E-ISSN: 2616-3470
P-ISSN: 2616-3462

www.surgeryscience.com
2019; 3(4): 529-532
Received: 23-08-2019
Accepted: 26-09-2019

Dr. Siddhesh Vora
Associate Professor, Department of General Surgery, CU Shah Medical College and Hospital, Surendranagar, Gujarat, India

Dr. Chintan Godhani
Resident Doctor, General Surgery, CU Shah Medical College and Hospital, Surendranagar, Gujarat, India

Dr. Soham Patel
Resident Doctor, General Surgery, CU Shah Medical College and Hospital, Surendranagar, Gujarat, India

Corresponding Author:
Dr. Siddhesh Vora
Associate Professor, Department of General Surgery, CU Shah Medical College and Hospital, Surendranagar, Gujarat, India

Rapunzel syndrome in a 7-year-old boy: A case report

Dr. Siddhesh Vora, Dr. Chintan Godhani and Dr. Soham Patel

DOI: https://doi.org/10.33545/surgery.2019.v3.i4i.298

Abstract
Background: Trichobezoar is a condition in which hair mixed with food material is formed in stomach. Rapunzel's syndrome is a rare variant of trichobezoar where the mass in stomach extends through pylorus into duodenum & varying length of small intestine & sometimes into colon.
Case presentation: 7-year-old male patient presented in Surgery OPD with recurrent attacks of acute epigastric pain, vomiting and loss of appetite; was diagnosed with Rapunzel Syndrome, especially where the trichobezoar was not suspected at all because of the negative history of trichophagia. The purpose of reporting this case is the rare occurrence of such condition discussing the presentation, diagnostic modalities & the ideal option of surgical treatment.
Conclusion: Trichobezoar or Rapunzel syndrome in children is rare and there are many factors associated with trichophagia. The clinical presentation is usually late but can be diagnosed by ultrasound, CT scan or Upper GI Scopy. In spite of several therapeutic options used, laparotomy is still considering the treatment of choice.

Keywords: Trichobezoar, Rapunzel syndrome, Trichophagia, Upper GI Scopy, Laparotomy

Introduction
The term bezoar refers to swallowed material (either food or foreign body) that fails to clear from the stomach and accumulates into masses of concretions [1]. Bezoars can be classified into many types: phytobezoar (vegetable); trichobezoar (hair); lactobezoar (milk/curd), pills (Pharmacobezoar) and miscellaneous (wool, cotton, sand, paper, etc). Although the prevalence of bezoars in humans is low, if untreated, have associated mortality rates as high as 30% primarily because of gastrointestinal bleeding, destruction or perforation. The most frequent type of bezoar in children and teenage girls is trichobezoars, while phytobezoars are more often found in adult [2, 3].
The pathogenesis of bezoars is not consensual. Human hair is resistant to digestion as well as peristalsis & it is believed that the smooth surface of hair does not allow for its propagation through peristalsis, being trapped between the mucosal folds of the stomach. Over a period, continuous ingestion of hair leads to its impaction together with mucus and food, causing the formation of a trichobezoar. However, the stomach of normal individuals is able to clear even large foreign bodies in up to 80 to 90% of the cases which may imply that bezoar formation occurs in the presence of altered gastric anatomy or physiology (e.g. after gastric surgery) or continued ingestion of the offending substance in case of underlying psychiatric problem, such as trichotillomania (the irresistible urge to pull one's own hair) and swallow it (trichophagia) and also pica in some cases [4].
Usually, the trichobezoar is confined within the stomach but in some cases extends through the pylorus into jejunum & various lengths of the intestine & this is called "Rapunzel Syndrome". The name "Rapunzel" syndrome comes from the German fairy tale in the collection assembled by the Brothers Grimm (a 12-year-old princess who was prisoned into a tower with neither stairs nor doors by a witch, the prince then rescues her by climbing up the tower's walls with the help of Rapunzel's long tresses) [5].
Trichobezoars are very difficult to be recognized in early stages due to their nonspecific presentation or even lack of symptoms but later it continues to grow in size and weight due to the continued ingestion of hair and may present with signs and symptoms of acute abdomen. These include abdominal pain, nausea, bilious vomiting, hematemesis, anorexia, early satiety, weakness, weight loss and palpable abdominal mass, depending on the degree of obstruction [2, 3]. This condition if untreated has increased 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. The diagnosis of trichobezoars is based on imaging study. Although ultrasonography is effective in detecting an epigastric mass, CT-scan is more accurate in revealing a characteristic bezoar image and allowing the identification of the presence of additional gastrointestinal bezoars. The definite diagnosis is established by endoscopy [2, 6].

