CASE REPORT

Rapunzel Syndrome- A case report With Review of Literature

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ABSTRACT
A 15-year-old girl with average IQ has had abdominal pain for the past 6 months, which has been gradual in onset, progressive dull aching kind, and low in intensity. She had been experiencing tremendous discomfort and weight in her belly for the past ten days, as well as a hard mass in her upper abdomen. In addition, the patient had a history of weight loss. There was no history of nausea, vomiting, or other symptoms that could indicate intestinal obstruction. Patient was scheduled for a laparotomy with gastrostomy under general anaesthesia after obtaining adequate consent due to the bulky nature of the trichobezoar. For four days after surgery, the patient was kept on hyperalimentation and ryle's tube aspiration. A single Packed red blood cell (PRBC) was transfused. On the fourth day, the patient was allowed to eat and was discharged on the seventh day in good condition, with the recommendation to see a psychiatrist. The recovery period was unremarkable. On Day 10, the sutures were removed.

Keywords: Children, Rapunzel syndrome, Trichobezoar, Trichophagia

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INTRODUCTION

The majority of bezoars are made up of indigestible organic matter, such as hair trichobezoars, vegetable and fruit phytobezoars, or a combination of both, however other uncommon substances have been identified in the literature. Trichobezoars are widespread in people with psychiatric disorders, although only around half of them have a history of trichophagia. Trichobezoars are the most frequent kind of bezoar, accounting for 55 percent of all cases. Hair strands are held in the folds of the gastric mucosa, and peristalsis leads them to become entangled until a ball forms that is too enormous to pass into the stomach, resulting in gastric atony.

These hairs become matted together and form a stomach shape, usually in a single mass. Rapunzel syndrome [1,2] is a rare occurrence in which they infiltrate the small intestine and cause intestinal blockage. Patients who go untreated may develop gastric outlet obstruction, GI bleeds, and, in rare cases, stomach necrosis and perforation.

In the 1812 fairy tale by the Grimm Brothers, a maiden named ‘‘Rapunzel" had her long hair spill out of her prison tower, allowing her prince to free her [3]. The illness is known as Rapunzel Syndrome because the tail of a trichobezoar extending into the small intestine resembles Rapunzel's hair (RS) [4].

In individuals with a history of psychiatric disease, a thick compact mass of hair (trichobezoar) was observed in the stomach with extension into the intestine through the duodenum, which was originally described in the literature by Vaughan et al. in 1968[4]. Hair pulling is referred to as trichotillomania, and hair chewing is referred to as trichophagia. In young psychiatric patients, the appearance of RS might range from asymptomatic to gastrointestinal ulcers, blockage, or perforation. In this report a 15-year-old adolescent girl presented with pain in the abdomen for 6 months.

CASE PRESENTATION

A 15-year-old girl with average IQ has had abdominal pain for the past 6 months, which has been gradual in onset, progressive dull aching kind, and low in intensity. She had been experiencing tremendous discomfort and weight in her belly for the past ten days, as well as a hard mass in her upper abdomen. In addition, the patient had a history of weight loss.

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There was no history of nausea, vomiting, or other symptoms that could indicate intestinal obstruction.

The girl was 158 cm tall and weighed barely 28 kg when she was examined. She had no patchy baldness or halitosis and appeared pale. Except for a large solid palpable (12 cm) spherical lump extending from the epigastrium to the upper umbilical region with smooth margins, mobile with respiration, slight side to side mobility, tender on touch, no local rise in temperature, no signs of peritonitis, dull on percussion, bowel sound present, systemic examination was within normal limits.

Outside ultrasonography (USG) revealed an ill-defined curvilinear echogenic line extending from the epigastric lesion to the left lumbar legion, leaving an acoustic shadow, suggesting trichobezoar. An overdistended stomach with a big heterogeneous intraluminal mass with mottlet gas appearance (figure 1) was seen in the stomach, extending through the pylorus into the duodenum and further into the proximal jejunum, according to a contrast enhanced CT whole abdomen performed in our institution. The duodenum is 3.5 cm long and dilated. The mucosa of the jejunum seems moderately edematous. Although the small intestinal loops are not dilated, the symptoms suggest trichobezoar (Rapunzel Syndrome).

