

A Rare Presentation of Rapunzel Syndrome without Intestinal Obstruction with Review of Literature

Neha Garg¹, Surender Verma², Pradeep Garg³, Anil Mehta⁴, Vijay Pal¹, Shivika Agarwal¹

From ¹Junior Resident, ²Assistant Professor, ³Senior Professor, ⁴Senior Resident, Department of General Surgery, PGIMS Rohtak, Haryana, India

ABSTRACT

Trichobezoar with Rapunzel syndrome is an uncommon diagnosis. Its clinical presentation without intestinal obstruction is rare. It is predominantly found in mentally retarded or emotionally disturbed youngsters. They typically cause abdominal pain and vomiting but can also present as asymptomatic abdominal mass progressing to obstruction and perforation; however, our case of trichobezoar presented with early satiety and loss of weight. The diagnosis was confirmed on computed tomography scan. The trichobezoar was removed by the anterior gastrotomy under general anesthesia. The post-operative period was uneventful. We recommend conventional laparotomy and anterior gastrotomy for large size trichobezoars extending to the duodenum and jejunum as the treatment of choice. Psychiatric consultation is recommended to prevent relapse.

Key words: Anterior gastrotomy, Rapunzel syndrome, Trichobezoar, Trichophagia, Trichotillomania

Bezoars are defined as the accumulation of concretions of human or vegetable fibers in the gastrointestinal tract. The word bezoar is derived from the Arabic word “bedzehr” or the Persian word “Padzhar” meaning protecting against poison [1]. There are different types of bezoars depending on their composition such as trichobezoars, phytobezoars, lithobezoars, pharmacobezoars, lactobezoars, plasticobezoars, and cottonbezoars [2]. Trichobezoar is the most common type of bezoar. These consist of hairballs or hair-like fibers caused by swallowing hair or other indigestible materials. These are associated with underlying psychiatric disorders and commonly present in adolescents during the 2nd decade of life in females. It mainly occurs in females due to the presence of traditional long hairs.

Human hairs are resistant to digestion and peristalsis due to their smooth surface and thus accumulate between the mucosal folds of the stomach. Over time, the impaction of hair with mucus and food leads to the formation of trichobezoar usually in the gastric body and hence found prepylorically [3,4]. Some of them may pass through the pylorus into the duodenum, jejunum, ileum, and even into the colon. This condition is termed as Rapunzel syndrome [5]. We are reporting here the case of Rapunzel syndrome in a 24-year-old girl with trichobezoar with a long tail that passed into the jejunum.

CASE REPORT

A 24-year-old unmarried girl presents with a complaint of pain in the upper abdomen for 2 years. The pain was mild, dull, and colicky in nature, and there were no aggravating or relieving factors. There was a history of early satiety, chronically decreased appetite, and weight loss for the past 6 months. There was the fullness of the abdomen after meals. There was no history of acid reflux, diarrhea, fever, melena, hematemesis, or any recent illness. The patient’s mother admitted that she had a history of pulling out of her hair and swallowing them when she was 10 years old. The patient also had a history of slightly retarded mental development. The patient had poor academic performance and unable to do simple calculations; however, she was able to do her household work properly. The patient had a history of seizures 2 years back, but she was currently not on any antiepileptic or antipsychotic medications. Her menstrual history was normal.

On examination, the patient was thin built, malnourished, and was looking anxious. Her height was 140 cm and weight was 40 kg. Her vitals were as follows: Pulse rate 96/min, respiratory rate 16/min, and blood pressure 108/70 mmHg. She had no patchy alopecia or halitosis. There was no evidence of jaundice or lymphadenopathy. The abdominal examination revealed a hard, non-tender, well-defined, and palpable mass from the epigastrium to the periumbilical region measuring approximately 8×6 cm. The mass was non-pulsatile and was moving well with respiration.

Correspondence to: Surender Verma, 117-A, Sector 14, Rohtak, Haryana, India. E-mail: drsrurn@gmail.com

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There was no evidence of intestinal obstruction. Per rectum examination was normal.