Case report

A 7-year-old boy presented with history of recurrent crampy epigastric pain along with nausea, non-bilious vomiting & loss of appetite. Initial Family history was unremarkable & parents denied any abnormal feeding habit with no other medical or surgical history.

On examination the child was well built, no sign of malnutrition was apparent & there was no alopecia. Abdominal examination revealed mild abdominal distention with a soft, non-tender mass in the epigastric region which moved with respiration. Bowel sounds were normal. Laboratory values were within normal limits.

Ultrasound revealed a large a heterogeneous mass within the stomach (Figure 1). Upper GI Scopy revealed a Trichobezoar occupying almost the whole gastric cavity (Figure 2). Retrospective detailed history taking from the boy revealed a long history of hair & fur eating mostly from ingesting hair, carpet & clothing fibres from the ground with evidence of subnormal mentality.

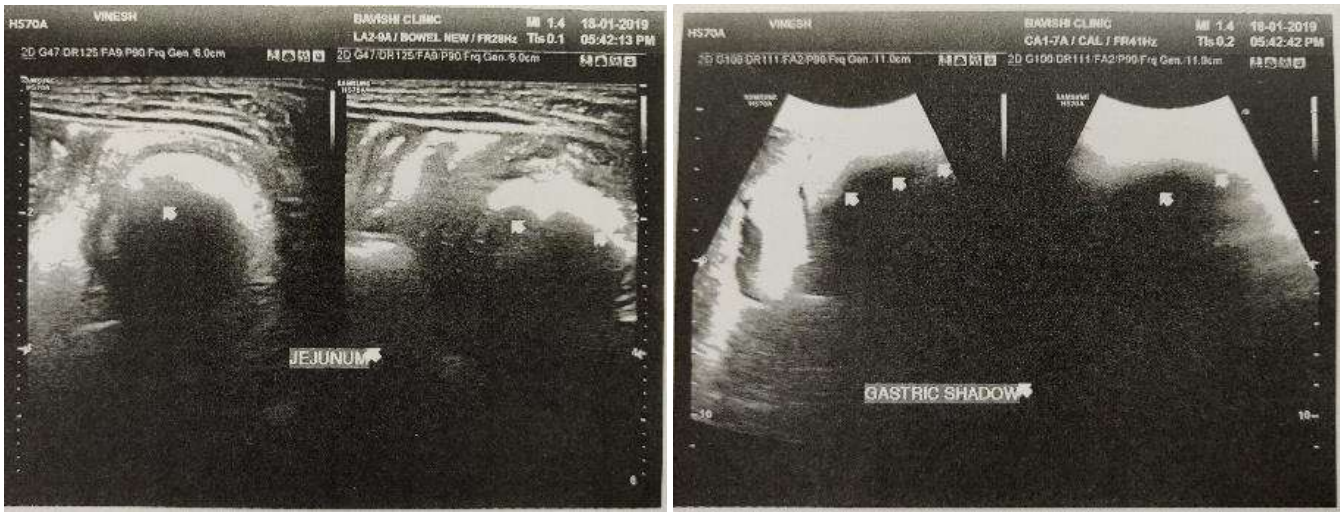

Fig 1: Abdominal Sonography

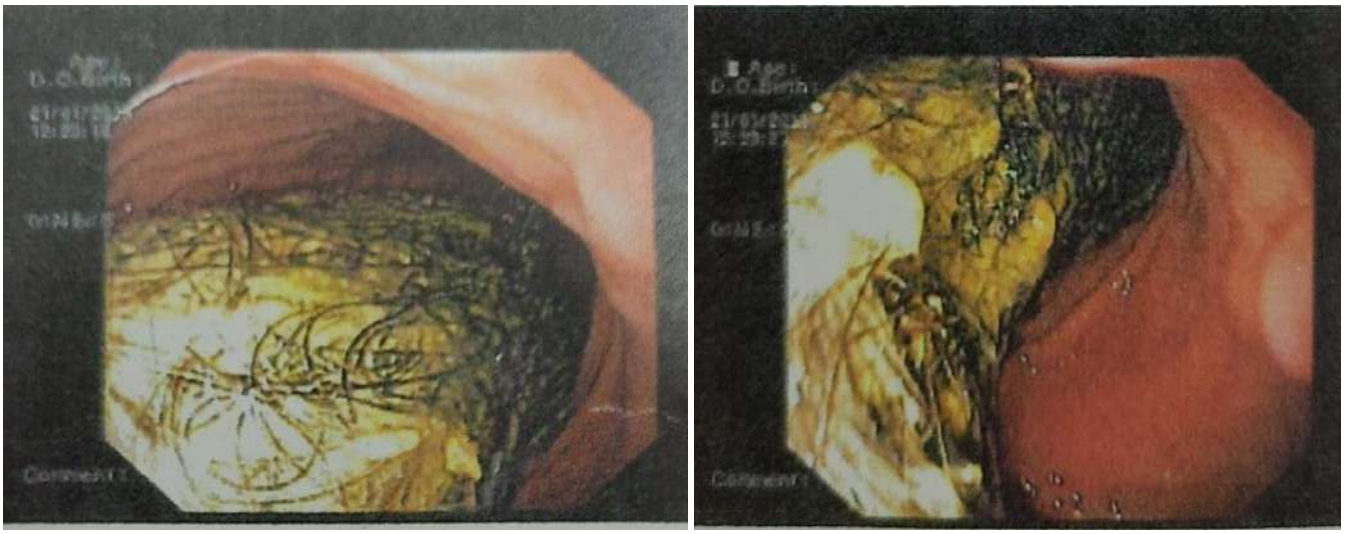

Fig 2: Gastro-Duodenoscopy Report

A nasogastric tube was inserted which drained about 40 ml of gastric secretion over 12 hours.

An Open Laparotomy was performed using supraumbilical midline incision & anterior gastrotomy was done (Figure 3). Mass was delivered through this incision without causing injury to the bowel (Figure 4). It was J-shaped, foul-smelling black bunch of synthetic hairs about 75cm long & 850gm weight (Figure 5). Gastrotomy closure done in two layers followed by abdominal fascia & skin closure.

The postoperative period was good & uneventful. Patient was kept Nil Per Oral (NPO) for four days, receiving i.v fluids & electrolyte supplementation along with broad-spectrum antibiotics. Oral feeds were started gradually in the fifth postoperative day. He was discharged home on seventh postoperative day. Patient was advised psychiatric consultation on discharge.

Fig 3: Delivery of trichobezoar through Anterior Gastrotomy

