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Case Report

Rapunzel Syndrome with Concealed Gastric Perforation - A Rare Presentation

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ABSTRACT

An unusual form of bezoars extending from the stomach to the small intestine or beyond has been described as Rapunzel syndrome. 25years female presented with history of mild epigastric abdominal pain and multiple episodes of vomiting since 4 days. Imaging detected large gastric trichobezoar with pneumoperitoneum suggestive of perforation. Patient was vitally stable with no peritoneal signs. She underwent laparotomy for removal of a trichobezoars weighing 700 grams. She was detected to have gastric ulcer with concealed perforation near lesser curve of stomach which did not require specific surgical management. Patient had good recovery.

Key words: Bezoars, Perforation, Pneumoperitoneum, Rapunzel syndrome, Trichobezoars.

CASE REPORT

25years old female presented to surgical out-patient department with 4 days history of mild epigastric abdominal pain and vomiting. She did not have history of hematemesis or malena. She gave history of early satiety and loss of appetite and weight since last 2 month. On examination patient had stable vital parameters. On abdominal palpation she had a firm, large 10x15 centimeter sized lump in the epigastric and left hypochondriac region, oval in shape, freely mobile in craniocaudal direction with restricted mobility in transverse direction. There was no tenderness, guarding or rigidity. We noticed that patient had short uneven stubby hairs with patchy alopecia. On enquiry relatives gave history of trichotillomania (habit of eating hairs). She refused history of any psychiatric consultations for same.

Erect X-ray abdomen was unremarkable. USG raised suspicion of foreign body in stomach probably trichobezoar. Gastroscopy confirmed the diagnosis. CECT scan of abdomen showed gastric trichobeazoar with characteristic entangled appearance occupying almost entire gastric lumen with reactive thickening of the stomach wall with presence of free intraperitoneal air near lesser curvature of stomach spreading in lesser sac reaching upto gastro-esophageal junction suggestive of sealed off perforation(fig.1,2). A discrete 2 cm sized foreign body with similar appearance as trichobezoar was noticed in proximal jejunum on CT scan.

Patient underwent upper midline laparotomy for removal of trichobezoars. Anterior gastrotomy was done to access the bezoar. Trichobezoar was found to occupy entire stomach (fig.3) with its thin Rapunzel tail extending distally in duodenum and reaching till proximal jejunum in continuity with 2 cm discrete bezoar in jejunum as seen on imaging which was removed in toto

and in continuity (fig.5). Trichobezoar and its long Rapunzel tail were removed through gastrotomy itself without needing additional jejunal enterotomy. On removal of the trichobezoars; a 2 cm deep ulcer was visible over posterior surface of gastric antrum near lesser curvature (fig.4). Base of this ulcer was densely adherent posteriorly to the pancreas and there was no obvious perforation seen in the stomach or anywhere else in GI tract. Thus we concluded that peumoperitoneum near lesser curve of stomach was due to concealed perforation at the site of deeply penetrating ulcer caused by large trichobezoar. Ulcer was left alone. Anterior gastrotomy closed in double layer.

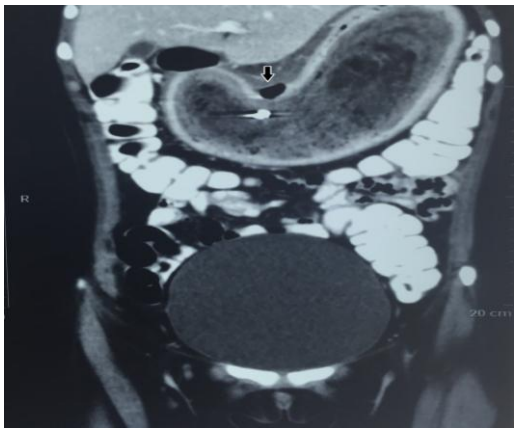

Figure 1: coronal CECT showing gastric trichobezoar With free intraperitoneal air (arrow) near lesser Curve due to concealed perforation

