

Case Report

A giant trichobezoar with Rapunzel syndrome: a case report

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ABSTRACT

Bezoar is a conglomeration of foreign material in the intestinal tract. Trichobezoar is accumulation of hairs in the gastric chamber and Rapunzel Syndrome is accumulation of hairs in the small intestine. This commonly occurs in children and adolescents who are commonly suffering from a psychiatric disorder called Trichotillomania in which they have the habit of eating hairs which is called Trichophagia. Authors are presenting a case of a fourteen years old girl who kept on eating her hairs and nails to develop a giant Trichobezoar with a short segment of Rapunzel Syndrome.

Keywords: Rapunzel syndrome, Trichobezoar, Trichotillomania

INTRODUCTION

Bezoars are concretions of foreign and intrinsic material in the gastrointestinal tract. The term is thought to be derived from the Arabic word Badzehr or the Persian word Padzhar which means antidote.¹ Bezoars can be classified into six types: Phytobezoars, Trichobezoars, Lithobezoars, Pharmacobezoars, Plasticobezoars and Lactobezoars.²

Trichobezoars are commonest type of Bezoars comprise 55% of all. Trichobezoar consists of hairballs or hair like fibres caused by chewing and swallowing them. It is found in the stomach and if the Trichobezoar passes through the pylorus into the duodenum, jejunum, ileum and even into the colon in a condition called Rapunzel Syndrome.^{3,4}

Rapunzel Syndrome named after a fairy tale written in 1812 by the Brother Grimm about a young maiden, Rapunzel, with long tresses who lowered her hairs to the ground from the prison tower to permit her young prince to climb up to her window and rescue her.⁵ Trichobezoars

commonly occur in patients with psychiatric disturbances mainly Trichotillomania, but only 50% will have history of Trichophagia.⁶ Hair strands are retained in the folds of the gastric mucosa, peristalsis causes them to become enmeshed until a ball is formed that is too large to leave the stomach, resulting in gastric atony due to the large size of the Trichobezoar.

The hairs become matted together and assume the shape of stomach, usually as a single mass, very rarely they might invade through the small intestine and leads to intestinal obstruction which is known as Rapunzel Syndrome.

CASE REPORT

Authors report a case of fourteen years old girl who walked in the outdoor clinic with complain of pain epigastrium, vomiting, constipation, lump, abdomen and decreased appetite since last two months. On taking history of her illness she revealed that inspired by her friend she started cutting and eating her own hairs and nails till she could eat no more hairs and nails or food.

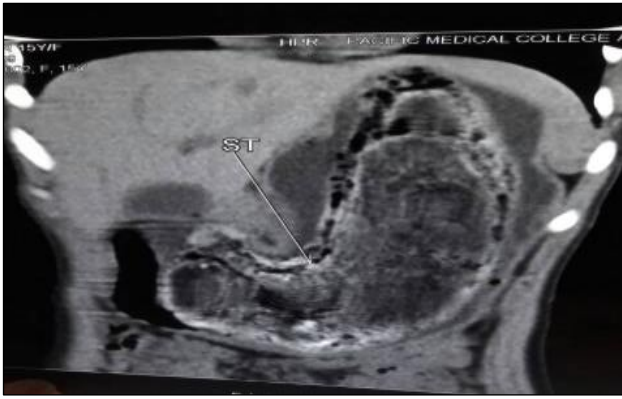


Figure 1: CECT abdomen showing Trichobezoar.

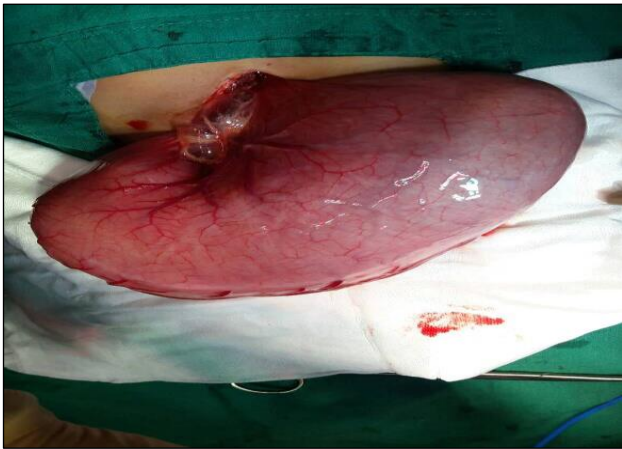


Figure 2: Stomach with lump.

