



Multiple small bowel perforation in a young adult female due to Rapunzel Syndrome



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Ozlem Zeliha Sert

Gastrointestinal Surgeon, Haydarpasa Training and Research Hospital, Istanbul, Turkey

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INTRODUCTION: *Rapunzel's syndrome is the state of the gastric trichobezoar (hair ball) extending to the small intestine. Although it is seen very rarely, it is a psychiatric and mental illness especially in young women. We present here, a young female with recurrent trichotillomania.*

CASE PRESENTATION: *In a 24-year-old patient who underwent laparotomy due to recurrent trichobezoar, which completely filled the stomach and extended to the small intestine, caused perforation in two places in the small intestine, was removed by gastrotomy and enterotomy. One of the perforated areas was very close to ligament of Treitz, and perforation occurred due to the ulcer caused by trichobezoar. Post-operative central total parenteral nutrition was given and oral intake was started at the end of the first week. The patient was referred to psychiatry and antipsychotic medication was started. She was discharged on the 9th postoperative day without any problem.*

CONCLUSION: *Although there are no signs of peritonitis in such Rapunzel syndrome cases, a detailed exploration should be made by keeping in mind that trichobezoar will cause ulceration and perforation. In addition, as the primary cause is the underlying psychiatric disorder, patients should be referred to the psychiatry clinic to prevent recurrence.*

KEY WORDS: Adult, Rapunzel syndrome, Surgery, Trichobezoar, Trichotillomania

Introduction

Bezoars are piles that accumulate in the digestive system as a result of the collection of non-digestible substance¹. Rapunzel syndrome is a quite rare clinical entity caused by trichotillomania². It is among the eating disorders and obsessive compulsive behaviors and it is a psychiatric disorder especially seen in young women as a result of unconsciously eating compulsive hair (trichophagia)³.

Hair accumulates in the mucosal folds of the stomach over time. These accumulated hair takes the shape of the stomach and become covered with mucosa and is called trichobezoar. If this gastric bezoar extends to the small intestine, it is called Rapunzel syndrome². Bezoars are usually treated with chemical dissolutions or endoscopic methods. However, surgery may be required in complicated cases and perforation⁴. Here we present a young woman with Rapunzel syndrome who has recurrent trichotillomania, causing multiple perforation in the small intestine.

Case Presentation

A 24-year-old female patient was admitted due to long-term dyspeptic complaints, nausea and vomiting. In her past medical history, she underwent surgery due to trichotillomania when she was 14-years. Interestingly, although the girl was referred to the psychiatry clinic, she did not make any application or receive any treat-

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Correspondence to: Ozlem Zeliha Sert, MD, Gastrointestinal Surgeon, Haydarpasa Training and Research Hospital, Istanbul, Turkey (e-mail: drzozlemsert@gmail.com)

ment. In addition, she is married and has a child. On examination she had tachycardia and a mass was palpated in the abdomen. At the time of application, blood test showed elevated white blood cell(WBC) count of 17400 μ L and elevated c-reactive protein(CRP) 6.6 mg/L with hypoalbuminemia(25mg/dl) and normal range of electrolytes.

After that, contrast enhanced abdominopelvic computer tomography(CT) showed a heterogeneous mass about 20x12x7 cm, filling the gastric cavity and same materials were found in the jejunum caused pseudo-invasion (Fig. 1). Next, gastroscopy was performed under general anesthesia. Gastroscopy revealed a large, black, mucus coated trichobezoar, almost occupying nearly completely the gastric cavity. Gastroscopic removal was attempted but was not successful. Therefore, surgical exploration decision was made. Since it was in the Covid-19 pandemic period, laparotomy was planned instead of laparoscopic approach. First, an anterior gas-

trotomy was performed and the giant trichobezoar covering the stomach was removed (Fig. 2). Then, gastro-tomy was closed in double coats with 2/0 polyglactin suture. In the continuation of the exploration, it was observed that there was a perforation in approximately 2x2 cm mesenteric area due to the ulcer in the jejunal segment close to the ligament of Treitz. Later, a 1x1cm diameter perforation was observed in the proximal ileum (Fig. 3). It was observed that trichobezoar continues in this area and the cause of perforation was thought to be due to trichobezoar necrosis. The trichobezoar was removed and the perforated area in the ileum was transversely closed with 3/0 polyglactin. Then, the perforated area in the jejunum was also fully sutured, and a soft drain was placed in the paracolic area and the Douglas space, and the operation was terminated. Histopathology reported as trichobezoar and ulcerated areas. Total parenteral nutrition was given to the patient who could not take oral food for a long time. Oral water was started