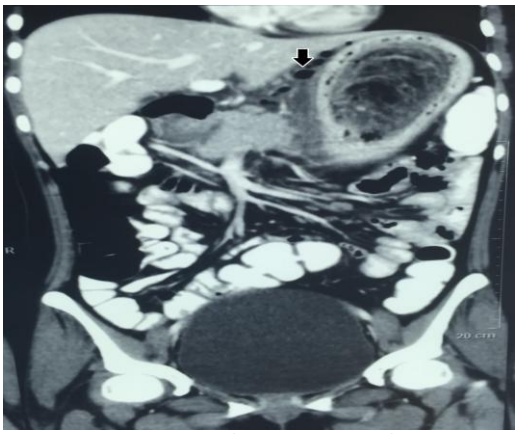

Figure 2: coronal view of CECT showing free intraperitoneal air (arrow) suggestive of perforation hollow viscous

Figure 3: intraoperative photo of gastric trichobezoar being removed by anterior gastrotomy

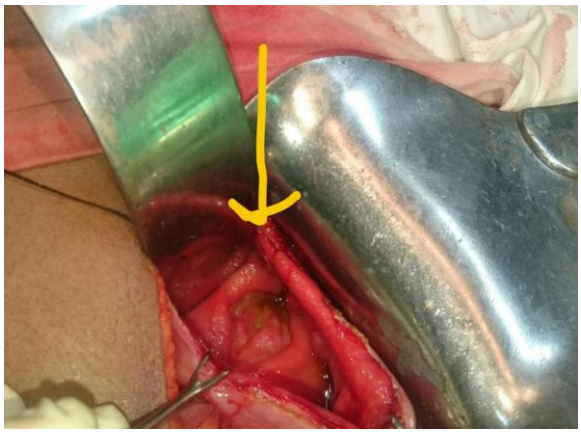

Figure 4: posterior gastric wall deep penetrating ulcer (yellow arrow) seen after removal of trichobezoar

Figure 5: specimen of trichobezoar with Rapunzel tail

Patient started orally after fifth postoperative day and discharged after suture removal on tenth day. Simultaneous psychiatric consultation was done and treatment started to avoid recurrence. She remained symptom-free, appetite improved and gained weight on one month follow up. Follow up gastroscopy showed good healing of ulcer.

DISCUSSION

Bezoars were originally described in reports dated 1000 years BC as found in the stomach of mountain goats in the Western

region of Persia. The term bezoars, as used today, are applied to concretions of various foreign or intrinsic substances found in the stomach and intestines of both humans and animals. Bezoars are of various types, trichobezoars, consisting of hair; phytobezoar, consisting of fibers, seeds and skin of vegetables; trichophytozeoar, a combination of both and concretions composed of ingested shell ; lithobezoar of mud and stones and lactobezoar of milk curd. [(1)] Additionally to the classical trichobezoars (hair bolus) and phytobezoar (concretion of vegetable fibers), bezoars may also present different compositions such as stones, plastic, cotton, medicines (antacids) and resin fibers. [(2)] Out of these trichobezoars are the commonest.

The first case of human trichobezoars was described in 1779, by Baudamant [(3)] and the first surgical removal of a trichobezoars was performed in 1883, by Schonborn. Highest incidence of bezoars present in the women. While 80% of cases occur before the age of 30 years, more than 90% of bezoars occur in adolescent girls with history of trichophagia present in 50% of cases. Trichobezoars are commonly seen in young girls with psychiatric disorders. Trichotillomania and pulling of hairs with trichophagy, and coexistent gastric motility disorders are usually seen in these patients. It has been estimated that only 1% of patients with trichophagia develop a trichobezoar. [(4)]

Trichobezoars symptoms depend on the site and size of the mass, besides the degree of functional involvement of the affected segment. Stomach trichobezoars may cause nonspecific dyspeptic symptoms such as post-prandial abdominal pain, nausea and emesis, halitosis, gastrointestinal bleeding, anemia and malnutrition. [(5)]

