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## CASE REPORT

# Delayed Recognition of Gastric Perforation in a Case of Rapunzel Syndrome

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### ABSTRACT

A 15 year old girl with a previous history of psychiatric symptoms and trichophagia (denied any trichotillomania), presented to the surgical emergency department with severe epigastric pain. She was initially managed with masterly inactivity as no indication for exploration was found. Later on investigations revealed gastric mass and ill-defined gastric perforation. Exploratory laparotomy revealed a perforation in anterior wall of stomach which was extended and a huge trichobezoar was found extending upto jejunum. Bezoar was removed and perforation repaired in two layers and covered by omentum. Postoperative recovery was uneventful. She was discharged on 10<sup>th</sup> postoperative day and referred to psychiatry department for further management.

**Key words:** Rapunzel syndrome, gastric perforation, delayed recognition

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### INTRODUCTION

Rapunzel syndrome is a rare presentation of a trichobezoar with only 27 cases having been reported in the literature so far. Amongst its various complications, gastric perforation is the one least reported. We searched the "MEDLINE" database for the case reports limited to human studies, including both male and female patients, aged 6-18 years, published in English literature, with the keywords "RAPUNZEL SYNDROME" and "GASTRIC PERFORATION". Surprisingly, we came across only one case in Jan 1996<sup>[1]</sup>. We herein report an interesting case of Rapunzel syndrome complicated by gastric perforation. We also review the literature for the incidence of Rapunzel syndrome, its various clinical and radiological presentations, the probable mechanisms, complications, prognostic factors and the management strategies.

### CASE REPORT

A 14 year old girl presented to the surgical emergency department with 2 weeks history of severe epigastric pain, radiating to whole abdomen, vomiting, anorexia, abdominal distension and off and on constipation. On physical examination, she was irritable, pale, ill-looking, with a palpable mass in left hypochondrium and epigastrium extending upto umbilicus, moving with respiration.

On USG, an echogenic mass filling part of epigastric and upper paraumbilical area was seen.

CT scan revealed retained food particles in the form of a ball in stomach, also extending into 1<sup>st</sup> and 2<sup>nd</sup> part of duodenum, along with multiple air fluid levels in the central abdominal loops (Fig 1-3). Also there was an ill-defined perforation from posterior wall of stomach communicating with peritoneal cavity due to which she was explored by a midline umbilicus saving incision to reveal a 3\*4cm sized perforation in anterior wall of stomach and a huge mass comprising of hair in the stomach (Fig 4) extending upto jejunum, weighing around 1550g and measuring 20\*10\*9cm with a tail of about 36cm (Fig 5). There was 200ml of pus in right subphrenic and subhepatic space. Mass was extracted and perforation repaired in two layers. Omentum placed over the repair.

Post-operatively, she gave history of trichophagia for last 4 yrs or so. She was a neglected child as her mother died when she was 4, she had a sister who got married when she was 10, and other family members were busy in their own lives. There was also history of other psychiatric symptoms including depressed mood, irritability, and use of abusive language, driving guests and family out of home, beating elder brothers, disturbed sleep and appetite.

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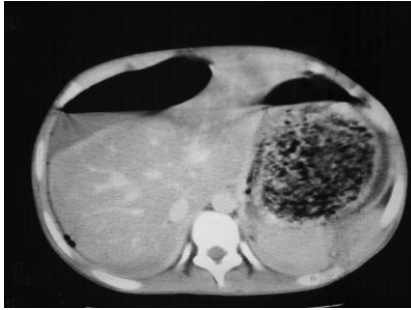


Fig 1: CT scan showing matted hair ball with intervening air in stomach

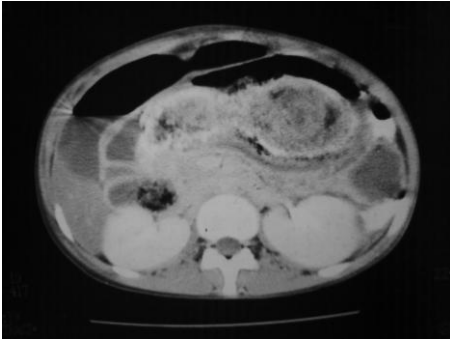


Fig 2: CT scan showing pylorus and 2<sup>nd</sup> part of duodenum with trichobezoar along with enmeshed air pockets

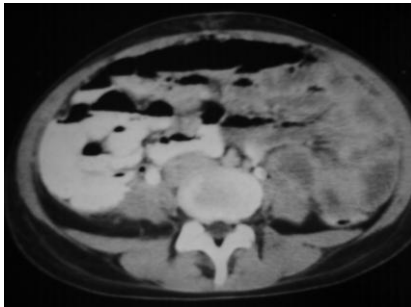


Fig 3: CT scan showing multiple air fluid levels



Fig 4: Peroperative photograph of bezoar



Fig 5: Complete trichobezoar with tail.



