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Case Report

A RARE CAUSE OF GASTRIC PERFORATION IN CHILDHOOD – RAPUNZEL SYNDROME

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ABSTRACT
Trichobezoar is a rare pathology in which swallowed hairs heap together in the stomach. Rapunzel syndrome is an unusual form of bezoars extending from the stomach to the small intestine or beyond. We report a case with Rapunzel syndrome causing acute life threatening gastric perforation and successful laparotomy removal of a giant gastro-duodenal trichobezoar in a 6-years old girl with a history of trichotillophagia.

Keywords: *Trichobezoar, Trichophagy, Rapunzel Syndrome, Trichotillomania*

INTRODUCTION
A bezoar is an indigestible accumulation of foreign materials in the gastrointestinal tract. Trichobezoars are rare conditions almost exclusively seen in young females (Iwamuro *et al.*, 2015) associated with psychiatric disorders such as trichotillomania and trichophagia. The Rapunzel syndrome is a rare type of trichobezoar that extends into the small intestine, in addition; incidentally, parts of the tail can break off and migrate to the small intestine, causing intestinal obstruction (Ahmad *et al.*, 2016). The main cause of trichobezoar formation is the resistance of human hair to peristalsis and digestion. Therefore, it accumulates between the mucosal folds of the stomach. The continuous ingestion of hair together with mucus and food particles increases its volume.
The most common presenting symptoms of trichobezoar include a palpable abdominal mass, vomiting and noticeable hair loss, abdominal pain, abdominal distension and vomiting. Untreated cases may result in serious complications like gastric erosion and ulceration, gastric and small bowel perforation, gastric outlet obstruction, pancreatitis, obstructive jaundice, protein-losing enteropathy, intussusception and death (Bouwer and Stein, 1998).

CASES
A 6 year old female child was presented to us in the emergency department in shock with complaints of severe pain abdomen, abdominal distension, absolute constipation, fever and recurrent vomiting from the last four days.
On abdominal examination there were gross distention, severe guarding, rigidity and tympanic note on percussion. Clinical diagnosis of bowel perforation with peritonitis was done. Patient was resuscitated with IV fluids and diagnosis of intestinal perforation was confirmed by abdominal x-ray having gas under the diaphragm.
After resuscitation patient was assigned for emergency laparotomy. On exploration, almost 750 ml of purulent fluid came out. Perforation site was searched meticulously. On the anterior wall of stomach one small perforation and few impending perforations were discovered. There was a hard lump inside the stomach.
Incision on the anterior stomach wall extending into the perforation site revealed a large trichobezoar having shape of stomach with extension into duodenum and jejunum. It was delivered through the anterior gastrotomy wound.
Anterior gastrotomy was closed after refreshing margins of perforation. Thorough peritoneal lavage done and the abdomen was closed. Retrospectively parents confirmed history of compulsive hair pulling and hair eating.

Patient condition gradually improved after surgery. There was not any post-operative complication. Patient was discharged in stable condition with psychiatric counselling and treatment.

Figure: *Trichobezoars*

DISCUSSION

Bezoars are incrustation of foreign material in the gastrointestinal tract, mainly in the stomach. Although the stomach is the most common location, bezoars have also been found in the duodenum, jejunum, ileum, colon, appendix and Meckel's diverticulum (Meier and Furtwaengler, 2015). Up to 90% of the all trichobezoars occur in girls. Males are rarely affected (Lynch *et al.,* 2003). Rapunzel syndrome is a rare form of trichobezoar, with a long tail, which may extend up to the jejunum or beyond. Trichobezoars can become large and form as a cast of the stomach. Trichobezoars result from compulsive pulling out of hair and then swallowing the hair. Patients frequently have accompanying comorbid mood, anxiety disorders and psychiatric problems. It has been estimated that only 1% of patients with trichophagia develop a trichobezoar (Sidhu *et al.,* 1993).

The reason why hair is collected in the stomach is not fully understood. Owing to its indigestibility and resiliency, it get hold up by the pylorus, and the churning action of the stomach which helps entangle new hair into the already formed casts.

Clinical manifestations vary, being dependent on the location and size of the trichobezoar, from asymptomatic patients to acute abdomen. A bezoar may also lead to mechanical obstruction, gastric perforation, gastrointestinal bleeding. Perforation and peritonitis are to a great extent responsible for an associated mortality of about 28% (Naik *et al.,* 2007).

Diagnosis is presented by characteristic history of trichophagia and palpable epigastric lump. Clinical suspicion should be high for trichobezoar in women with psychiatric problems presenting with abdominal pain. Diagnosis is confirmed by ultrasonography, upper GI contrast study, CT scan of abdomen and upper gastrointestinal endoscopy (Belsky *et al.,* 2014).

Management and treatment of bezoar involves removal of the mass and prevention of recurrence by addressing the underlying physical or emotional cause. Trichobezoars do not respond to enzymatic dissolution. Basing on its consistency, size and location, bezoar removal may occur via endoscopy or surgery (Caiazzo *et al.,* 2016). The patient's long-term prognosis is excellent if behavioural therapy is used to control trichophagia, and psychological/psychiatric follow up is maintained.

Conclusion

Trichobezoar should be considered in adolescent females presenting with non-specific abdominal complaints. Endoscopy can be invoked as a diagnostic modality for these patients, which can be removed safely with endoscopy. Conventional laparotomy is the treatment of choice in children with trichobezoar and to be the only valid treatment for children with Rapunzel syndrome. The literature provides no

evidence of superiority of endoscopy or laparoscopy. In addition to the acute surgical treatment, psychiatric consultation is needed in order to prevent relapses.

Conflict of Interest*:* None
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