Case Report

An Unusual Cause of Anaemia: Rapunzel Syndrome, A Case Report

Aysegul Ertinmaz Ozkan¹, Nilay Danis², Mustafa Altundal³, Nurhayat Ozkan Sevencan⁴, Fatih Karatas⁵, Burcak Kayhan⁶

¹,⁴,⁶ Department of Internal Medicine, Education and Research Hospital, Karabuk University, Karabuk, Turkey; ² Department of Gastroenterology, Education and Research Hospital, Karabuk University, Karabuk, Turkey; ³ Department of General Surgery, Education and Research Hospital, Karabuk University, Karabuk, Turkey; ⁵ Department of Medical Oncology, Education and Research Hospital, Karabuk University, Karabuk, Turkey

Correspondence: Nilay Danis. Email: nilaydanis17@gmail.com

Abstract

Trichobezoars present with stomach ache and with a mass in the stomach. It’s common in the young and middle-aged women having psychiatric disorder, presenting with stomach ache and existence of mass in the stomach. Although it’s one of the rare causes of anaemia it should be considered when dealing with cases of chronic and unresponsive anaemia

Keywords: Rapunzel syndrome; Iron Deficiency Anaemia; Trichobezoar; Trichotillomania.

Introduction

Trichobezoar, commonly known as a hairball is a wad of swallowed hair which is frequently seen to occur in young and middle aged women suffering from
Psychiatric disorders Trichobezoars most commonly located in the stomach can become gigantic masses in the stomach over a period of years extending into Duodenum and Jejunum. Such cases are called as having Rapunzel Syndrome (RS).\(^1\) The most commonly observed features of Rapunzel Syndrome are stomach ache and Iron Deficiency Anaemia. But in younger patients it can present with signs and symptoms of gastric perforation, hemorrhage and gastrointestinal obstruction.\(^2\) Although Trichobezoars are a rare cause of Iron Deficiency Anaemia and gastric pain it should be kept essentially in mind while dealing with patients having psychiatric disorders.

Considering its high morbidity and mortality rate, we are presenting here a Case Report of a 19-year-old female patient having Trichobezoar in her stomach which extended up to her duodenum.

**Case Report**

On 10\(^{th}\) of November 2018 a 19-year-old female patient reported to the Internal Medicine outpatient Department of Karabuk University Faculty of Medicine Hospital, with the complaints of fatigue and loss of appetite extending over a period of last four months. She was generally a weak and pale looking female fully conscious having a BP of 100/70 mm Hg and a pulse of 96 beats / minute.

On her general physical examination she was found to have anaemia but had no signs of oedema, icterus and cyanosis and her lymphnodes were not found to be enlarged. Her laboratory findings were as follows; Hb: 3.3g/dL, Htc: 9.4%, MCV: 68 fl, Serum Iron: 16 mg/dL, Total Iron-Binding Capacity: 430 mg/dL, Ferritin: 5 ng/mL, and Vitamin B12: 396pg/mL. The other biochemical and haematological blood parameters were found to be normal. Hypochromia, microcytosis, anisocytosis, and poikilocytosis were found to be present on her Peripheral blood smear. Since she had all the signs and symptoms of anaemia she was transfused 4 units of packed red blood cells.
On her Abdominal computed tomography (CT abdomen) a foreign object trapping air was identified as filling the gastric lumen (Figure 1). On her Esophagogastroduodenoscopy a giant ulcer of 3 to 4 cm in diameter and a Trichobezoar covering the luminal surface of the stomach extending into the bulbus was identified which could not be removed endoscopically (Figure 2). The patient underwent gastrostomy and a Trichobezoar, approximately 25 x 15 cm in dimensions and weighing 950 gms, was removed (Figure 3). On the pathological examination of ulcer it was found to be non-malignant. The patient later confessed to be having Trichophagy for which she was referred to the Psychiatry Department.

