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Case Report

Jejunal perforation secondary to trichobezoar (Rapunzel syndrome): a rare presentation

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ABSTRACT

Trichobezoar is a rare disorder. It usually occurs in young and adolescent females associated with some psychiatric illness. Trichobezoar usually accumulate in the GI tract and most commonly in stomach but it can migrate through the pylorus into the jejunum, ileum and colon. Once the bezoars extends from the stomach into the jejunum or further on, it is referred to as "Rapunzel syndrome". Though initially asymptomatic but over a period of time it may cause gastric mucosal erosion, ulceration, and perforation of the stomach or the small intestine. If unrecognized, tichobezoar may present with intussusceptions, obstructive jaundice, protein-losing enteropathy, pancreatitis and even death. Small trichobezoar may be extracted by endoscopic fragmentation but bezoars like Rapunzel Syndrome, on the other hand, need open surgical removal. Counseling by a psychiatrist is an important part of management to prevent recurrence.

Keywords: Trichobezoar, Trichotillomania, Rapunzel syndrome

INTRODUCTION

It derived from Greek word trich, which means hair and bezoars, which means collections of indigestible material. Swain first described trichobezoar while conducting an autopsy in 1854. Once the bezoars extends from the stomach into the jejunum or further on, it is referred to as "Rapunzel Syndrome," which was first described in 1968 by Vaughan et al.[6] Trichobezoar are often associated with psychiatric illness like trichotillomania and trichophagia. It usually occurs in young and adolescent females. Trichobezoir usually accumulate in the GI tract and most commonly in stomach but it can migrate through the pylorus into the jejunum, ileum or even in the colon.[8] On the basis of their contents, bezoars are classified into phytobezoars (composed of non-digestible food materials such as seeds and pits), trichobezoars (composed of hair), lactobezoars (composed of lactose), and pharmaco-bezoars (composed of medications).[4] Though initially asymptomatic but 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. The most common complications reported over the years, include gastric mucosal erosion, ulceration, and perforation of the stomach or the small intestine.[8] In this case, jejunal perforation is present, which is a very rare presentation of rapunzel syndrome. Even most of the surgeons do not encounter this disease throughout their life. Trichobezoir has to be considered as part of differential diagnosis of abdominal pain in young adolescents girls with history of trichotillomania or trichophagia, or on consistent with hair pulling behavior**.** However, prevalence of the condition is very less 0.06% to 4%.

CASE REPORT

A 35 year-old woman, reported to the emergency department with the complaints of severe pain in abdomen, abdominal distension and obstipation for the

past 4 days. There was history of colicky abdominal pain on and off from last 6 month and no any co morbidity was present as HTN, DM, and TB. Patients have history of mental illness since childhood. No menstrual irregularity was present. On general examination, patient had fever, tachycardia and with low blood pressure. On local examination, there was generalized tenderness in abdomen with guarding and rigidity, bowel sound was absent and on per rectal examination rectum was empty.

Investigation

After initial resuscitation, an erect abdominal radiograph was taken, which revealed gas under right dome of diaphragm (Figure 1). On ultrasonography, moderate amount of fluid was present. On the basis of clinical parameter and radiological investigation, diagnosis of perforation peritonitis was made, and an emergency laparotomy was planned.

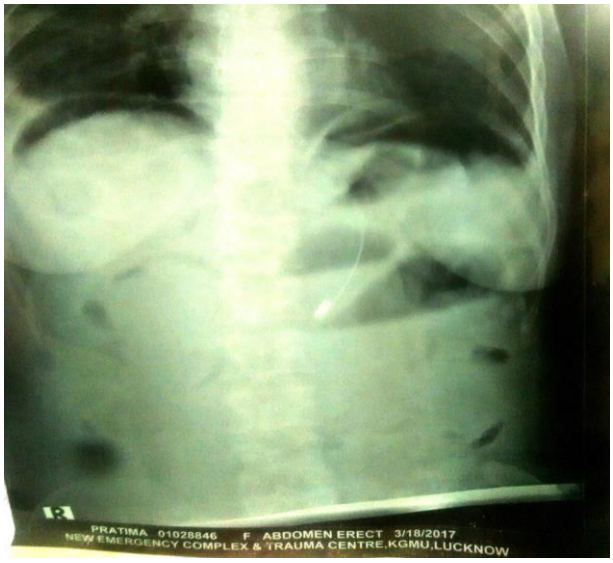

Figure 1: X-ray gas under right dome of diaphragm suggestive of pneumoperitoneum.

