

Case Report

‘Rapunzel syndrome’ and a giant gastric trichobezoar in young girls: a report of two case.

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Abstract

A bezoar is a mass of undigested material within the gastrointestinal tract. The term bezoar derives from the Arabic word Badzehr, which means antidote [1]. Bezoars were used as antidotes against plague, snake-bite, leprosy, and epilepsy by physicians from 12th to 18th century [2], Rapunzel syndrome is an unusual presentation of a trichobezoar, with female predominance and a mean age of 10.8 years. The patients commonly present with abdominal pain, nausea, vomiting and signs of obstruction. The distal end of the bezoar may be in the jejunum, ileum or the colon. Here, we report a 23-year-old college student, of this uncommon syndrome, who had a trichobezoar in the stomach with a tail extending into the ileum,we also describe a case of a young female with a giant gastric trichobezoar that weighed 1230 grams and a small intestinal trichobezoar (375 g).

Keywords:

Trichobezoar, Rapunzel syndrome, intestinal obstruction,

Background

Bezoar is a tightly packed collection of undigested material that is unable to exit the stomach, Most bezoars are of indigestible organic matter such as hair (Trichobezoars) or vegetable and fruit (phytobezoars),or a

combination of both but other rare substances has been also been described in literature. A trichobezoar is a mass of undigested hair within the gastrointestinal tract. Trichobezoars are often associated

with trichotillomania (hair pulling), and trichophagia (hair swallowing). Trichotillomania may be unconsciously or unintentionally done and is part of the DSM IV psychiatric classification of impulse control disorders [3, 4]. Only 50% will have history of trichophagia. Trichobezoars have been described in literature and they comprise 55% of all bezoars. Trichobezoars most commonly occur in adolescent females

Case presentation

Case 1

A 23-year-old, college student girl, was referred to the surgical clinic in 2007, with a history of abdominal pain, distension, weight loss and attacks of vomiting followed meal and fluid intake. This history was on and off for almost six months. She was treated in different primary health care as a case of gastroenteritis and parasitic infestation. Despite the treatment she was receiving there was no improvement in her condition, in fact she was getting worse.

In the surgical clinic, abdominal palpation revealed an oblong mobile well-defined mass occupying the upper half of the abdomen, the mass was not tender and was firm in consistency. Routine blood investigations were normal. An erect plain X-ray of the abdomen showed few gas filled

[5]. The site of hair pulling is most commonly from the scalp, but can occur from the eyelashes, eyebrows, and pubic area [6].

In very rare cases the Rapunzel Syndrome hair extends through the pylorus into the small bowel causing symptom and sign of partial or complete gastric outlet obstruction [15-19].

small bowel loops with a few air fluid levels. Ultrasound examination of the abdomen revealed multiple minimally dilated fluid filled bowel loops.

Upper Gastrointestinal endoscopy revealed a Trichobezoar occupying almost the whole gastric cavity, an attempt to remove it by foreign body forceps failed and the forceps was barley pulling few fibers of this huge ball of hair.

Conservative management for sub-acute small bowel obstruction with intravenous fluids and nasogastric decompression failed to improve her clinical condition over 24 h. Midline exploratory laparotomy was then performed to open the stomach (Gastrotomy) between two Vicryl stay suture. A huge

Trichobezoar was identified which took the shape of the stomach (Figure 1).



Figure 1:midline Exploratory Laparotomy showing stomach and jejunum loop

There was a long tail of hair extending through the pylorus into the small bowel(Figure 2).



Figure 2: a long tail of hair extending through the pylorus into the small bowel.

By this feature the diagnosis was clear of a Rapunzel syndrome. A distal enterotomy was performed and the remaining part was dislodged gently. Both the opening in the gastric wall and small bowel was closed with

continuous Vicryl. The patient had an uneventful postoperative course and was discharged after six days. The patient was advised to visit the psychiatric clinic evaluation and management.

Case 2

A 20-year-old, single female, college student, patient presented with chief complaints of epigastric discomfort and vomiting for six months, progressive loss of appetite for four months, pain and lump in abdomen for two months. She had lost weight, but no history of dysphagia, hematemesis or melena. On examination, the patient was thin built well looking female with normal basal parameters.

Abdominal examination revealed a well-defined, smooth and hard intra-abdominal lump of 8 x 10cm in the epigastric region. The lump was having palpable curved (convex) lower border but upper border not reachable (fingers could not be insinuated between the lump and costal margin). The lump was moving with respiration and dull on percussion. Differential diagnosis of

gastric trichobezoar and gastric malignancy was made clinically.

Abdominal ultrasound showed a large mass in the epigastric region with echogenic anterior margin and dilated loops of bowel.

Plain radiograph of the abdomen showed multiple air fluid levels with dilated small intestinal loops and a sizable soft tissue density within the stomach are seen (**Figure3**).



Figure 3: Plain radiograph of the abdomen showed multiple air fluid levels with dilated small intestinal loops and a sizable soft tissue density within the stomach.

Endoscopy showed mass in the stomach. Contrast Enhanced Computed Tomography (CECT)

showed grossly distended stomach with heterogeneous intraluminal content, 17.5X7.9 cm, showing areas of curvilinear

band like hyper densities and scattered lucencies suggestive of Gastric Trichobezoar. The small bowel was diffusely dilated due to a focal intraluminal mass.

The suspicion of a trichobezoar was confirmed by upper digestive endoscopy that demonstrated the presence of a foreign body described as a "hair bolus" hindering endoscopic access (**Figure 4**).



