

# Trichobezoar and Rapunzel syndrome: A review

## Abstract

Rapunzel syndrome is a rare variant of trichobezoar where the long tail of accumulated hairs extends to the small intestine. Gastrotomy of a 7-year-old female child revealed a mass consisting of hairs and remains of threads that occupied the entire stomach extending up to the jejunum. This is a unique case report of Rapunzel syndrome with habit of eating threads in addition with hair. Treatment and psychological support of the mental as well as physical disorder is important for prevention of its recurrence.

**Key words:** Rapunzel syndrome, trichobezoar, trichotillomania

## INTRODUCTION

Trichobezoars usually occurs in children and adolescent women with the history of trichotillomania (pulling hairs), trichophagia, gastric dysmotility and usually with a baseline paranoid-type mental disorder. The extension into the small bowel through the pylorus in the form of a tail - the Rapunzel syndrome, is a rare occurrence. Clinical presentation is deceptive and vague, ranging from an abdominal mass to gastrointestinal symptoms. The presenting case represented the rare condition of the Rapunzel syndrome in a 7-year-old child treated surgically. This case is very unique because the bezoar is not exactly trichobezoar, but it was a mixture of hairs and threads.

## CASE REPORT

A 7-year-old female child had presented with the complaints of intermittent abdominal pain, nausea, vomiting, and early satiety. Three months back, her parents had noticed that she has the habit of picking and eating hair from her head and threads from her cloths. Milestone development was reported normal, except delayed eating (other than breast milk) at the age of 3 years. History of irritable behavior was present. On examination, there present a movable and firm mass of size 7 cm × 3 cm size. Barium follow-through revealed filling defect with whorled, stippled and mottled appearance, extending up to the proximal part of jejunum. During planned laparotomy, a mass was palpated in the stomach, which was removed by gastrotomy. The mass consisted of hair and remains of threads that occupied the entire length of the gastric chamber and extending through duodenum up to 5 cm of jejunum [Figure 1]. Biopsy revealed numerous hairs, hair follicles in the background of amorphous degenerated vegetative material. Trichotillomania was diagnosed on psychiatric assessment and treated accordingly. Now she is maintaining well.

## DISCUSSION

Bezoar is an assortment of foreign material in the intestinal tract (usually stomach). It is mainly caused by the presence of indigestible substances in the lumen, gastric dysmotility including previous gastric surgery such as vagotomy and partial gastrectomy or antrectomy; and some substances that encourage stickiness and concretion formation. It is common in young females usually with mental retardation and an underlying psychiatric disorder, who chew and swallow their hair (trichobezoar), vegetable fibers (phytobezoar), and persimmon fibers (diospyrobezoar). Although the exact cause of trichotillomania is not clear; certain psychosocial, behavioral, and biological theories have been proposed such as childhood trauma, stress, and neurochemical imbalances (like of serotonin).<sup>[1,2]</sup> The first case was reported by Baudomant in 1779. Surgical removal was first performed in 1883 by Schonbern. Trichobezoars are usually symptomless until they reach a large size. They may present with varied upper gastrointestinal symptoms, wasting and cachexia. Large bezoars are often palpable and may be indentable (Lamerton's sign). Trichobezoars weighing 2500 g have been reported.<sup>[3]</sup> The main complications of trichobezoar include ulceration, gastrointestinal perforation, obstruction, intussusception and diarrhea and vitamin

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**Figure 1:** Specimen obtained after gastrotomy

B12 deficiency (secondary to bacterial overgrowth by colonization of the bezoar). Conventional radiography shows a mass of opaque soft tissue in a dilated stomach and a calcified rim may delineate the edge of the bezoar. Ultrasonography may reveal curvilinear mass with bright echogenic band that does not allow transmitting the ultrasound waves and generate a shadow over the left upper quadrant. The high echogenicity of hair and the presence of multiple acoustic interfaces (created by trapped air and food) limit its diagnostic ability. Both contrast radiography (barium meal) and upper gastrointestinal tract endoscopy are the procedures of choice for establishing the diagnosis. Upper gastrointestinal contrast radiography typically shows an intragastric mass with barium in the honeycomb interstices and might detect other complications such as gastric ulcers. A mottled gas pattern (air bubbles retained within the bezoar) is characteristic on computed tomography scan.

When trichobezoar extends from the stomach into the duodenum, the proximal small intestine or to the ascending colon it is called Rapunzel syndrome. It is named after a tale written in 1812 by the Brothers Grimm about a young maiden, Rapunzel, with long hair who lowered her hair to the ground from a castle, which was a prison tower, to permit her young prince to climb up to her window and rescue her. The first case was published in 1968 by Vaughn. The tail may extend to the ileocecal junction. After a thorough search of literature, we found total 64 cases of Rapunzel syndrome reported [Table 1]. The additional complications are jaundice, acute pancreatitis secondary to obstruction of the ampulla of Vater and peritonitis. Chemical dissolution, mechanical fragmentation and laser-ignited mini-explosive technique were used successfully for small bezoars.<sup>[4]</sup> Laparoscopic techniques are also becoming fashionable and large bezoars can be milked into the caecum before removal. Open surgery is essential, especially if there is an extension into the bowel. Resection of the bowel is required if enterotomy (single/multiple) removal is not feasible. A thorough search of the stomach and whole intestine for retained bezoars is mandatory. Recurrence mainly occurs because the underlying psychological trigger was not corrected. Untreated bezoars have mortality of 75% and there is a 4% mortality.<sup>[5]</sup> To the best of our knowledge, only two cases of Rapunzel syndrome with bezoars of threads of cloths are reported.<sup>[6,7]</sup> We are the third one to report the habit of eating thread in a case of Rapunzel syndrome.

## CONCLUSION

- Rapunzel syndrome is a rare entity. It can occur with the habit of eating threads of cloths also. This possibility should be considered in young female with chronic pain abdomen

**Table 1: Brief review of Rapunzel syndrome**

Article (s)	Special features
Mohite <i>et al.</i> <sup>8</sup>	Case report of on table diagnosis of Rapunzel syndrome in a case operated for gastric perforation. They review 27 cases of Rapunzel syndrome
Rabie <i>et al.</i> <sup>4</sup>	Reported that only 24 cases have been reported in the literature. One of them was pregnant and had small bowel intussusception and perforation, a very rare combination.
28 simple case reports	No special feature described
Dindyal <i>et al.</i> <sup>9</sup>	Case report describing unusual radiologic sign associated with a retroperitoneal perforation of the third part of duodenum – the comma sign.
Salem <i>et al.</i> <sup>10</sup>	Case report of Rapunzel syndrome that was complicated by pancreatitis.
Matejů <i>et al.</i> <sup>11</sup>	Reported death resulting from Rapunzel syndrome due to gastric perforation and parental neglect.
Janes <i>et al.</i> <sup>12</sup>	Recurrent Rapunzel syndrome in a 37-year-old woman.
Jain <i>et al.</i> <sup>7</sup>	Case report of a large trichobezoar in a 3-year-old male child who presented to the emergency department with THREADS protruding from mouth with no sign of hair loss on body.

- Treatment of the underlying behavioral or neuropsychological disorder should be given equal importance during the treatment of the disease.

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