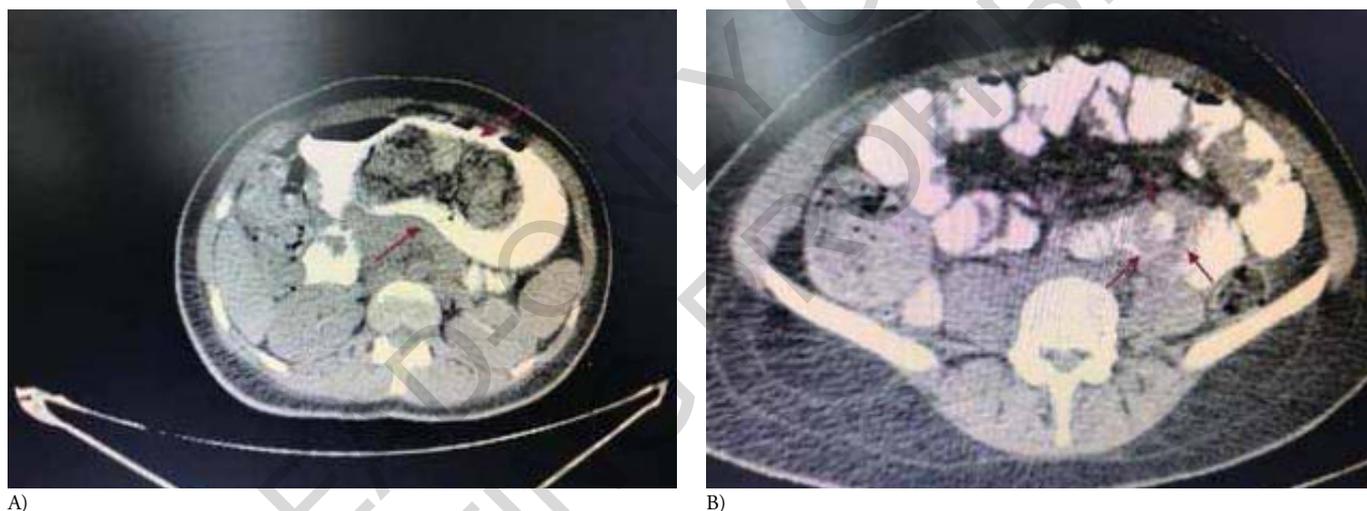


Fig. 1: Axial sections of contrast enhanced abdominopelvic CT. A) The arrow shows a large, heterogeneous solid appearing mass in the gastric cavity. B) The arrow shows a heterogeneous solid non-enhancing mass in the jejunum.



Fig. 2: A) The arrow shows anterior gastrotomy. B) Photograph of removed trichobezoar. C) Removed trichobezoar after enterotomy.



A)



B)

Fig. 3: A) Perioperative view of perforation area of proximal jejunum. B) Perioperative view of perforation area of proximal ileum.

at the end of the first week, and then solid food was gradually started. As a result of the psychiatric consultation, the antipsychotic drug risperidone tablet was started. She was discharged postoperative 9th day uneventfully. No problems were encountered in the follow-up two weeks later.