On clinical examination of the girl she was anaemic, emaciated having a palpable lump in epigastrium which was smooth, firm, moving with respiration. Ultrasound and CECT abdomen showed a large Trichobezoar extending into duodenum (Figure 1). She underwent Laparotomy and anterior Gastrostomy was done to reveal a huge Trichobezoar (Figure 2), a giant hairball occupying the whole gastric cavity and extending into the pylorus and duodenum. There was no evidence of gastric necrosis or intraperitoneal infection.



Figure 3: Extracted specimen of Trichobezoar.

The Trichobezoar was removed intact (Figure 3) and abdominal cavity closed after two layered closure of gastrostomy. The patient had an uneventful post-operative course and was discharged after seven days. She is constantly being followed up in the outdoors by surgical and psychiatric doctors counselling her on regular basis.

DISCUSSION

Trichotillomania is characterised by recurrent and impulsive pulling of one's hair for pleasure, gratification, or relief of tension. Children and women less than 30 years of age are the ones who mostly suffer from this type of psycho disorder.

Bezoars are concretions in the gastrointestinal tract increase in size by continuous accumulation of non-absorbable food or fiber. Most Bezoars in common are Trichobezoars from swallowed hairs from the head, dolls or brushes. Trichobezoars have been described in literature and they comprise 55% of all Bezoars.⁷ Only 50% will have history of Trichophagia. As the proteins of the follicles denature, they discolour, turning black or brown and an unpleasant odour seems to be emitted.⁸

Patients with Trichotillomania around 30% will engage in Trichophagia, and of this only 1% will go on to eat their hair to the extent requiring surgical removal. In present case the girl inspired by her friend started eating her own hairs and nails to the extent when she could eat no more because the stomach was occupied full of huge giant Trichobezoar with extension into the duodenum.

Patients may present with abdominal pain (37%), nausea and vomiting (33.3%), abdominal distension, obstruction (25.9%), peritonitis (18.3%), diarrhoea or constipation, anorexia, weight loss (7.4%), abdominal mass, hematemesis and intussusceptions (7.4%).⁹ Complications of Trichophagia include gastrointestinal obstruction (26%), bleeding (10%), perforation, malabsorption and nutritional deficiencies. If Bezoars are left without treatment, the mortality rate can reach 30% because of the associated complications.¹⁰

The standard treatment remains surgical intervention for large, symptomatic and complicated Trichobezoar like Rapunzel syndrome. Endoscopic removal is an option for small Trichobezoar. New reports on the use of video laparoscopy are encouraging.

Enzymatic dissolution, Nd-Yag laser and extracorporeal shock wave fragmentation are novel approaches to be further evaluated.¹¹ Fragmentation carries the risk of small bowel obstruction, protein losing enteropathy, pancreatitis, steatorrhea and perforation. Antidepressant treatment help control the obsession while behavioural therapy and family involvement provides the means for habit-reversal training.

CONCLUSION

Trichobezoar although being a rare disorder is now being commonly diagnosed, thanks to the latest diagnostic gadgets but a big thanks to few of these patients who disclose in their history of food habits they have perverted to. Authors know an entity called Pica which is a common feature of anaemia. It is still under study whether trichotillomania with trichophagia is a form of Pica. Early diagnosis and treatment of Trichobezoar is of eminent importance in order to avoid later fatal complications such as gastric perforation, intestinal obstruction and necrosis. Surgical removal of Trichobezoar, psychiatric consultation, coupled with long term follow up are essential to prevent recurrence, above all the nutritional status of the patient has to be given the utmost importance.

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