Figure 4: upper digestive endoscopy that demonstrated the presence of a foreign body described as a "hair bolus" hindering endoscopic access.

Prompted by the investigations, we enquired further, to which she admitted about habit of hair eating and auditory hallucination, since the age of 11. A young sister, is under psychiatrist care, with a diagnosis of schizophrenia.

Based on the above findings we decided to perform surgery for the trichobezoar.

Hospital psychiatrist was consulted for evaluation of any associated psychiatric illness and counselling/treatment. An informed consent was taken from the patient and her father. Trichobezoar was removed by upper midline Exploratory Laparotomy with anterior gastrotomy and enterotomy, under GA (**Figure 5**).



Figure 5: huge Trichobezoar was identified which took the shape of the stomach.

A giant trichobezoar was removed through an anterior gastrotomy. Gastric mucosa inspected and there was no sign of any complication (ulceration, haemorrhage etc.).

The mass weighed 1230 g and measured 23x17 cm (**Figure 6**).



Figure 6: The mass weighed 1230 g and measured 23x17 cm.

There was another small mass (375 g) in the end of jejunum which was removed through an enterotomy. In our patient, it is likely that parts of the tail of the gastric trichobezoar had broken off and migrated to the small intestine, causing intestinal obstruction,

Discussion

Trichobezoar, a hair ball in the proximal gastrointestinal tract, is a rare condition almost exclusively seen in young females [7, 8]. Human hair is resistant to digestion as well as peristalsis due to its smooth surface. Therefore it accumulates between the mucosal folds of the stomach. Over a period of time, continuous ingestion of hair leads to the impaction of hair together with mucus and food, causing the formation of a trichobezoar. In most cases the trichobezoar is confined within the stomach. In some cases, however, the trichobezoar extends through the pylorus into jejunum, ileum or even colon. This condition, called Rapunzel syndrome, was first described by Vaughan et al. in 1968 [9].

This syndrome was originally described by Vaughan et al. in 1968 [9]. The commonly accepted definition is that of a gastric

trichobezoar with a tail extending to the jejunum, ileum or the ileocecal junction. The common presentation of trichobezoar is in young females usually with an underlying psychiatric disorder. In our case the presentation is in a very young age with hair extending down to the small bowel, causing symptoms, which could mimic gastrointestinal infections and infestation especially in endemic areas. Rapunzel Syndrome is a rare form of trichobezoar. It is named after a tale written in 1812 by the Brothers Grimm about a young maiden, Rapunzel, with long hair who lowered her hair to the ground from a castle, which was a prison tower to permit her young prince to climb up to her window and rescue her.

Majority of cases of trichobezoar present late, due to the low index of suspicion by the physician. In a series of 131 collected cases of trichobezoar, a palpable abdominal mass

the gastrotomy and enterotomy were in two layers and the abdomen was closed with drainage. The patient presented a satisfactory progression and was discharged at the 7th postoperative day, being referred to the psychiatric ambulatory for follow-up.

Majority of cases of trichobezoar present late, due to the low index of suspicion by the physician. In a series of 131 collected cases of trichobezoar, a palpable abdominal mass

was present in (87.7%), abdominal pain (70.2%), nausea and vomiting (64.9%), weakness and weight loss (38.1%), constipation or diarrhea (32%) and hematemesis (6.1%). The laboratory investigations revealed low hemoglobin in about 62% (average) [20]. When not recognized, the trichobezoar continues to grow in size and weight due to the continued ingestion of hair. This increases the risk of severe complications, such as gastric mucosal erosion, ulceration and even perforation of the stomach or the small intestine. In addition, intussusception, obstructive jaundice, protein-losing enteropathy, pancreatitis and even death have been reported as complications of (unrecognized) trichobezoar in the literature [10-14]

The complications of Rapunzel syndrome ranges from attacks of incomplete pyloric obstruction to complete obstruction of the bowel to perforation and peritonitis. [21-23]. Patient with Trichotillomania (a psychological condition that involves strong

Conclusion

Trichobezoar is a rare condition that may pose a diagnostic challenge. Although rare, trichobezoar should not be overlooked as a differential diagnosis in young female

urges to pull hair), around 30% will engage in trichophagia, and of these, only 1% will go on to eat their hair to the extent requiring surgical removal [24]. Less than half of the patients give a history of trichophagia. There has been few cases of recurrence following successful surgery [25].

While, the gold standard in the diagnosis of trichobezoar remains upper GI endoscopy which also allows therapeutic intervention, a CT scan of the abdomen will usually reveal the trichobezoar. The diagnostic accuracy of CT is reported to be 73 to 95% [26]

In the early stages endoscopic removal is not without risk of bowel perforation and should be reserved for small Trichobezoars only [26]. Other methods including the use of laser ignited mini-explosive technique were used successfully [26]. Laparoscopy has been also used with limited success. Open surgery still remains the corner stone of large Trichobezoar removal especially if it has an extension into the bowel, which might be missed with other methods of treatment.

patients, with history of trichophagia, whom should undertake endoscopic examination as early as feasible. Diagnosis can be easily made with the use of CT scan and endoscopy. Management almost

always requires surgical removal. Also, recurrences are known. A psychiatric evaluation, counselling and treatment are helpful in preventing these recurrences.

Many patients with trichobezoar have psychiatric pathology with emotional

problems, family discord, and history of neglect or mental retardation. Thereby, counseling by a psychiatrist is an important part of management to prevent recurrence.

Conflict of Interest:

None.

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None

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