Fig 4: Delivery of trichobezoar through Anterior Gastrotomy

Discussion

Trichobezoar is a rare condition almost exclusively seen in young females. Trichobezoars constitute about 55% of all bezoars, 90% of cases occur in adolescent girls as mentioned in the classic review of literature [3]. While the patient in this reported case was a boy with history of ingesting hair, carpet and clothing fibres from the ground. Rapunzel syndrome is a rare form of trichobezoar with only forty-nine cases formally reported in the medical literature.

It has been suggested that impairment of the sieving and grinding mechanisms of the stomach, more than the emptying, is the factor contributing to the formation of bezoars [4].

Factors associated with trichophagia and trichobezoar formation in early childhood include child neglect, abuse, or bereavement, mental retardation, psychiatric conditions and underlying behavioural disorder leading to hair pulling (trichotillomania) and pica [5]. Our patient has no trichotillomania, but suffers from trichophagia

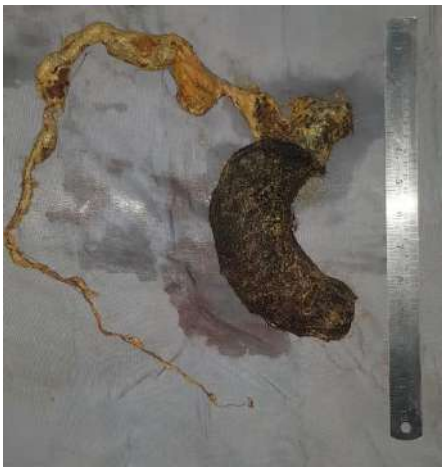

Fig 5: Completely extracted stomach-shaped trichobezoar with is tail extending to small bowel

Trichobezoar has no pathognomonic symptoms or signs and the patient may remain asymptomatic for many years. The most common presenting complaints include bloating, nausea, early satiety, abdominal pain and weight loss. Trichobezoar may present as an emergency with perforation of either the stomach or the intestine due to reduced blood supply to the gastric mucosa and part of the intestine because of the presence of large eroding or obstructing mass of bezoar, which may cause ulceration and later develop into perforation. Less commonly, patients may also present with peritonitis, hematemesis and intussusception [7]. Other complications include obstructive jaundice, acute pancreatitis and gastric emphysema [8].

Bezoars can be diagnosed by ultrasound, contrast films, CT scan or endoscopy.