Her hemoglobin was 10.2 mg/dl and white blood cell count was $5.9 \times 10^9/L$. The rest of the routine blood examination and biochemistry was within normal limits. The plain radiograph showed air-fluid levels with a density within the stomach. Abdominal ultrasound showed a hyperechoic curvilinear mass with dense acoustic shadow. A computed tomography (CT) scan demonstrated a large intraluminal iso- to hypodense area with whorl appearance and multiple air and loculi in the stomach and duodenum extending into the proximal jejunal loop with multiple calcific foci and marked distension of the stomach and duodenum suggestive of trichobezoar (Fig. 1). There was no thickening of the gastrointestinal wall or any other sign of necrosis. Upper gastrointestinal endoscopy revealed a giant gastric trichobezoar.

Endoscopic therapy was not attempted as the hairball was large and was made of tight coils of long hairs with extension beyond the pylorus. Thus, due to the bulky size of trichobezoar, an exploratory laparotomy was planned. The patient was given general anesthesia and an orotracheal tube was inserted. An upper midline incision was given. The stomach was opened with a longitudinal incision along the anterior wall of the stomach (Fig. 2a). The stomach was filled with large trichobezoar and a

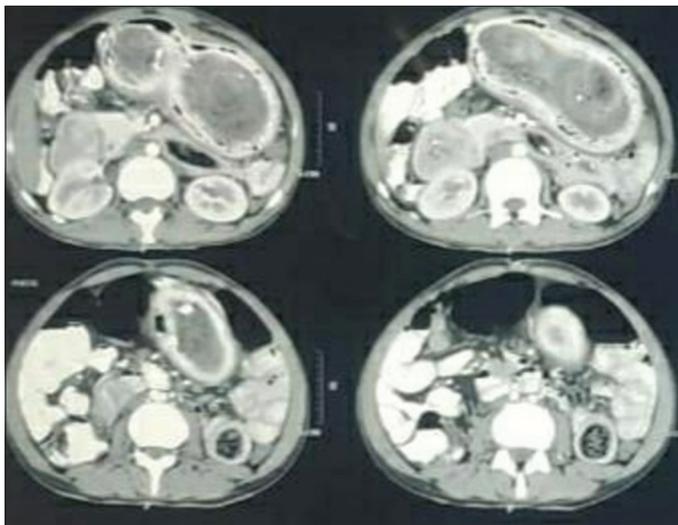


Figure 1: Computed tomography – transverse plane showing trichobezoar

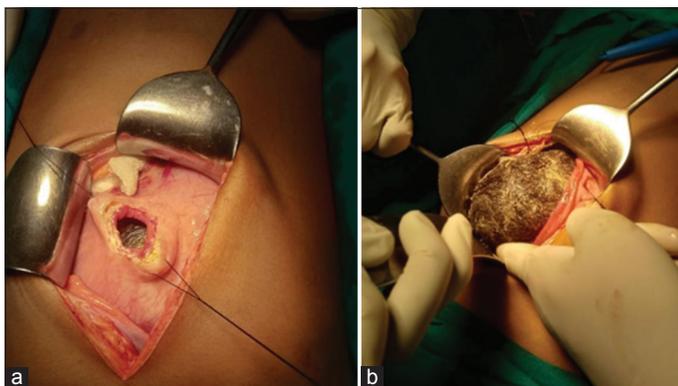


Figure 2: (a) Trichobezoar visible through anterior gastrotomy incision; (b) large Trichobezoar protruding through the stomach

large tail extending through the duodenum into the upper part of jejunum (Fig. 2b). There was no evidence of gastric necrosis or intraperitoneal infection. Hence, a diagnosis of Rapunzel syndrome was confirmed. The mass was engaged in one piece like a fetus which is delivered from the uterus (Fig. 3).

The total length of trichobezoar was 100 cm and weight was 2850 g. The mass was a perfect cast of the stomach, pylorus with a tail. Ryle's tube was inserted in the intraoperative period and was removed on the 4th post-operative day. The patient was allowed orally on the 5th post-operative day. There was no post-operative complication. The wound was clean and healed well. The psychiatric consultation was done and the mother was educated regarding behavior management. On the follow-up, the patient's appetite improved and had gained weight with no active complaints. Consent was taken from the patient regarding the publication of the case.