The hairs swallowed collect in the stomach to form a hair ball, escaping peristaltic propulsion because of their slippery surface, are retained in the folds of the gastric mucosa. As more hair accumulates, peristalsis causes it to be enmeshed into a ball. The stomach is not able to push these hairs out of its lumen because the friction surface of the bezoars is insufficient for propulsion by peristalsis. The frequent location of these trichobezoars is in the stomach is due to the holdup by the pylorus and the churning action of the stomach which helps to entangle new hair into the already formed cast. Rarely, however these bezoars can get broken and pass into the intestine, extending up to the ileocecal junction, a condition termed as Rapunzel Syndrome. Rapunzel syndrome is a rare clinical scenario that arises when a large stomach trichobezoars mass extends through the duodenum and into the small intestine with or without mechanical intestinal obstruction. It derives its name from the long haired heroine in the fairy tale by the Grimm brothers.

As this ball gets too large to leave the stomach, gastric atony may result. The ball of hair becomes even more matted together and assumes the shape of the stomach, usually as single solid mass. [(6)] The bezoars have a glistening shiny surface due to the mucus that covers it. Decomposition and fermentation of fats give the bezoars, and the patient's breath, a putrid smell. [(7)] The acidic contents of the stomach denature the hair protein and give the bezoars its black color. [(8)] Affected patients occasionally remain asymptomatic for many years. Symptoms develop as the bezoars increases in size to the point of obstruction. Not surprisingly, most of the cases have been reported in countries where women traditionally have long hair. The most common presenting signs are abdominal pain, nausea and vomiting, obstruction, and peritonitis. Less commonly, patients have presented with weight loss, anorexia, haematemesis and intussusceptions. Complications of large eroding or obstructing bezoars additionally include gastric ulceration, obstructive jaundice, acute pancreatitis and gastric emphysema. [(9,10)] Other malabsorption related complications include protein-losing enteropathy, iron deficiency, and megaloblastic anaemia. Less common

presentations and complications include pancreatitis, obstructive jaundice. Appendicitis secondary to trichobezoars has also been reported. [11]

The gold standard for diagnosis is upper gastrointestinal endoscopy. In addition to providing direct visualization, this procedure allows sample taking and potentially therapeutic intervention. The diagnosis can be supported by radiological examination viz Barium swallow ultrasonography or computed tomography which may show a characteristic appearance. Contrast-enhanced radiography frequently demonstrates a non-adherent, spindle-shaped mass in the stomach, with the contrast mean surrounding, coating and infiltrating the trichobezoars, excluding the diagnosis of neoplasia.

The only satisfactory treatment for bezoars is early surgical intervention. Postoperative complications and death though rare is reported in the literature. The treatments of bezoars encompass removal of the mass and prevention of recurrence by addressing the underlying physical or emotional cause. Endoscopic therapy can be effective for bezoars composed of vegetable matter (phytobezoar) and milk curd (lactobezoars), as they are small in size, but is less likely to be effective for trichobezoars, particularly those that are large (>20 cm). Specialized bezotomes and bezotriptors (medical devices that pulverize bezoars either mechanically or with acoustic waves) have been used to fragment large and solid trichobezoars. [12] Dissolution with papain saline, acetylcysteine, and cellulase are generally used for small phytobezoar. The first report of the successful endoscopic removal of a trichobezoars was for one that was relatively small, weighing only 55g. [13] An analysis of the published case reports revealed that out of 40 cases in which endoscopic removal had been tried, only two (5%) were successful. In one of these, a trichobezoars was successfully removed whole from the distal esophagus. [14]

Large trichobezoars require surgical management by means of gastrotomy as the method of choice. Enterotomy may be required when bezoars extend into the small intestine. Traditionally, a gastric trichobezoars was removed by gastrotomy through an upper midline laparotomy. Since the advent of minimally invasive surgery, surgeons now use laparoscopic techniques for small to moderate-size bezoars. [15]

In some centers, laparoscopy is considered inferior to laparotomy for the treatment of a trichobezoars. *Nirasawa et al.* were the first to report on laparoscopic removal of a trichobezoars. [16] Various new techniques have been developed to tackle the issue of bezoars with variable successes like water jet, bedside coca-cola lavage, direct large channel endoscopic aspiration, Dormia basket, forceps, polypectomy snare, Nd: YAG laser therapy, use of modified needle - knife [bezotome] and modified lithotripter [bezotriptor]. [17] Various other methods like extracorporeal shock wave lithotripsy, intragastric administration of enzymes (pancreatic lipase, cellulose), and medications (metoclopramide, acetylcysteine) demonstrate varying success.

