

# A Rare Case Report of Rapunzel Syndrome: Celiac Disease with Generalized Edema

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## Abstract

Rapunzel syndrome is a rare disease. In this syndrome, hairs the person has eaten become tangled and trapped in their stomach and can cause symptoms such as gastric outlet obstruction, small intestine obstruction, gastrointestinal bleeding, perforation of the gastrointestinal wall, abdominal mass, nausea and vomiting. A 46-year-old woman presented with generalized edema and abdominal pain. The patient was diagnosed with celiac disease several years ago on a gluten-free diet. In the experiments, the patient had low serum albumin and anemia. In transit of the small intestine, the patient had a dilated stomach and duodenum containing a mass, which was diagnosed by endoscopy and could not be extracted with an endoscope due to the large mass. As a result, the patient was referred to a surgeon and underwent laparotomy and the mass was extracted.

**Keywords:** Edema; Celiac Disease; Endoscopy; Trichobesar; Rapunzel Syndrome

## Introduction

Bezoar is an indigestible eaten substance that accumulates gradually in the gastrointestinal tract, especially in the stomach, but can be found in other sites of the gastrointestinal tract from the esophagus to the rectum [1]. Bezoars are divided into several categories: hair accumulation (trichobezoar), vegetables (phytobezoar), drugs (pharmacobezoar), persimmon (disopirobazvar), milk proteins (lactobozvar). The prevalence of bezoar has been reported in different studies. Gastric bezoars have been reported with an incidence of 0.3% in upper endoscopy and incidence of 0.4 to 4.8% has been reported in patients presenting with small bowel obstruction [2]. The prevalence and type of bezoars vary depending on the age, geographical locations, eating habits and culture of the people of each country. Therefore, in countries such as South Korea and Japan where the people have a diet high in vegetables, the

prevalence of phytoplankton is more common in children and adolescent girls [3]. In a healthy person, bezoar formation is uncommon and usually some degree of dysfunction of the stomach and also the nature of the eaten substance are involved in its formation [4]. Other risk factors for bezoar formation include diabetes, hypothyroidism, and lack of healthy teeth [5]. Although sometimes asymptomatic, bezoars may cause serious symptoms, such as abdominal discomfort or pain, dysphagia, small bowel obstruction, gastric ulcer, ileus, gastrointestinal bleeding, abdominal mass, pancreatitis, etc [6-8]. Among most types of bezoar, trichobezoar is commonly found in people with Pica and other psychiatric conditions such as emotional disturbances, learning disabilities, and a history of inattention or mental retardation. These people pull out their hair and swallow it [9-11]. One of the rare manifestations of trichobezoar is Rapunzel syndrome, which refers to the spread of bacillus to the small intestine, so that it can continue even until the

cecum [12-14]. Rapunzel Syndrome takes its name from a long-haired girl in the nineteenth-century folk tale of the Brothers Grimm, who was imprisoned in the castle tower and can only be released by a Prince rising through her long hair. Common symptoms of this syndrome include abdominal pain, vomiting, nausea, weight loss, malnutrition, vomiting of blood, diarrhea, or constipation. In this article, we present a case of a 45-year-old woman with celiac disease and Rapunzel syndrome who was admitted with generalized edema.

## Case Presentation

The patient was a 45-year-old woman with a known case of celiac disease who presented with edema of both lower limbs and abdomen one month ago. The patient has had a slight swelling in her legs for several months, which has gradually increased and reached to the thighs and lower abdomen. The patient also reported occasional nausea and vomiting with abdominal pain, which usually occurred one

hour after eating. The patient has anorexia in the last three months and mentions about 5 kg of weight loss. She has a history of celiac disease from several years ago, who had adherence on a gluten-free diet. The patient underwent laparotomy four years ago due to small bowel obstruction caused by Trichobezoar. The patient also reported a history of depression that had been under the supervision of a psychiatrist for some time but had not been followed up.