Discussion

Rapunzel syndrome is a very rare clinical entity and is frequently seen especially in young women with trichotillomania and trichophagia. There is usually an underlying psychiatric disorder or mental retardation. Trichobezoar cases have also been reported in children in the literature^{2,5,6}. Hair cannot be digested in the human stomach, it accumulates over time between mucosal folds and is covered with mucus, and because they are resistant to peristalsis, they cannot pass into the duodenum and turn into giant masses that take the shape of the stomach^{3,7,8}. Methods such as chemical solvents, motility enhancers, endoscopic removal are used in the treatment of trichobezoar. In fact, in a case report, they reported that a 13-year-old girl injected Coca-Cola into the trichobezoar from different directions by the endoscopic method in her stomach and that the trichobezoar was dissolved¹. However, surgery is required if these methods fail or in the state of perforation. Minimally invasive method or open method can be used surgically. With the widespread use of minimally invasive methods, laparoscopic or robotic approach is generally preferred⁹. In our case, because of the endoscopic method failed, laparoscopic exploration was considered, but laparotomy was preferred because the laparoscopic approach was not recommended in the Covid-19 pandemic. In addition to the gastric trichobezoar filling the stomach in exploration, there was a perforated area in the proximal jejunum caused by trichobezoar-induced perforation, and in the proximal of the ileum caused by

trichobezoar compression. Similarly, small bowel perforations caused by trichobezoar have been reported in the literature¹⁰. In a 25-series review published by Gok et al, recurrent bezoar cases were reported in 2 patients. They stated that only one patient had gastrotomy and enterotomy simultaneously¹¹. In this case, it is an interesting case in terms of simultaneous gastrotomy and enterotomy. Like this case, in recent case reports, Tripodi et al. demonstrated that two cases of Rapunzel syndrome, they performed both gastrotomy and enterotomy¹². Another case report highlighted a 62-year-old woman underwent emergency surgery due to acute mechanical intestinal obstruction and giant phytobezoars were removed with gastrotomy and jejunotomy¹³. The first and most important step of this disease is to be able to make the correct diagnosis in a timely manner. Detailed clinical history of the patient should be taken and it should be kept in mind in the differential diagnosis as it may be confused with other abdominal diseases due to non-specific complaints. In early cases, it can be managed with conservative methods or endoscopic removal without the need for surgery^{3,11}. As a result, although Rapunzel syndrome is rare, it is known that it can cause multiple perforations in the gastrointestinal tract, which can cause obstruction in any segment. In most cases, conservative management, medical therapy or gastroscopy can be useful in the treatment modality. However, surgery is required in cases where these treatments are unsuccessful and complicated. In addition, patients should be referred to psychiatry for behavioral and medical treatment for underlying psychiatric disorders as well as surgical or medical therapy.

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Riassunto

L'estensione del tricobezoar gastrico (palla di peli) esteso dallo stomaco al tenue rappresenta la sindrome di Rapunzel. Sebbene la si incontri molto raramente, è espressione di una patologia psichiatrica e mentale che si riscontra soprattutto in giovani donne. Si presenta qui il caso di una giovane donna con tricotillomania ricorrente, dell'età di 24 anni.

È stata sottoposta a laparotomia per un trichobezoar ricorrente, che ha riempito completamente lo stomaco e si è esteso all'intestino tenue, ha causato perforazione in due punti dell'intestino tenue, ed è stato rimosso mediante gastrotomia ed enterotomia. Una delle aree perforate era molto vicina al legamento di Treitz e la perforazione si è verificata a causa dell'ulcera causata dal tricobezoario. Dopo un periodo di nutrizione parenterale totale centrale post-operatoria, il passaggio all'alimentazione orale è stata iniziata alla fine della prima settimana. La paziente è stata quindi indirizzata al reparto psichiatrico e trattata con farmaci antipsicotici. È stata dimessa il 9° giorno postoperatorio senza alcun problema.

Conclusione: sebbene non vi siano segni di peritonite in questi casi di sindrome di Rapunzel, è necessario effettuare un'esplorazione dettagliata tenendo presente che il tricoobozoo potrebbe causare ulcerazioni e perforazioni. Inoltre, poiché la causa principale è il disturbo psichiatrico sottostante, i pazienti dovrebbero essere indirizzati alla clinica psichiatrica per prevenire le recidive.

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