Case Report

Successful Laparoscopic Removal of a Huge Trichobezoar in Cases of Rapunzel Syndrome in Children

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ABSTRACT

Rapunzel syndrome is a very rare condition. The trichobezoar, in cases of Rapunzel syndrome, extend from the stomach into the duodenum and small bowel. Trichobezoars are usually encountered in young women with psychiatric problems, such as trichotillomania, trichophagia, or mental retardation and pica. Large trichobezoars, which are associated with Rapunzel syndrome, are removed during open surgery which requires large incisions. This Case Report describes 2 girls who had Rapunzel syndrome where the trichobezoars reached the jejunum and laparoscopic surgery was successful in the removal of the trichobezoars. Laparoscopic removal of a trichobezoar can be considered as a treatment option for children with Rapunzel syndrome.

Keywords: bezoar, children, laparoscopy, Rapunzel syndrome, trichobezoar

Introduction

Human hairs are effectively foreign bodies when they are ingested. Hair has a smooth surface resistant to peristaltic propulsion and digestion in the stomach. At first, ingested hair becomes caught between gastric mucosal folds where more hair accumulates and becomes entangled (due to peristalsis) trapping food, and this develops into hard hairballs coated in mucus, called trichobezoars [1]. The location of trichobezoars is generally confined to the stomach.

Rapunzel syndrome is primarily a psychological condition resulting in the formation of trichobezoars which form a tailed extension from the hairball mass in the stomach to the duodenum and small bowel [2]. Rapunzel syndrome named after the fairy tale heroine from a Brothers Grimm story who had very long, blond hair [3].

Though Rapunzel syndrome is very rare, the consequences of this condition need prompt treatment. In most of the reported cases of Rapunzel syndrome, trichobezoars (the foreign body), were removed during laparotomies. There were only a few reported cases where the condition was treated with laparoscopic surgery [1].

Here, we report 2 cases of Rapunzel syndrome which had caused huge gastric trichobezoars which were removed successfully by laparoscopic surgery.

Case Report

1. Case 1

A 13-year-old girl was referred to the Emergency Department due to worsening epigastric pain and vomiting for several days. She had intermittent epigastric pain and vomiting once a week for 3 months. She had a 5-month history of chewing her hair subconsciously. Her vital signs were stable, and she had mild tenderness and a palpable mass which were revealed...
in the epigastric area on physical examination. Laboratory examination showed normal findings except for the complete blood cell counts, which showed hypochromic microcytic anemia, with a hemoglobin concentration of 7.0 g/dL, and a mean corpuscular volume of 58.5 fL. A computed tomography (CT) scan revealed a 10 × 7 cm mass, which was considered a bezoar in the stomach (Figures 1A and 1B). An upper gastrointestinal endoscopy was performed but failed in the removal of the bezoar and revealed a huge trichobezoar with 2 ulcerative craters on the greater curvature side of the antrum. The patient underwent an emergency laparoscopic exploration using 4 trocars in the sub-umbilicus, right lower quadrant, right upper quadrant, and left upper quadrant. The gastric wall was opened at the anterior wall of the stomach along the greater curvature, and a trichobezoar that had passed into the small intestine, was identified (Figure 2). Rapunzel syndrome was diagnosed. After the trichobezoar had been removed from the gastrointestinal tract, it was retrieved with a Lapbag (Sejong Medical, Seoul, Korea), and the gastrotomy site was closed using a V-loc 3-0 (Covidien, New Haven, CT, USA) continuous suture (Figure 3). While retrieving the trichobezoar, the umbilical port was widened because the bezoar could not be passed through the 11 mm trocar site. Prophylactic antibiotics were used pre- and postoperatively (cefotetan 50 mg/kg). The postoperative course was uneventful. The patient was discharged on Day 8 after surgery and was treated for Rapunzel syndrome by psychiatric services to address her trichophagia, and her gastric ulcer was treated by gastroenterology services.