Incidence of recurrent trichobezoars is extremely rare. The recurrence of trichobezoars reported in literature occurred 2 or more years following its removal. [18] The effective treatment of trichobezoars lies not only in its surgical removal but also in the cognitive behavior therapy and pharmacotherapy with serotonin reuptake inhibitors.

REFERENCES

1. Lal MM, Dhall JC. Trichobezoar: A collective analysis of 39 cases from India with a case report. Indian Pediatr 1975; 12:351-353.
2. Barbosa AL, Bromberg SH, Amorim FC, et al. Obstrução intestinal por tricobezoar. Relato de caso e revisão da literatura. Rev Bras Coloproct. 1998;18:190-3
3. S Alsafwah, M Alzein. Small bowel obstruction due to trichobezoar: role of upper endoscopy in diagnosis.

Gastrointest Endosc. 2000 Dec; 52(6): 784-6.
4. Phillips MR, Zaheer S, Drugas GT. Gastric trichobezoar: case report and literature review. Mayo Clin Proc 1998; 73: 653–656.
5. Ibuowo AA, Saad A, Okonkwo T. Giant gastric trichobezoar in a young female. Int J Surg. 2006;6:e4-e6
6. Pace AP, Fearne C. Trichobezoar in a 13 year old male: a case report and review of literature, Malta Med J 2003; 15:39–40. 10.
7. Sidhu BS, Singh G, Khanna S: Trichobezoar. J Indian Med Assoc 1993; 91:100–101.
8. Sharma V, Sahi RP, Misra NC. Gastro-intestinal bezoars. J Indian Med Assoc 1991;89:338–339
9. Jiledar Singh G, Mitra SK. Gastric perforation secondary to recurrent trichobezoar. Indian J Pediatr 1996; 63:689–691.
10. Klipfel AA, Kessler E, Schein M. Rapunzel syndrome causing gastric emphysema and small bowel obstruction. Surgery2003; 133:120–121.
11. Kochar AS. Acute appendicitis associated with a trichobezoar. J Am Med Assoc 1984; 252:1681-1682.
12. Wang YG, Seitz U, Li ZL, Soehendra N, Qiao XA. Endoscopic management of huge bezoars. Endoscopy 1998; 30:371–374.
13. Saeed ZA, Ramirez FC, Hepps KS, Dixon WB: A method for the endoscopic retrieval of trichobezoars. Gastrointest Endosc. 1993, 39: 698-700. 10.1016/S0016-5107(93)70226-6.
14. Pogorelić Z, Jurić I, Zitko V, Britvić-Pavlov S, Biocić M: Unusual cause of palpable mass in upper abdomen-giant gastric trichobezoar: report of a case. Acta Chir Belg. 2012, 112: 160-163.
15. Naik S, Gupta V, Naik S, Rangole A, Chaudhary AK, Jain P, Sharma AK. Rapunzel syndrome reviewed and redefined. Dig Surg2007; 24:157–161.
16. Nirasawa Y, Mori T, Ito Y, Tanak H, Seki N, Atomi Y: Laparoscopic removal of a large gastric trichobezoar. J Pediatr Surg. 1998, 33: 663-665. 10.1016/S0022-3468(98)90342-6.
17. Blam ME, Lichtenstein GR. A new endoscopic technique for the removal of gastric phytobezoars. *Gastrointest Endosc.* 2000 Sep; 52(3): 404-8.
18. Kumar V, Khatri AK, Pandey M, et al. Recurrent trichobezoar: First reported case. Indian J Pediatr 1996; 63:257-258.

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