel obstruction. Abdominal pain continued and 9-days post-surgery another abdominal ultrasound revealed evidence of a loculated right lower quadrant with right paracolic and midline fluid collections. Percutaneous ultrasound-guided drainage was performed on the abdominal collections. Eleven days post-surgery, cultures came back positive for *Candida*, so amphotericin B was added to the treatment regimen. Subsequent ultrasounds failed to demonstrate improvement of the multi-loculated abdominal collections.

Due to persistent fever and elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), 25 days after the initial surgery, another laparotomy was required. The multi-loculated collections were drained and the peritoneal cavity

irrigated. During the surgery, fungus balls were noted within the peritoneal cavity (Figure 2). After laparotomy removal of the fungus balls, the fever subsided but the post-operative period was complicated by an enterocutaneous fistula initially managed conservatively by total parenteral nutrition (TPN) for 7 weeks, and subsequently surgically closed. The patient recovered well without additional complications. Psychiatric consultation was performed after surgery which was consistent with anxiety disorder. Behavior therapy was performed to prevent recurrence.

Discussion

Bezoars form when ingested material in the GI tract cannot be digested and builds up. Based on the type of ingested material, bezoars are classified into phytobezoars, pharmacobezoars, trichobezoars, lactobezoars, and foreign body bezoars [1]. Trichobezoars are relatively rare and caused by hair accumulation in the GI tract. These are almost exclusively seen in young females and are associated with psychiatric disorders. Trichobezoars are usually limited to the stomach but can rarely extend to the distal GI tract including the small bowel and even the colon, which is a phenomenon known as Rapunzel syndrome [2,3]. Patients with Rapunzel syndrome typically present with symptoms including abdominal pain, nausea, vomiting, GI bleeding, anorexia, weakness, weight loss, and an abdominal mass [2,3]. Anemia is associated with this syndrome as well. In Rapunzel syndrome, contrast upper GI studies and CT scans are helpful for diagnosis, and ultrasound can also identify the corresponding abdominal mass. Definitive diagnosis is performed with endoscopy [1,2].

Various treatment approaches have been described for trichobezoar/Rapunzel syndrome including laparotomy, laparoscopic

surgery, endoscopic removal of the bezoar, and chemical dissolution [2–4]. Classically, management of Rapunzel syndrome involves removal of the mass and recurrence prevention. Treatment further depends on the bezoar size and the type of ingested material. Endoscopic removal of the mass is successful in the case of phytobezoars and lactobezoars because of their typically small size. Endoscopic removal of trichobezoars, however, is often challenging and incomplete because of bezoar size. Currently, the main role of endoscopy in trichobezoars is diagnosis [2]. Only a few case reports of successful laparoscopic removal of bezoars in the pediatric population have been published [2]. The main advantages of laparoscopic bezoar removal are that it has a lower rate of postoperative complications, a shorter post-surgical hospitalization time, and better cosmetic outcome. Disadvantages include a longer surgery time and higher chance of peritoneal contamination [1,4]. Laparoscopic removal of bezoars can be challenging due to the size of the mass as well as satellite lesions [2]. Laparotomy is considered the standard treatment for pediatric bezoars/Rapunzel syndrome with a complication rate of 12% including GI tract perforation, wound infection, pneumonia, fecal leakage, and ileus [1,4].

Our patient's post-surgical fungal peritonitis and the presence of *Candida glabrata* fungus balls is unusual. Although *Candida* peritonitis is a relatively common complication of perforated peptic ulcers unrelated to bezoars [5], based on our knowledge, fungal peritonitis secondary to bezoars has been reported only once with positive cultures [1]. To our knowledge, peritoneal "fungus balls" have never been reported in English literature secondary to a bezoar. Although the fungal peritonitis is more common in patients with liver/renal failure and

immunosuppression, the only known medical condition in our patient was malnourishment. In our patient, no "gross" leakage and peritoneal contamination was detected during operation and peritoneal cavity irrigated. We believe fungal peritonitis was likely because of microscopic peritoneal contamination. Given the high mortality (21%) of secondary *Candida* peritonitis [5], we believe it is important to cover infections caused by *Candida* and other fungi with an antifungal regimen in pediatric patients with Rapunzel syndrome, pre- and post-operatively. In addition, our case demonstrated that prolonged fever and septic symptoms post-operatively warrants a search for peritoneal fungus balls that are not simply responsive to anti-fungal therapy and may necessitate repeat laparotomy.

Conclusions

We reported an unusual case of trichobezoar which was complicated by fungal peritonitis and fungus ball formation after laparotomy. Laparotomy is now the standard of care for large trichobezoars and Rapunzel syndrome, but can be complicated by fungal peritonitis and fungus ball formation within the peritoneal cavity. Consideration of this complication is important when managing these patients, especially in cases with signs of post-surgical peritonitis. Fungal peritonitis necessitates modification of the treatment regimen by adding anti-fungal medication and even repeat laparotomy to remove the fungus balls.

Conflict of Interest

None.

References:

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