2. Case 2

A 6-year-old girl was referred to the outpatient clinic due to vomiting and intermittent abdominal pain. She had vomited every morning, unrelated to meals for a few months. She was born prematurely, at 32 gestational weeks. She had no other medical history except for trichotillomania, which was diagnosed 4 years earlier but she had not been prescribed medications or other treatments because her parents thought it would improve over time. Her vital signs were stable, and the physical examination revealed no abnormal findings except for a palpable mass-like lesion in the upper abdomen. The laboratory test results were within the normal ranges. A CT scan revealed a huge bezoar in the stomach reaching to the duodenum (Figures 4A and 4B). An upper gastrointestinal endoscopy suggested a diagnosis of Rapunzel syndrome. A mass of hair filled the whole stomach and had reached into the duodenum (Figure 5). Prophylactic antibiotics were used pre- and postoperatively (cefotetan 50 mg/kg). The girl underwent laparoscopic exploration using 4 trocars. After opening the gastric wall along to the greater curvature of the anterior wall, the trichobezoar was removed with a Lapbag (Figure 6). Widening of the umbilical port was also needed. The gastrotomy site was closed using a V-loc 3-0 continuous suture.
The patient was discharged on Day 8 after surgery without complications. She began treatment with psychiatric services for trichophagia and trichotillomania.

**Discussion**

Trichobezoars are caused by the ingestion of hair and associated with Rapunzel syndrome. Trichotillomania and trichophagia are psychiatric disorders of Rapunzel syndrome involving hair pulling and the ingestion of the pulled hair, respectively. About 10% of the patients with trichotillomania had trichophagia as part of their compulsive behavior. The disorders are also associated with mental retardation, and pica. Therefore, after the removal of the trichobezoar foreign body, these patients need psychiatric treatment for trichotillomania and trichophagia to limit their compulsive behavior and avoid the recurrence of a trichobezoar [4,5].

Since the first report of Rapunzel syndrome by Vaughan et al in 1968 [6], about 50 cases have been reported. The patient ages have ranged from 4 to 19 years old, and except for 1 case, all patients were girls. Most cases were treated with open laparotomy for the removal of the trichobezoars resulting in a good prognoses, except for 3 patients who died [3].

Typically, patients showed nonspecific or a lack of symptoms initially. Thus, trichobezoars may not be recognized delaying diagnosis whilst the trichobezoar continues to increase in size and weight due to the continued ingestion of hair [1,7]. The most commonly reported symptoms are epigastric pain and discovery of a mass (70%). Nausea and vomiting (64%), hematemesis (61%), weight loss (38%), and diarrhea and constipation (32%) have also been reported. [8] The increase in size and mass of trichobezoars can lead to more severe complications, including mucosal erosion, ulceration, and perforation of the stomach or the small intestine. Obstructive jaundice, protein-losing enteropathy, intussusception, and pancreatitis also have been reported [1,7].

The gold standard for the diagnosis of a trichobezoar is endoscopy. However, it cannot reveal the co-existence of Rapunzel syndrome. Both CT and intraoperative findings and patient history are needed for diagnosing Rapunzel syndrome. The CT findings of a trichobezoar show a non-enhanced, well-described, heterogeneous, intraluminal mass with a mottled appearance. The heterogeneous and mottled appearances come from different densities of composition, including food debris and air bubbles [9].

The gold standard of treatment is traditionally open laparotomy. Endoscopic treatments have been tried but were only successful in a few patients and have limitations [8,10]. Laparoscopic treatment is also challenging however, smaller scars and less invasiveness are obvious advantages of the laparoscopic approach [5,11]. To prevent the complications caused by remnant trichobezoar, especially in Rapunzel syndrome where there is a long extension of the trichobezoar into the small bowel, inspection from the stomach to the small bowel is needed to remove all of the trichobezoar. It is more difficult and more time-consuming than an open laparotomy. The risks of contaminating the abdominal cavity and wound with hair fragments is also a problem. A wide extension of the trocar site is sometimes needed and increases the appearance of the scar [1,5,11]. Repairing the gastrotomy site would be more difficult than performing open laparotomy. It is not difficult or time-consuming for experts in laparoscopic surgery to navigate from the stomach to the small intestine. To remove the trichobezoar, a plastic retriever bag and an Alexis O wound protector/retractor (Applied Medical, CA, US) was used which helped to avoid contamination of the abdominal cavity and the wound, and reduced the extension of the trocar site. For repairing the gastrotomy site using a self-anchoring barbed suture like a V-Loc, made repair easier. In the 2 cases presented, the trichobezoars only reached the proximal jejunum. This may be a factor that has made laparoscopic removal possible. If the trichobezoar had reached the distal small intestine, it may be difficult to remove laparoscopically.

In summary, when girls or young women, especially those with psychological problems, have atypical abdominal pain or gastrointestinal symptoms, trichobezoar and Rapunzel
syndrome should be included in the differential diagnosis, although they were very rare. Laparoscopic removal of trichobezoars can be considered a treatment option for children with Rapunzel syndrome.

Conflicts of Interest

The authors have no conflicts of interest to declare.

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