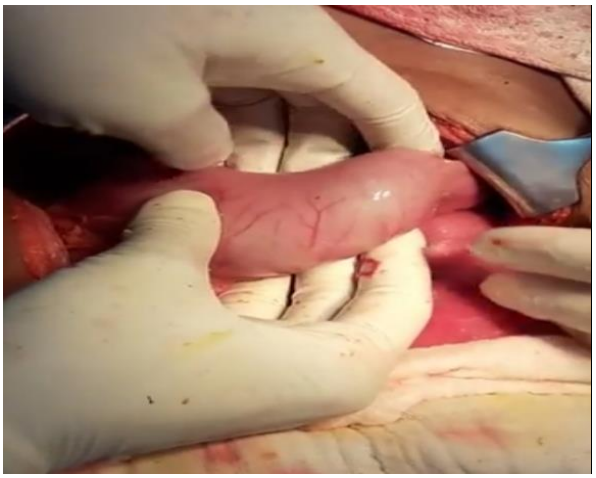

Figure 2: Trichobezoir as a solid mass in stomach.

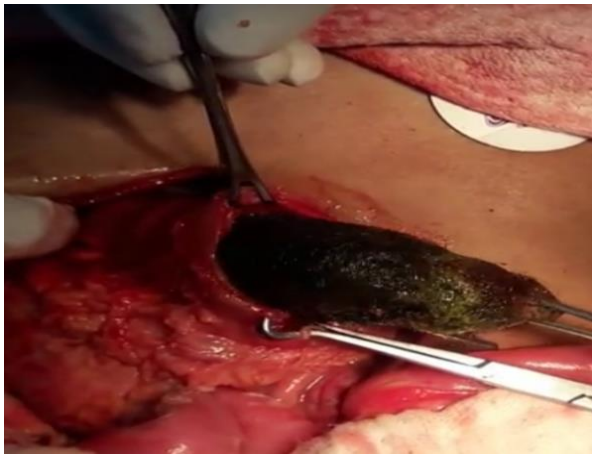

Figure 3: Removal of trichobezoir through gastrostomy.

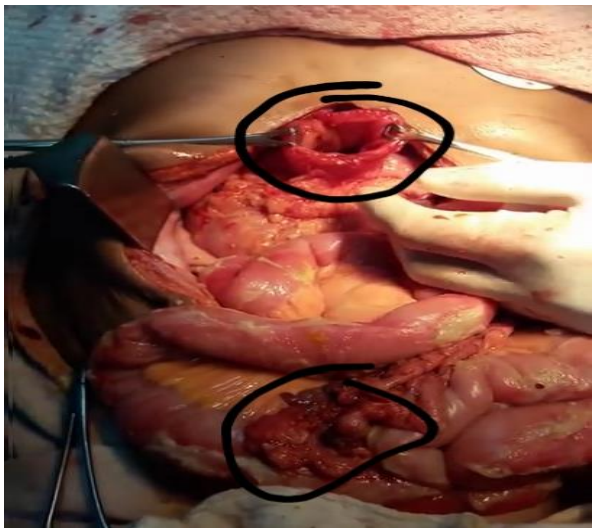

Figure 4: Upper black circle marked as gastrostomy incision, lower black circle marked as a jejunal perforation.

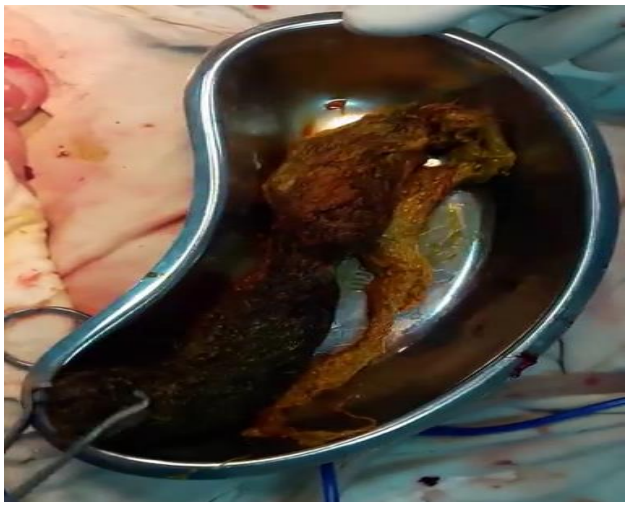

Figure 5: Tuft of hairs about 50 cm in length extending from stomach upto jejunum.

Differential diagnosis

If a female presented in surgical emergency with acute abdomen and signs of perforation peritonitis, a diagnosis of trichobezoar must be considered in differential diagnosis along with other common causes of perforation e.g. duodenal perforation caused by Helicobacter pylori, illeal perforation caused by typhoid or tubercular.

Treatment

On exploratory laparotomy, a large mass was felt in the stomach extending from pylorus up to jejunum (Figure 2). A large perforation about 1 cm with multiple small perforations was identified in the jejunal loop 1 feet distal to dudenojejunal junction (Figure 3). A separate gastrostomy was made in anterior wall of stomach (Figure 4). A long tuft of hairs, which had cast of entire stomach extending from pylorus up to jejunum, was removed (Figure 5). The mass was foul smelling and contained densely bunch of hair. The gastrostomy was repaired primarily in two layers, first layer interrupted via vicryl 2-0 and second layer continuous via silk 2-0. Diseased bowel containing jejunal perforation was resected and jejunojejunal anastomosis was done along with feeding jejunostomy. Three drain were placed in right sub hepatic, left paracolic gutter and in pelvic cavity.

