A BIZARRE TRICHOBEZOAR: A mystical charm

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ABSTRACT

We report a 5 year-old girl who presented with generalized body swelling, skin rashes, loose motions and severe vomiting worsening since 15 days. An initial diagnosis of Tuberculoses (TB) was made based on clinical and radiological findings. She was discharged on anti-tuberculosis medications once her symptoms improved. At 2 week follow-up she had gained weight however vomiting had not decreased. For further evaluation barium follow through examination was performed which showed multiple intraluminal filling defects nearly extending her entire small bowel. An initial diagnosis of bezoar was made which was confirmed during exploratory laprotomy, where a large trichobezoar extending from the stomach to 50 cm proximal to the ileocaecal junction was removed.

KEY WORDS: Rapunzel Syndrome, Bezoar, Trichotillomania, Trichobezoars

Introduction

Bezoars are collections of indigestible material in the gastrointestinal tract. They are of many forms most commonly occurring are phytobezoars which consist of plant and fruit material and Trichobezoars which occur from ingestion of hair. Patients with Trichobezoars are associated with trichotillomania, in which patients have an irresistible desire to pull out their hair to the point of alopecia. Rapunzel syndrome is a very rare complication of trichobezoar formation in which the mass of hair extends through the pylorus into the small bowel and can even reach the colon. Originally it was named by Vaughan in 1968 that derived its name from the long haired tower bound character in Grimm’s Fairy Tales. There are only 28 recorded cases in the literature worldwide.

Case Report

We present a case of a 5 year old Asian girl who presented with 15 days history of loose motions and vomiting. Due to severe vomiting and weight loss she had turned pale and appeared severely malnourished with anasarca, having weight less than 5th percentile and height less than 5th percentile. She also suffered from generalized body swelling and skin rashes since 1 week. Skin rashes were scaly erythematous with areas of hyperpigmentation mainly involving extremities and anterior abdominal wall. Her mother mentioned that she has had vomiting on and off for about a year and has had several hospital visits and admissions in the past, the last one 5 months back due to similar symptoms.

Chest radiological examination and ultrasound abdominal examination was unremarkable. Barium follow through examination was considered inadequate due to significant food particles and poor preparation did show some distal ileal loop thickening, however no obstruction or other pathology was seen. On the basis of history, clinical examination and radiological examination Tuberculosis (TB) was considered on top of the differential diagnosis, others being severe protein calorie malnutrition with either inflammatory bowel disease or malabsorption. Patient was started on anti TB medications and discharged when she showed signs of improvement. Early follow-up in 2 weeks showed some improvement in weight, but continued to complain of relentless vomiting. To reinvestigate barium follow through examination was performed with better preparation which showed multiple intraluminal filling defects extending from the duodenum to the ileum (Fig 1 and 2).
Discussion

Trichobezoars are commonly present in young females usually with an underlying psychiatric disorder. Formation of trichobezoars occurs when the hair strands are retained in the folds of the gastric mucosa because their slippery surface does not move forward with peristalsis. As more hair is added, peristalsis causes it to be enmeshed into a ball, too large to leave the stomach, causing gastric atony due to its large size. This large quantity of hair becomes matted together and assumes the shape of the stomach, usually as a single mass.

Rapunzel syndrome is a rare form of trichobezoar. It is named after a charming tale written in 1812 by the Brothers Grimm about a young maiden, Rapunzel, with long tresses who lowered her hair to the ground from the high tower to permit her young prince to climb up to her window and rescue her. This syndrome was originally described by Vaughan in 1968. Since then...
28 cases have been reported in the literature, with variable clinical features. 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 up to the ileocecal junction; others have said it is simply a trichobezoar with a long tail, which may extend to the jejunum, ileum or the ileocecal junction, and still others have defined it as a trichobezoar of any size which presents in the form of an intestinal obstruction.

We concluded from a review of several articles that the most important requirements to qualify a trichobezoar to be a Rapunzel Syndrome are 1) a tail; 2) extension of the tail at least to the jejunum; 3) symptoms suggestive of obstruction.

All the cases reported in the literature are females, except one. This may be attributed to the traditional long hair of females which makes them more prone to entanglement and hence formation of trichobezoar casts. The only male was reported by Hirugade et al. This patient used to eat hair of his sisters.

**Conclusion**

Rapunzel syndrome is an uncommon trichobezoar, with a tail extending into the small intestine. It has a varied presentation and is seldom diagnosed preoperatively.

**References**