Discussion
Bezoars are the masses created by the objects that cannot be digested in the gastrointestinal system. According to the objects they contain, Bezoars are categorized as Phytohezoars (consisting of the indigestible food particles in vegetable or fruit fibers), Trichobezoar (consisting of the combination of hair and food particles), Lactobezoars (milk protein), and Pharmacobezoars (various medicines). Phytohezoars are the most commonly known bezoars and constitute approximately 40% of all cases. Bezoars are mostly found in the stomach, esophagus, small intestine, large intestine, and bile ducts. Trichobezoars are the hairballs that mostly occur in children and adolescents, accumulating particularly in the stomach, because they cannot be digested in the gastrointestinal system. In these patients, psychiatric comorbidities may occur involving strong impulses, which lead patients to draw (Trichotillomania) and eat (Trichophagy) their own hair. Trichobezoars occur when the hair strands, which escape from the peristaltic impulses due to the slippery surfaces, are retained in the curls of the gastric mucosa, growing and extending into the distal sections of the bowel (Duodenum, Jejunum), because of the impacts of
peristalsis. This case was first identified as Rapunzel Syndrome by Vaughan et al in 1968.4

In our case, it was observed that Trichobezoar, which resulted in RS in a 19-year-old girl, led to severe anaemia, without any symptoms. In fact, according to the literature, most of the Trichobezoar cases are diagnosed late, although they often cause epigastric pain, nausea, vomiting, absence of appetite, loss of weight, malabsorption of trace elements. In cases with delayed diagnosis, malabsorption or severe anaemia caused by gastrointestinal bleeding might occur.5 As a rare complication, they can result in ileus, perforation, ulceration, bleeding, pancreatitis, obstructive jaundice, superior mesenteric artery syndrome, intussusception, and peritonitis, leading to a high morbidity and mortality. IFA, commonly observed in patients with trichobezoars, is a direct result of certain gastric ulcers due to the pressure impacts of the mass. If not treated, the mortality ratio associated with gastrointestinal bleeding, destruction, or perforation can increase up to 30%.2

Although a palpable mass in the abdomen initially gives rise to a suspicion of a malignant process going on, Trichobezoar should be kept in mind in the patients with any psychiatric disorders presenting with bad smell in breath and/or existence of a patched alopecia. Although barium radiographies or abdominal CT demonstrate Trichobezoar or RS, particularly with spotted intraluminal mass consisting of the hair entangled with air in the abdomen and small intestine, upper gastrointestinal endoscopy should be used for a final diagnosis.

Endoscopy, a Gold Standard method, is the easiest invasive method for the diagnosis as well as the treatment (removal of Bezoar). If it is impossible to remove Bezoar by endoscope, pharmacological approaches (i.e. Coke, motility stimulants) can be employed. There are data indicating that Bezoar, which was exposed to coke diet for 1 week, was observed to be partially dissolved.3, 6 However, since these treatment options are unsuccessful in most RS cases, surgical removal by laparotomy or laparoscopy should be employed or
considered as a treatment option. It was demonstrated that success ratio of laparoscopic treatment, which is 75%, reaches to 99% by laparotomy. Although the success level is higher with laparotomy, the risk of complications is higher too. In laparoscopy, the limiting factors are seeding and the risk of peritoneal contamination.

Although most of the patients with Trichobezoars have psychiatric disorders involving Trichotillomania and Trichophagy, the Trichobezoar occurs only in 1% of the patients with Trichophagy. Most of the Trichobezoar cases occur in women. Eighty percent of them appear in childhood/adolescence period. A recent study demonstrates that its prevalence increases in the 7-8 and 11-12.5 years of ages and the onset of disease is generally triggered as a reaction to negative emotions (stress, anxiety). It is estimated that 1 of 2000 people around the world has Trichotillomania, with 30% of them having Trichophagy. The adults with Trichotillomania have higher anxiety and depression ratios than the general population. As most of the patients have psychiatric pathologies accompanied by emotional problems and eating disorders, psychiatric consultation plays a significant role to prevent the recurrence of Bezoar. Although there is inconsistency in terms of the studies on pharmacotherapeutic treatment for Trichotillomania, there are studies which demonstrate that some patients may respond to selective serotonin reuptake inhibitors.

Conclusion

Rapunzel syndrome is a significant clinical entity, which results in serious complications including severe anaemia, particularly observed in women and adolescents. Early diagnosis and a suitable treatment may decrease its morbidity and mortality. Another important point is the prevention of its recurrence, and for which the patients should be referred to psychiatric clinics for a thorough psychological assessment along with treatment.
Disclaimer: None to declare.

Conflict of Interest: None to declare.

Funding Sources: None to declare.

Patient approval: An informed consent form was obtained from the patient for publishing of this paper.

References


Figure 1: The large, intraluminal solid mass on computed tomography.

Figure 2: A view from endoscope, giant gastric ulcer and a large mass filling the gastric lumen
Figure 3: The large, intraluminal solid mass that was removed from stomach