Patient was investigated preoperatively for fitness for surgery. Her hemoglobin content was 11.0 g/dL, Hematocrit- 32.5 % and a mean corpuscular volume (MCV) of 89.5 fL.The white blood cell count was 5.5 10^9/L. The routine blood examination and blood biochemistry examination results were within normal limits.

Patient was scheduled for a laparotomy with gastrostomy under general anaesthesia after obtaining adequate consent due to the bulky nature of the trichobezoar. Long anterior gastrotomy was performed intraoperatively, and trichobezoar filling the entire dilated stomach was discovered. The distal boundary reached the proximal jejunum, which was beyond the duodenum. There was no sign of gastric necrosis or infection within the peritoneal cavity. As a result, the Rapunzel syndrome diagnosis was confirmed.

Extraction was attempted but failed due to inability to mobilise the lower limit. The distal section of the bezoar was moved proximally into the duodenum after an enterotomy in the proximal jejunum. By pulling at the stomach side and pressing from the jejunal side, simultaneous extraction of bezoar in its whole without breaking was tried and successfully retrieved (figure 2).

For four days after surgery, the patient was kept on hyperalimentation and ryle's tube aspiration. A single Packed red blood cell (PRBC) was transfused. On the fourth day, the patient was allowed to eat and was discharged on the seventh day in good condition, with the recommendation to see a psychiatrist. The recovery period was unremarkable. On Day 10, the sutures were removed.

Figure 1: CECT whole abdomen showing stomach with a big heterogeneous intraluminal mass

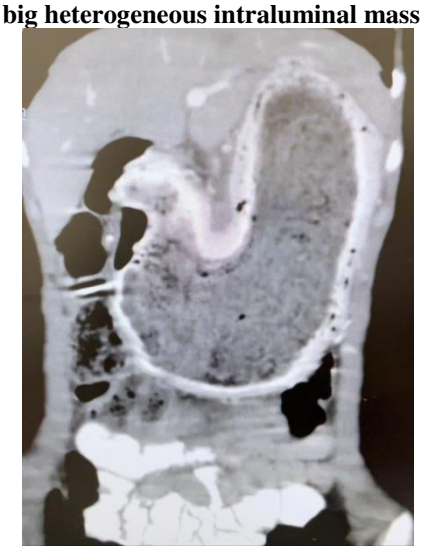
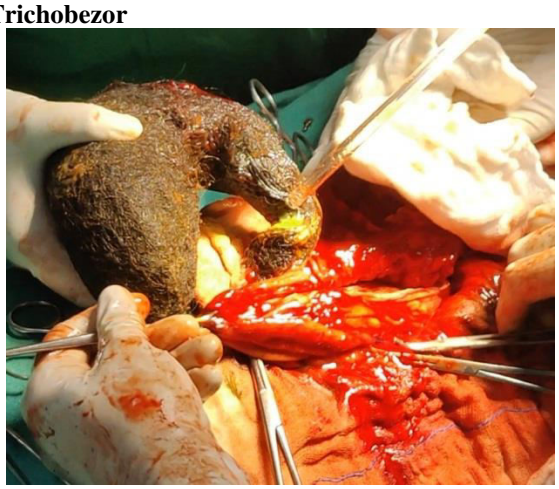


Figure 2: Intraoperative Enbloc Retrival of Trichobezor



DISCUSSION AND REVIEW OF LITERATURE

A bezoar is a clump of food or foreign material that forms in the gastrointestinal tract and stays there. The word "bezoar" is derived from the Arabic word "bedzehr" or the Persian word "Padzhar," both of which signify "protection against poison." Bezoars from animal stomachs have been utilised as precious stones and antidotes to poisons throughout history. It's a part of Chinese traditional medicine. Phytobezoars (induced by vegetables), trichobezoars (caused by hair), lactobezoar (caused by milk), and miscellaneous are the four groups (caused by medication; pharmacobezoars, tissue papers and or fungus). In 1779, during an autopsy of a patient who died of gastric perforation and peritonitis, the first mention of a bezoar in a human was made [4,5].