Fig 6: other view of the bezoar

## DISCUSSION

The term “trichobezoar” is a combination of “trich” meaning hair in Greek, and “bezoar” meaning poison antidote in Arabic or Persian<sup>2,3</sup>. Trichobezoar is a rare disorder in which swallowed, hair accumulate in the stomach, thus forming a hair-ball over the years.

Rapunzel syndrome is a rare form of trichobezoar. It's named after a tale written in 1812 by the Brothers Grimm about a young maiden named Rapunzel, with long tresses<sup>4,5</sup>. Trichobezoar was first reported in 1779 by Baudamant<sup>2,6</sup> and Rapunzel syndrome was first described by Vaughan and colleagues in 1968<sup>2,7</sup>. Various criteria have been used by different clinicians to report their cases as rapunzel syndrome. Some have defined it as a gastric trichobezoar with a tail extending upto ileocaecal junction, others have said it's simply a trichobezoar with a long tail which may extend to the jejunum, ileum or the ileocaecal junction and still others have defined it as a trichobezoar of any size

presenting in the form of intestinal obstruction. The common features in all these cases are:

1. A trichobezoar with a tail
2. Extension of the tail at least to the jejunum
3. Symptoms suggestive of obstruction<sup>4</sup>.

More than 90% of those afflicted are females<sup>3,7</sup>, and over 80% are under 30 years of age<sup>7</sup>. So far the syndrome has been reported in 23 females and 1 male patient with a mean age of 10.8 years<sup>4</sup>. Overall mortality is about 30%<sup>2</sup> mainly due to perforation and peritonitis. Approximately 10% patients show psychiatric abnormalities<sup>3</sup>.

The reason why hair collect in the stomach is not fully understood, DeBakey and Ochsner suggested that entrapment in the gastric folds is the initiating event. Formation of trichobezoar occurs when the hair strands are retained in the folds of gastric mucosa because the slippery surface prevents propulsion by peristalsis. As more hair is added, peristalsis causes to be enmeshed until a ball, too large to leave the stomach, forms causing gastric atony due to its large size. This large quantity of hair becomes matted with together and assumes the shape of stomach, usually as a single mass. The mucous covering the bezoar gives it a glistening shiny surface. Decomposition and fermentation of fats in the interstices gives it a putrid smell. The acidic contents of the stomach denature the hair protein giving it its black colour regardless of the original colour of hair<sup>4</sup>.

The symptoms<sup>2,4,8,9</sup> of Rapunzel syndrome include abdominal pain (37%), fullness, nausea and vomiting (33.3%), constipation, weight loss (7.8%), anorexia & hematemesis (7.4%) and early satiety. Signs<sup>2,5,8-10</sup> are abdominal mass, tenderness (peritonitis), severe halitosis and patchy baldness.

Bezoars are associated with several complications<sup>4,5,9</sup>, intestinal obstruction (25.9%), peritonitis (18.3%), ulceration, gastrointestinal bleeding (6%), enteropathy, steatorrhea, pancreatitis, intussusception, appendicitis and obstructive jaundice. Free perforation of stomach or small intestine resulting from pressure necrosis of intestinal wall is an unusual complication<sup>9</sup>.

Diagnostic modalities include abdominal plain films, contrast upper GI radiography, ultrasound, CT scan, upper GI endoscopy and laparoscopy. Endoscopy and laparoscopy have also got therapeutic potential<sup>2,8</sup>. CT scan has a higher accuracy rate. Typical bezoar images on CT demonstrate a well-circumscribed intraluminal mass, composed of concentric whorls of densities with

pockets of air enmeshed within it (Fig. 3). Endoscopy can differentiate a trichobezoar from a phytobezoar.

Promising minimally invasive treatment options are extracorporeal lithotripter, endoscopic lithotripter and laser fragmentation. Laparoscopic extraction<sup>2</sup> laparoscopic assisted endoscopic removal<sup>2</sup> may be superior in terms of cosmesis, risk of wound infection and anaesthesia time, their role and success rate needs to be defined. Open surgery remains the gold standard<sup>2</sup> by gastrotomy & enterotomies (single/multiple-when required) or resection of bowel if unable<sup>3</sup>.

An operative mortality of 10.4% is reported by DeBakey and Ochsner<sup>3</sup> whereas non-operative treatment of symptomatic trichobezoar has been associated with a 70% mortality rate<sup>7</sup>. So far, the recommended treatment for a large/complicated trichobezoar is surgical<sup>5</sup>. A psychiatric assessment & long term follow up by endoscopy or contrast study are advocated with parental/spouse counselling as a regular part of treatment to prevent recurrence<sup>4</sup>.

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