Rapunzel Syndrome

RAPUNZEL SYNDROME; A RARE CAUSE OF UPPER GASTRO INTESTINAL OBSTRUCTION

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ABSTRACT

Rapunzel syndrome is a rare form of trichobezoar that extends into small intestine and beyond. This condition usually occurs in mentally retarded or emotionally disturbed young females. Case of a young female is presented.

Keywords: Trichobezoar, Rapunzel syndrome, Intestinal obstruction, Trichophagia

INTRODUCTION

Bezoars are concretions in the gastrointestinal tract that increase in size by continuous accumulation of non-digestible foods or fibres. Trichobezoars are collections of swallowed hair in the stomach, predominantly found in emotionally disturbed or mentally retarded females. Rapunzel Syndrome is a rare, unusual form of trichobezoar extending from the stomach into the small intestine or beyond. We present here a case of Rapunzel syndrome presenting as recurrent pyloric obstruction in a young female.

CASE REPORT

A 15 year old girl was admitted in surgical department Military Hospital Rawalpindi with recurrent attack of colicky central abdominal pain for last 4 years. Each episode lasted from a few hours to a day or so and was relieved by vomiting which often contained hair and threads. Her latest episode lasted for four days. Her bowel habits and appetite were normal. She was otherwise in good health with no pallor. The abdominal examination revealed a hard, non-tender mobile elliptical mass in epigastrium. Complete blood count and metabolic work up were normal.

Upper GI endoscopy revealed a mass of entangled hair and threads in stomach, blocking access to duodenum. Further questioning from mother revealed that the patient had been in the habit of pulling and chewing hair of her dolls and from combs since 4 years of age. She also ate threads from clothes and pieces of paper but did not pull out her own hair. She was dull in her class and had been failing in 5th grade for last two years. Her mother was counselled regarding surgery.

Upper midline laparotomy was made and stomach was opened to extract the trichobezoar. While pulling out the mass of entangled hair and threads with a serpentine thick tail (a cast of pylorus, duodenum and jejunum) small intestine was observed to concertina towards the duodenojejunal junction (figure-1). The trichobezoar was found to be extending up to mid-jejunum and was removed in two pieces by making an enterotomy. The total length of the trichobezoar was 170 cm (figure-2). Post operatively patient made a good recovery and was referred for psychiatric evaluation.

DISCUSSION

Bezoar is a collection of indigestible organic or inorganic material in the gastrointestinal tract; commonly the stomach. Depending upon their composition they are named as phytobezoar (vegetables and fruits), pharmacobezoars (medication, resins), trichobezoar (hair), diospyrobezoar (persimmon fibres) and lactobezoar (milk curd).

Trichobezoar was first described in 1779 by Baudomant and comprises 50% of all bezoars. A sparsely reported entity “Rapunzel syndrome” was first described by Vaughn et al in 1969. It is a rare form of gastric trichobezoar that extends...
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as a tail to the jejunum, ileum or the ileo-caecal junction causing small intestinal obstruction. It is named after 'Rapunzel' – the maiden in a German fairy tale, whose long hair flowed out of her prison tower allowing her prince to rescue her.

Trichobezoars form when hair strands are retained in the folds of the gastric mucosa due to their indigestibility and escape peristaltic propulsion because of their slippery surface. As more hair accumulate, peristalsis causes them to be enmeshed into a ball forming the trichobezoar. Sometimes it may extend as tail into small intestine causing Rapunzel syndrome.

Most patients with trichobezoars suffer from psychiatric disorders including trichotillomania (strong urge to pull out their own hair) and trichophagia (eating of hair). Only rarely do these patients chew hair from other sources including hair from wigs. Trichotillomania has a prevalence of 0.6%-1.6%. Patients with trichotillomania usually pull hair from the scalp, but eyelashes, eyebrows, legs, armpits, and pubic regions are also targets for pulling. Of the patients suffering from trichotillomania around 30% will engage in trichophagia, and of these, only 1% will go on to eat their hair to the extent requiring surgical removal. Our patient did not have trichotillomania and ate hair of dolls have from combs.

The common presentation of trichobezoar is in young females usually with an underlying psychiatric disorder and may be attributed to the traditional long hair in females. One reported male case ate the hair of his sisters. Affected patients occasionally remain asymptomatic for many years. Symptoms develop as the bezoar increases in size to the point of obstruction. Sometimes the bezoar may fragment and small pieces pass into small intestine causing intestinal obstruction, intussusception or perforation.

Majority of cases of trichobezoar present late, due to the low index of suspicion by the physician. Severe halitosis and patchy alopecia provide clues on physical examination. Presentation range from early satiety, abdominal pain, nausea and vomiting, weakness and weight loss, constipation or diarrhoea, hematemesis to obstruction and perforation. Complications by a large eroding or obstructing bezoar additionally include gastric ulceration, obstructive jaundice, acute pancreatitis and gastric emphysema. Other malabsorption-related complications include protein-losing enteropathy, iron deficiency, and megaloblastic anaemia. Palpable epigastric mass remains the commonest physical sign. Our patient had recurrent bouts of colicky abdominal pain probably due to recurrent pyloric obstruction by trichobezoar and she had a palpable mass in epigastrium.

Figure 1: Jejunum pulled into a concertina (white arrow) due to the long tail of trichobezoar (red arrow) being pulled out from gastrostomy.

Figure 2: The trichobezoar removed was a perfect cast of stomach and duodenum with tail extending to a total length of 170 cm into jejunum.
Ultrasound is effective in diagnosing bezoars in up to 88% of cases. An abdominal CT with contrast diagnoses 97% of trichobezoars, which appear as free-floating defects in the stomach often with air pockets. The gold standard for diagnosis is upper gastrointestinal endoscopy. In addition to providing direct visualization, this procedure allows sampling and potentially therapeutic intervention. The management of a bezoar need to encompass removal of the mass and prevention of recurrence by addressing the underlying physical or emotional cause to prevent recurrence. Depending on its consistency, size and location, bezoar removal may occur via endoscopy or surgery. Phytobezoars may be dissolved with various enzymes including cellulase, papain, acetylcysteine or carbonated beverages. Trichobezoars are often resistant to enzymatic dissolution and require endoscopy for removal. Specialized bezotomes and bezotriptors (that pulverize bezoars either mechanically or with acoustic waves) have been used to fragment large and solid trichobezoars. Laser initiated mini-explosive technique has also been used for endoscopic management. Laparoscopic removal has been done in a few cases. Open surgery still remains the corner stone of large trichobezoar removal, especially if it has an extension into the bowel (as in present case), which might be missed with other methods of treatment. There are no formal guidelines for the treatment of trichotillomania in children. Common treatment modalities include medication (clomipramine, or fluoxetine) and cognitive behaviour therapy. Habit–reversal training is usually used as the initial therapy in children with up to 74% response rate.

**CONCLUSION**

Trichobezoar, an under-diagnosed entity, has to be considered in the differential diagnosis of abdominal pain and a non-tender abdominal mass, and a history of pica should always be obtained. Many of these patients have psychiatric pathology with emotional problems or mental retardation. Counselling by a psychiatrist is an important part of management to prevent recurrence.

**REFERENCES**