Since the first description, less than a hundred cases of Rapunzel syndrome have been described in the literature. The trichobezoar in Rapunzel syndrome spreads from the pylorus into the small intestine [4]. The majority of trichobezoar cases are documented in females with traditional long hair, but one male case

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was reported who ate his sister's hair [6]. The majority of trichobezoar patients have psychiatric disorders such as trichotillomania (hair plucking) and trichophagia (eating hair).

Ultrasound and computed tomography are two diagnostic techniques (CT). Although CT scans have a high accuracy rate, ultrasonography in these situations is extremely sensitive [7]. Upper gastrointestinal endoscopy is still the best way to diagnose intragastric trichobezoar because it allows you to see the hair threads and try extraction [8]. Trichobezoar has been treated using a variety of therapy approaches.

Cristina Oana Marginean described a 13-year-old girl who was admitted to the hospital with abdominal pain, anorexia, and weight loss. The patient weighed 32 kg and had generalised baldness, a palpable lump in the epigastric area, and abdominal pain when palpated. Anemia was discovered in the laboratory tests. A stomach intraluminal tumour with a superior hyperechoic arc was discovered on abdominal ultrasonography. The upper digestive endoscopy revealed a mass made up of hair, mucus, and food that occupied the gastric cavity and extended into the duodenum, confirming the Rapunzel syndrome diagnosis [9].

Rajesh Godara et al [10] described an 18-year-old female who had been suffering from dull painful epigastric discomfort, vomiting, loss of appetite, and weight loss for 18 months. She was malnourished and had a palpable bulge in her epigastrium when examined. A big trichobezoar was discovered after further investigation and imaging. She had a gastrostomy and a hair ball was removed. The recovery period was unremarkable.

A twenty-year-old girl had a stomach trichobezoar with a slender tail reaching into the duodenum, according to Shrestha R et al [11]. Trichophagia had been a problem for her in the past. She had epigastric pain and was vomiting. An attempt at endoscopic removal was made, however it failed. Following that, the patient had an exploratory laparotomy and a gastrotomy. She made a full recovery and was discharged with the recommendation of a mental consultation.

Badawy MK [12] described a three-year-old Caucasian child who had been experiencing intermittent abdominal pain, distention, non-bilious vomiting, and constipation for two weeks. Her doctor prescribed laxatives, but she had no noticeable side effects. Due to her reduced oral intake and weight loss, she was subsequently admitted to the local hospital. She underwent a laparotomy and gastrostomy to remove the trichobezoar and recovered quickly. She was given an iron supplement and was directed to a nutritionist as well as child mental health services.

A 17-year-old girl presented to the emergency clinic after experiencing a syncopal episode, according to Jamaall Jackman et al [13]. On inspection, there was a lump in the left upper quadrant with no symptoms of peritonism. A huge trichobezoar was discovered on contrast-enhanced CT, which resulted in stomach rupture and intra-abdominal free fluid. The patient underwent laparotomy and gastrotomy, and had an uneventful recovery with psychological examination prior to discharge. Despite their rarity, bezoars should be considered in our differential diagnosis since, due to their size and weight, they can appear in a variety of ways. With large gastric bezoars, there is a risk of stomach perforation in this scenario.

CONCLUSION

When young girls come with abdominal pain, vomiting, and anaemia, Rapunzel syndrome should be evaluated as a differential diagnosis. These issues can be avoided with early detection. Recurrence can be avoided with psychiatric diagnosis, counselling, and therapy. Trichobezoar is frequently found alongside psychiatric disorders.

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