

## The Rapunzel Syndrome: An Unusual Trichobezoar Presentation

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### Abstract

An unusual form of bezoar extending from the stomach to the small intestine or beyond has been described as Rapunzel syndrome. Trichobezoar with Rapunzel syndrome is an uncommon diagnosis in children with less than 40 cases reported. It is predominantly found in emotionally disturbed or mentally retarded youngsters, because of its rarity its imaging features and review of literature is reported.

**Keywords:** Abdominal mass, Emotional disturbance, Mental retardation, Rapunzel syndrome, Trichobezoar

### Introduction

Bezoars are concretions of human or vegetable fibers that accumulate in the gastrointestinal tract. The word “bezoar” comes from the Arabic word “bedzehr” or the Persian word “padzhar,” meaning “protecting against a poison.” At different times in history, bezoars from animal guts were used as precious stones, antidotes to poisons and today as part of traditional Chinese medicine. The first reference to a bezoar in a human was in 1779 during an autopsy of a patient who died from gastric perforation and peritonitis.

In humans, the most common type of bezoar is the trichobezoar, which is mostly made of

hair. However, bezoars can also be made of vegetable or fruit fibre (phytobezoars), milk curd (lactobezoars), or any indigestible material.

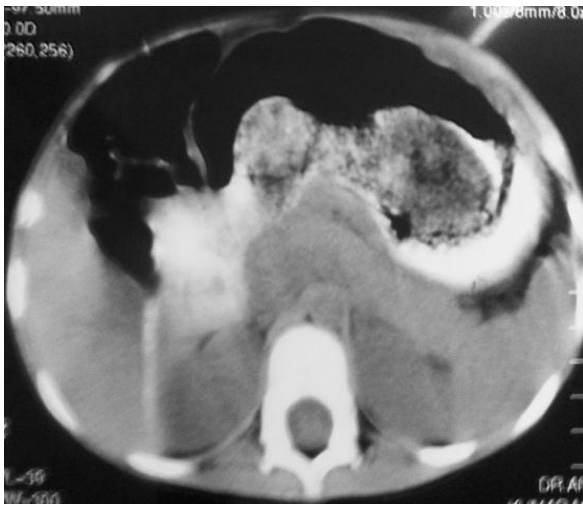
Trichobezoars, unlike other bezoars, are not associated with alterations in gastrointestinal motility but with underlying psychiatric disorders, and most commonly present in adolescents and during the second decade of life. Rapunzel syndrome is an unusual and rare form of trichobezoar extending into the small intestine. The name “Rapunzel” syndrome comes from the Grimm Brothers’ fairy tale of a 12-year-old princess who was shut into a tower with neither stairs nor doors by an enchantress who climbed up the tower’s walls with the help of Rapunzel’s long tresses.

Most cases of trichobezoar are reported in females, which may be attributed to the traditional long hair in females. One reported male case ate the hair of his sisters. A cotton bezoar with Rapunzel syndrome was recently

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reported in an 18-year-old male. The majority of these cases presented between 13 and 20 years of age.



**Fig.1: CT abdomen showing hypodense mass lesion in the body of stomach.**

### Case presentation

A 6 years old female child presented with pain abdomen since 1 month. Examination revealed a palpable mass in the epigastrium. Earlier ultrasonography done was reported as intussusception. CT abdomen (Fig.1) showed a hypoechoic mass lesion with linear streaks within it in the body of stomach measuring about 8x7 cms. Barium meal (Fig.2) revealed an irregular filling defect in the lumen of stomach extending into the duodenum with linear streaks of barium in the filling defect.

A laparotomy was performed which showed a transmural ulcer adherent to the anterior abdominal wall. Gastrotomy was done and a gastric trichobezoar of 12x5cms which has taken the shape of the stomach has been extracted along with 48cms tail.

### Discussion

The term *Rapunzel syndrome* was coined by Vaughan and associates in 1968, for an unusual manifestation of a trichobezoar in which the mass extends from the stomach and duodenum through a large portion of the small intestine.<sup>1</sup> The term *bezoar* comes from the Arabic *badzehr* or from the Persian *panzer*, both meaning counterpoison and antidote.<sup>2</sup> Hindus used bezoars in the 12th century BCE for rejuvenating the old, neutralizing snake venom and other poisons, and treating vertigo, epilepsy, melancholia, and even plague.

Causes of a bezoar can include the presence of indigestible material in the lumen, gastric dysmotility (including previous surgery such as vagotomy and partial gastrectomy, etc.), and certain other substances that encourage stickiness and concretion formation. Bezoars occur mainly in young women and are named according to the substances that are swallowed and produce them, including hair (trichobezoar), phytobezoar (vegetable fibers), diospyrobezoar (persimmon fibers), or pharmacobezoar (tablets/semi-liquid masses of drugs).<sup>3</sup>

Bezoars mostly originate in the stomach, probably related to a high-fiber diet. They generally cause nonspecific symptoms, such as epigastric pain, dyspepsia, and postprandial fullness. The patient may present with a palpable, firm, nontender epigastric mass, which may be discovered on routine physical examination in an asymptomatic patient or may be an operative surprise. Patients may also present with GI bleeding (6%) and intestinal obstruction or perforation (10%).<sup>4,5</sup> Some patients with trichobezoar show overt psychic/mental

disturbance or personality maladjustment. Though their presence may be suspected clinically and on ultrasound, trichobezoars are best identified on barium studies and CT. On ultrasound, the characteristic appearance persists irrespective of the angulation of the transducer, the alteration of position of the patient, or the administration of clear water. This excludes the clinical possibility of a pancreatic pseudocyst, a splenic or renal mass, a noncalcified gastric tumor, a gastric duplication cyst, or a gastric outlet obstruction. However, a heavily calcified mass (such as a teratoma, a neuroblastoma, or an impacted mass of feces) may produce a similar ultrasound image.<sup>6</sup>

Apart from the mottled filling defect, an upper GI study may show the positive density of the mass with a lacelike pattern due to residual contrast medium on delayed films. The fluoroscopic examination with the patient in the erect position shows the swallowed barium held up in the cardiac end of the stomach for a few seconds, as though it were forming a cap to something inside the organ. Then the contrast can be seen to suddenly diffuse slowly downward on either side of a nonopaque foreign body and follow the regular contours of the greater and lesser curvatures to map out the normal contour of the stomach.

On CT, in addition to the characteristic appearance as described above, the free mobility of the mass in the stomach lumen can be seen on supine and prone views. CT best reveals the size and configuration of the bezoar and most accurately identifies its location. Further, the highly characteristic CT appearance permits ready differentiation from other pathologies (such as intra- or extragastric neoplasms) that would be

difficult on plain radiography or on ultrasound.<sup>7</sup>



**Fig.2: Barium meal showing filling defect in the lumen of stomach extending into the duodenum with linear streaks of barium in the filling defect.**

The treatment of large bezoars and concretions is essentially surgical. Gastric bezoars may be retrieved endoscopically or via gastrotomy. Duncan et al<sup>8</sup> recommended bezoar extraction by multiple enterotomies in cases of Rapunzel syndrome. It is mandatory to do a thorough exploration of the rest of the small intestine and the stomach to look for retained bezoars. Other methods like modified needle-knife (bezotome) and a modified lithotripter (bezotripter) have also proven very successful. The patient's psychological problems also need to be dealt with.

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