DISCUSSION

Trichobezoar rarely presents as Rapunzel syndrome. It was first described by Vaughan *et al.* in 1968 [5]. It is essentially associated with a history of pulling and ingestion of hairs or other indigestible materials. Gastric dysmotility may be another factor associated with it [6]. These disorders are often the result of psychological problems or mental retardation [7]. These are mostly seen in females, as seen in our case also. We have demonstrated the case of a patient who was diagnosed and treated for Rapunzel syndrome. The parents of the patient revealed that the patient had a habit of picking and swallowing hairs. These are often not recognized by the initial presentation and the diagnosis is often delayed.

Symptoms of trichobezoar can be non-specific such as abdominal pain which can be due to distension and ulceration, fatigue, nausea, vomiting, early satiety, weight loss, halitosis, and epigastric mass [8]. In cases of Rapunzel syndrome, the presence of a tail in the intestine can result in peristaltic movements, leading to colicky abdominal pain and this was also the cause of colicky pain in our case. Other psychiatric disorders, such as pica, obsessive-compulsive disorder, depression, and anorexia nervosa, may also be associated with it.

When not recognized early, it continues to grow in size and weight, leading to a risk of severe complications such as gastric mucosal erosion, ulceration, and even perforation of the stomach or small intestine. In addition, intussusception, obstructive jaundice, pancreatitis, and even death has been recorded as



Figure 3: Stomach trichobezoar with a long tail

complications. On physical examination, a palpable, well-defined, and non-tender mass can be found in the epigastrium. Abdominal imaging examinations include plain radiographs, ultrasound, upper gastrointestinal barium series, CT scan, and magnetic resonance imaging. However, the final diagnosis should be confirmed through upper gastrointestinal endoscopy [9].

Upper gastrointestinal endoscopy is considered as the gold standard for the diagnosis of trichobezoar [10]. The acidic contents of the stomach denature the hair proteins and gives the bezoar its black color [11]. However, it may not show the presence of a coexisting Rapunzel syndrome. Hence, the diagnosis of Rapunzel syndrome can be made by a combination of CT scan and endoscopy before surgery. When not recognized, these can result in severe complications the most common of which are gastric or intestinal perforation. Other complications include gastric ulcer, obstruction, intussusception, obstructive jaundice, gastric emphysema, protein-losing enteropathy, iron deficiency anemia, megaloblastic anemia, and reactive pancreatitis [12]. Due to a significantly large size of trichobezoar, the blood supply to the mucosa of the stomach and part of the intestine is hampered resulting in a perforation.

The treatment depends on the size of the bezoar, their location, and their composition [13]. Although medical treatment and enzymatic degradation appear attractive due to non-invasiveness, these are ineffective [9]. Other treatment options include removal by endoscopy, laparoscopy, or by laparotomy. Due to a high success rate, low complication rates, low complexity, and the ability to examine the entire gastrointestinal tract in a short period of time, laparotomy with the anterior gastrotomy is the treatment of choice in Rapunzel syndrome.

When the gut loops are damaged or show necrosis or perforation, bowel resection may be required. Endoscopy should not be tried as it can result in breaking off parts of bezoar and their slippage into the small intestine which cannot be removed endoscopically [14]. However, small trichobezoars or phytobezoars can be removed endoscopically. Laparoscopy was found successful incidentally in trichobezoar extending until the duodenum, but the removal of the tail requires careful manipulation of the bowel to prevent perforation. Sometimes multiple enterotomies can occur.

Although the recurrence of Rapunzel syndrome is extremely rare, there is no doubt that the prevention of recurrence is important. The most common reason for recurrence is the loss to follow-up the patients and, therefore, did not complete their psychological treatment. Hence, professional psychotherapy and long-term follow-up are critical after surgical removal of trichobezoars [15,16].

CONCLUSION

Non-obstructing Rapunzel syndrome is a rare variety; however, it should always be considered as a differential in female patients presenting with vague abdominal pain, recurrent vomiting, weight loss and history of hair pulling, and eating. The diagnosis is confirmed by the upper gastrointestinal endoscopy and CT scan. Laparotomy with anterior gastrotomy is the treatment of choice. Counseling by psychiatrists is an important part to prevent recurrence.

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