Outcome and follow-up

Postoperative period was uneventful and patient was orally allowed on 3rd postoperative day. On 6th postoperative day, patient developed bile leak from midline. Laparostomy was done immediately. Patient was kept nil orally and total parentral nutrition was started. Bile output monitored per day regularly and amount decreased continuously from 300 to 50 ml per day. Patient again allowed orally on 20th postoperative day and discharge with stable vitals. Now psychiatry treatment was started after consultation. The patient had never diagnosed nor treated for any psychiatric illness previously.

DISCUSSION

Trichobezoar is usually seen in adolescent girls, often with an underlying psychiatric illness. The most common presentation is abdominal pain (37%), nausea and vomiting (33.3%), obstruction (25.9%), and peritonitis (18.3%). Less frequently, patients have presented with weight loss (7.4%), hematemesis, anorexia or intussusceptions.[1] a preoperative diagnosis of trichobezoar may be suggested in a patient presenting with severe halitosis, patchy alopecia, a previous history of trichotillomania and trichophagia. Trichobezoir can cause severe complications, such as gastric mucosal erosion, ulceration and even perforation of the stomach or the small intestine. Other associated complications of are malabsorption related, which include protein-losing enteropathy, iron deficiency anemia, and megaloblastic anemia, pancreatitis, intussusceptions.[7] Epigastric pain was present in every reported case. Other common but inconsistent symptoms were guarding, rigidity, rebound tenderness, vomiting (bilious and nonbilious), fever, hypotension, tachycardia. Poor appetite, weight loss and anemia were common but not ubiquitous. Perforation is a most common complication of trichobezoars of either the stomach or the intestine.

The exact pathophysiology explaining why patients with trichobezoars perforate their stomach is unknown, but pressure necrosis and irritation of the gastric mucosa have been implicated. As the size of the trichobezoar increases, the blood supply to the mucosa of the stomach and part of the intestine is hampered, leading to ulceration and eventually perforation. Anemia, hypoalbunemia and hypovolemia may also increase the risk of perforation in case of long standing trichobezoir. The evolution of gastric trichobezoir is still not fully understood. Hair strands because of their slippery surface, escape peristaltic propulsion and are retained in the folds of the gastric mucosa. The hairball lies dormant and the trichobezoar continues to grow in size. Ultimately, the gastric trichobezoir attains the shape of the stomach, usually as a single solid mass.[7]

A chest or abdominal X-ray was the first imaging modality used. Ultrasound and computed tomography were useful in better visualizing a gastric mass, abdominal free fluid, with gastric perforation or perigastric abscess formation detected via extravasations of oral contrast. However upper gastrointestinal endoscopy is considered to be the gold standard for the diagnosis of the trichobezoar and also able to differentiate the nature of trichobezoir.[2]

Various treatment modality are available that include endoscopic, laparoscopic or laparotomy. Nirasawa et al were the first to report on laparoscopic removal of a trichobezoir.[3] In this case, we opted for laparotomy as we did not know what we are dealing with. Moreover, there was sign suggestive of perforation. Though laparoscopy is superior to laparotomy in view of cosmetic results, less postoperative complication and less hospital stay, but in the laparotomy careful examination of the entire gastrointestinal tract are easier to prevent secondary intestinal obstruction due to migration of residual bezoars. Laparotomy is still considered as the gold standard treatment and hence most commonly used. Psychiatric consultation is must to prevent recurrence of the disease.[5]

Learning points

Trichobezoar (Rapunzel syndrome) may be presented as a case of perforation peritonitis. Jejunal perforation is a very rare presentation of trichobezoar (rapunzel syndrome). Open surgery is still preferred mode of treatment.

CONCLUSION

Jejunal perforation is a very rare complication of rapunzel syndrome. Even most of the surgeons do not encounter this disease throughout their life. Trichobezoir has to be considered as part of differential diagnosis of abdominal pain in young adolescents girls with history of trichotillomania or trichophagia, or on consistent with hair pulling behavior. Once diagnosed, often incidentally with the help of radiologic imaging, the literature to date argues for an open approach to removal of the hair mass as opposed endoscopic or laparoscopic approaches. Small trichobezoars may be extracted by endoscopic fragmentation but Bezoars like Rapunzel syndrome, on the other hand, need open surgical removal. Surgical outcomes are excellent for this entity. Many of these patients have psychiatric pathology with emotional problems, family discord, and history of neglect or mental retardation. Counseling by a psychiatrist is an important part of management to prevent recurrence.

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