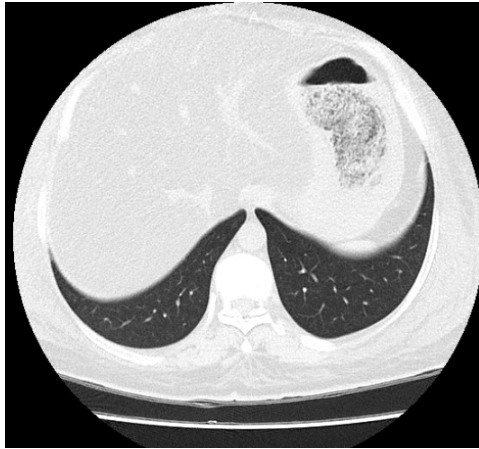
On physical examination, clinical signs are normal and bilateral edema of the legs is seen with ascites and the appearance of orange peel on the lower half of the abdomen. On the inner part of the right ankle, there was a superficial wound with a dark appearance without discharge. There was no special point in the examination of the head, neck and chest regions. In the abdomen with deep touch there was a slight tenderness in the epigastrium but no rebound sensitivity. There was no organomegaly and lymphadenopathy. Table 1 lists the patient's tests.

Lab Data	Value	Normal Range
White blood cells	8400	3.54-9.06 × 10 <sup>3</sup> /mm <sup>3</sup>
Hemoglobin	10.6	12.0-15.8 g/dL
Platelets	276	165-415 × 10 <sup>3</sup> /mm <sup>3</sup>
Hematocrit	36.3	35.4%-44.4%
Mean corpuscular volume (MCV)	69.5	79-93.3 fL
Partial thromboplastin time (PTT)	38.6	26.3-39.4 s
Prothrombin time (PT)	15.8	12.7-15.4 s
International normalized ratio (INR)	1.17	
BUN	12	7-20 mg/dL
Creatinine	0.86	0.5-1.9 mg/dL
Aspartate transaminase (AST)	48	12-38 U/L
Alanine transaminase (ALT)	64	7-41 U/L
Alkaline phosphatase (ALP)	278	33-96 U/L
Total bilirubin	0.85	0.3-1.3 mg/dL
Stool levels of alpha-1-antitrypsin	821	< 248 mcg/g
Albumin	1.72	4.0-5.0 g/L
Na	133	136-146 meq/L
K	3.8	3.5-5.0 meq/L
Calcium	6.7	8.7-10.2 mg/dL
Ferritin	18	10-150 ng/ml
Reticulocyte count	0.7	0.5-1.5 %
TSH	7.7	0.30-4 µIU/ml
C-Reactive Protein (CRP)	2.81	< 1 Negative
Erythrocyte Sedimentation Rate (ESR)	4	0-30 mm/hr
Anti-transglutaminase antibodies (Anti TTG Ab)	21	Negative < 18

**Table 1:** Hematologic laboratory tests of the presented case.

The image of a hypoechoic mass with marked hyperechoic dimensions of approximately 22 x 20 mm and without vasculitis was clearly visible in the 7-segment liver on ultrasound. Atypical hemangioma was present in the first diagnosis and adenoma in the next diagnosis. No pathological findings were reported elsewhere in the abdomen and pelvis, so a triphasic CT scan was requested to differentiate the

liver lesion, which confirmed hemangiomas. Intra luminal lesion was reported in CT of the stomach (Figure 1). Due to the history of celiac disease in the patient, generalized edema, normal value of Anti TTG Ab and weight loss, with the possibility of celiac complications such as small bowel lymphoma for the patient, small bowel transit was requested (Figure 1).



(A)



(B)

**Figures 1:** A: CT view which revealed Gastric intraluminal mass with gastric, B: duodenal dilation with intraluminal mass like lesion.

Due to the patient's epigastrium, the presence of a lesion in the CT scan and transit of the patient's small intestine, an upper endoscopy was performed for the patient, which

showed a large bezoar tricot that was extended to the distal jejunum (Figure 2).



(A)



(B)



(C)

**Figure 2:** Endoscopic view of trichobezoar in gastric lumen which extended to distal part of jejunum. A: Trichobezoar in distal part of D3, B: Antrum of stomach C: Body of stomach.

## Discussion

Most trichobezoars are more common in girls in the second decade of life and is usually detected due to nonspecific complaints in the upper endoscopy; so far it has been seen in a handful of men [15-18]. In trichobezoars, swallowed hair accumulates in the stomach, and gastric acidity breaks down

hair proteins and darkens the hair by oxidation. Later, the hair becomes entangled with the accumulated food particles. As a result, colonized bacteria and fermentation of undigested fat can lead to bad breath. Classically, trichobezoar is caused by eating hair (trichophagia), which can be related to the patient's mental state [19]. The patient reported a history of depression that was treated for some time. After

the diagnosis, the patient was under the supervision of a clinical psychologist. Also, due to laparotomy history, the possibility of small bowel obstruction in this patient is high, and on the other hand, due to the possibility of intestinal motility disorder, it can lead to the formation of a bezoar. The most common manifestations of Rapunzel syndrome are epigastric pain, premature satiety, nausea, anorexia, weight loss, intestinal obstruction, and intussusception. Also cases of obstructive jaundice [20], pancreatitis [21], appendicitis [22], and intestinal perforation have been reported [23,24]. On abdominal examination, the mass may be felt in the upper abdomen (Lamerton's Sign).