The ultrasonography has limited role in diagnosis because the high echogenicity of hair and the presence of multiple acoustic interfaces created by trapped air and food limit the ultrasonography of the trichobezoar, it shows a typical curvilinear trichobezoar with bright echogenic band. CT Scan vividly demonstrates trichobezoars as free-floating filling defects within the stomach, especially in the presence of orally administered contrast medium.

A definitive diagnosis of gastric bezoars is established by endoscopy as was done in this case. The proper therapy for any bezoar necessitates removal and prevention of recurrence.

They can be removed by Open Laparotomy, Upper GI Scopy & Laparoscopically. Small bezoars may be amenable to nasogastric lavage or suction, a clear liquid diet and the use of prokinetic agents. Endoscopic retrieval and fragmentation can be used for proximal bezoars however, the procedure can be technically challenging and also during the procedure fragments may migrate distally and cause small bowel obstruction [10]. Specialized Bezotomes & Bezotriptors (medical devices that pulverize bezoars either mechanically or with acoustic waves) also have been used to fragment large & solid trichobezoars.

Conclusion

Laparotomy is still considered the treatment of choice in many cases due to relatively low complication rate, low complexity &

the ability to carefully check the entire gastrointestinal tract for satellites in a relatively short period of time [9]. Successful laparoscopic removal requires significantly longer operation time as compared to conventional laparotomy. The risk of spilling contaminated hair fragments out of bowel lumen into the peritoneal cavity makes the laparoscopic approach even less attractive.

Laparotomy is mandatory for bezoars that have complication of perforation, haemorrhage or are too large to be managed less invasively. Additionally, in Rapunzel syndrome, we see no other valid option than conventional laparotomy. The standard approach is a gastrotomy and then gastric trichobezoars can be easily extracted with careful palpation of the duodenum and jejunum for hair balls that may have broken off from the primary mass as was the case in this patient! During laparotomy, often the bezoar along with its tail can be retrieved through single gastrotomy, but in some cases, multiple enterotomies may be required to avoid the risk of gut perforation because the long trichobezoars are usually adherent with the gut wall and are difficult to be removed through single enterotomy. In this case report, the long bezoar was removed easily via the single gastrotomy without complications.

After removal of trichobezoar, the prognosis is good if psychiatric therapy to control pica and habitual trichophagia is successful. Recurrence of trichobezoar is expected if the underlying emotional stress factor is not resolved [11].

Declaration

Funding: No Funding Sources

Conflict of interest: None

Ethical Approval: The Study was approved by Institutional Ethics Committee

References

1. Williams RS. The fascinating history of bezoars. Med J Aust. 1986; 145:613-614.
2. Sehgal VN, Srivastava G. Trichotillomania ± trichobezoar: revisited. Eur Acad Dermatol Venereol. 2006; 20:911-915.
3. DeBakey M, Ochsner W. Bezoars and concretions: A comprehensive reviewof the literature with an analysis of 303 collected cases and a presentation of8 additional cases. Surgery. 1939; 5:132-160.
4. Coulter R, Antony MT, Bhuta P, Memon MA. Large gastric Trichobezoar Ina normal healthy woman: Case report and review of pertinent literature. Southern Med J. 2005; 98:1042-1044.
5. Gonuguntla V, Joshi DD. Rapunzel syndrome: a comprehensive review of an unusual case of trichobezoar. Clinical medicine and research. 2009; 7:99-102.
6. Carr JR, Sholevar EH, Baron DA. Trichotillomania and trichobezoar: A clinical practice insight with report of illustrative case. J Am Osteopath Assoc. 2006; 106:647-652.
7. Anantha RV, Chadi SA, Merritt N. Trichobezoar causing intussusception: Youngest case of Rapunzel syndrome in a boy in North America. J Ped Surg Case Reports. 2013; 1:e11-e13.
8. Klipfel AA, Kessler E, Schein M. Rapunzel syndrome causing gastric emphysema and small bowel obstruction. Surgery. 2003; 133:120-121.
9. Gorter RR, Kneepkens CM, Mattens EC, Aronson DC, Heij HA. Management of trichobezoar: case report and literature review. Pediatric surgery int. 2010; 26:457-463.
10. Alsafwah S, Alzein M. Small bowel obstruction due to trichobezoar: role of upper endoscopy in diagnosis. Gastrointest Endosc. 2000; 52:784-786.
11. Memon SA, Mandhan P, Qureshi JN, Shairani AJ. Recurrent Rapunzel syndrome-a case report. Medical Science Monitor. 2003; 9:CS92-94.