

An unusual cause of anaemia: Rapunzel syndrome, a case report

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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.

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Introduction

Trichobezoar, commonly known as a hairball. It 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 RS are stomach ache and Iron Deficiency Anaemia (IDA). But in younger patients it can present with signs and symptoms of gastric perforation, haemorrhage and gastrointestinal obstruction.² Although Trichobezoars are a rare cause of Iron Deficiency Anaemia (IDA) 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 upto to her duodenum.

Case Report

On 10th of November 2018, a 19-year-old female patient applied to the Internal Medicine outpatient clinic 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 Blood pressure of 100/70 mm-Hg and pulse 96 rate / minute.

On physical examination she looked pale, however oedema, icterus, cyanosis and lymphadenomegaly. Her laboratory findings were as follows; Hb: 3.3g/dL, HCT: 9.4%, MCV: 68 fl, Serum Iron: 16 mg/dL, Total Iron-Binding Capacity: 430 mg/dL, Ferritin: 5 ng/mL, and Vitamin B12:396 pg/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 asymptomatic anaemia, 4 units of packed red blood cells were transfused to her.

On abdominal computed tomography, a foreign object trapping air was identified as filling the gastric lumen (Figure-1). Esophagogastroduodenoscopy revealed a giant ulcer of 3 to 4 cm in diameter and a Trichobezoar covering the luminal surface of the stomach extending into the



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

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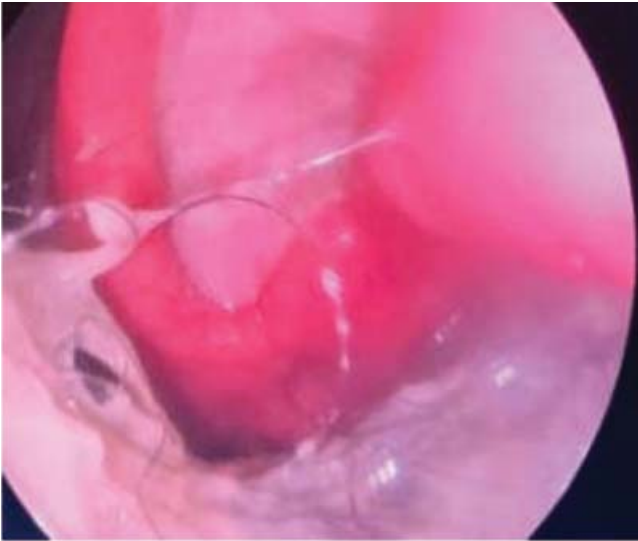


Figure-2: A view from endoscope, giant gastric ulcer and a large mass filling the gastric lumen.



Figure-3: The large, intraluminal solid mass that was removed from stomach.

bulbus was identified which could not be removed endoscopically (Figure-2). The patient underwent gastrotomy and a Trichobezoar, approximately 25 x 15 cm in dimensions and weighing 950 grams, was removed (Figure-3). On the pathological examination of ulcer there were not malignant histological findings. On psychiatric evaluation two weeks after the surgery, the patient admitted to having Trichophagy.

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 Phytobezoars (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). Phytobezoars are the most commonly known bezoars and constitute approximately 40% of all cases.³ Trichobezoars 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 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, with weakness. 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. IDA, 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, obstruction, 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 is, a gold standard method, and the easiest invasive method for the diagnosis as well as the treatment (removal of Bezoar). If it is impossible to remove Bezoar with endoscope, pharmacological approaches (i.e. coke, motility stimulants) can be applicable. In the literature, coke diet for 1 week in patients with Bezoars, provided partially regression of Bezoars.^{3,6} However, since these treatment options are unsuccessful in most RS cases, surgical removal with laparotomy or laparoscopy should be applied or considered as a treatment option. It was demonstrated that success ratio of laparoscopic treatment, which is 75%, reaches to 99% with laparotomy.⁷ 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, however, 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).⁸ It is estimated that 1 of 2000 people around the world has Trichotillomania, with 30% of them having Trichophagy.² The adults with Trichotillomania have higher anxiety and depression ratios than general population. Because of 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 no consensus in the literature for the treatment of Trichotillomania, there are studies which demonstrate that some patients may respond to selective serotonin reuptake inhibitors.⁹

Conclusion

Rapunzel syndrome is an important clinical entity, which

results in serious complications including severe anaemia, particularly observed in women and adolescents. Early diagnosis and an appropriate treatment may decrease its morbidity and mortality. Another important point is the prevention of its recurrence, and therefore the patients should be referred to psychiatric clinics for a sufficient psychological assessment along with treatment.

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