In a review of previous articles, hypoalbuminemia was reported in a handful of cases of Rapunzel syndrome, which was associated with generalized edema in only one case [25-29]. This is the first case of Rapunzel syndrome to be presented with generalized edema in an adult; two cases of generalized edema of this syndrome have previously been reported with generalized edema in children [29].

Protein Loss Enteropathy (PLE) 1 is a rare disease characterized by protein loss through the gastrointestinal tract and results in decreased serum protein levels. Most excreted protein is albumin, but other proteins such as immunoglobulins and ceruloplasmin are also lost [30,31].

### **Gastrointestinal Disorders That May Cause PLE Include**

Abnormalities of the lymphatic system, such as primary intestinal lymphectasia or secondary to obstruction (cancer or colitis) or increased lymph pressure (congestive heart failure) leading to leakage of protein-rich lymph. 2. Mucosal damage, resulting in increased mucosal permeability due to mucosal erosions such as inflammatory bowel disease, some intestinal bacterial infections such as Salmonella and Shigella, or non-ulcerative intestinal diseases such as celiac disease, mantir disease, and allergic gastroenteritis [30].

The diagnosis of PLE is based on clinical suspicion and confirmation by increasing the alpha level of a fecal antitrypsin, as noted in our patient. In our case, enteropathy was the protein loss manifestation of a large trichobezoar that caused partial obstruction and malabsorption of the intestine.

Protein-losing enteropathy, inadequate food intake, malabsorption, and bacterial overgrowth may lead to long-term, slow-growing hypoalbuminemia [32,33]. The pathophysiology of trichobezoar formation begins with the long-term stay of swallowed hair fibers, which later cause gastrointestinal peristaltic movements to form a ball. The hair mass develops so much that it makes it impossible to empty

the stomach. Regardless of the color of the ingested hair, due to enzymatic oxidation of stomach acid, trichobezoar stands usually on black hair and often have an unpleasant odor due to fat fermentation [9]. In uncomplicated cases, blood tests are usually in the normal range. Some patients may develop iron deficiency anemia and hypoalbuminemia due to malabsorption.

Diagnostic studies include ultrasound, plain photo assessment or barium study, CT scan, and upper endoscopy. Ultrasound can show a highly echogenic band formed by a mixture of swallowed hair fibers with food and air in the trichobezoar. Ultrasound is usually non-diagnostic [34]. Plain and abdominal barium imaging may be useful in confirming the clinical diagnosis and determining gastrointestinal obstruction secondary to trichobezoar. CT scan has the highest diagnostic accuracy so that the trichobezoar can be seen as a heterogeneous mass and hypodense [5,35-37].

The therapeutic goal of trichobezoars is to completely eliminate the mass and prevent its recurrence. Dissolution of the mass with chemicals (Coca-Cola drinks) or removing it by endoscope is not effective in Rapunzel syndrome and large trichobezoar [38].

Nowadays, surgeons are very interested in laparoscopic methods due to fewer complications, more willingness of patients and shorter hospital stay. Several cases of these methods have been performed in patients with trichobezoar, but the standard treatment for Rapunzel syndrome is still open surgery [21,39,40]. Our patient underwent open surgery after correction of albumin with intravenous albumin and correction of INR with intravenous vitamin K, and after extraction of bezoar and supportive and nutritional cares, she was under the supervision of a psychiatrist and clinical psychologist.

### **Conclusion**

Rapunzel syndrome is difficult to diagnose and requires high diagnostic suspicion. This is the first case of Rapunzel syndrome in an adult with generalized edema. Internal medicine specialists should consider trichobezoar disease in the differential diagnosis of enteropathy of protein loss in young women, especially if there is a behavioral disorder in the patient's history. Early diagnosis and treatment are important to prevent potentially fatal complications. Psychological and psychiatric evaluation and follow-up are also recommended to treat and prevent recurrence.

### **Conflict of Interests**

The authors declare that there is no conflict of interest among them.

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