CASE REPORT / OLGU SUNUMU

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The Rapunzel Syndrome in a Four-year-old Girl

Dört Yaşındaki Bir Kız Çocuğunda Rapunzel Sendromu

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ABSTRACT

Trichobezoar is a rare pathology in which swallowed hair accumulates in the stomach. An unusual form of bezoar extending from the stomach to the small intestine or beyond is described as Rapunzel syndrome (RS). Trichobezoars typically cause abdominal pain and nausea, but can also present as an asymptomatic abdominal mass, progressing to abdominal obstruction and perforation. Trichobezoar with RS is an uncommon diagnosis. It is predominantly found in emotionally disturbed or mentally retarded young people. The diagnosis may be suspected in young females with abdominal pain, epigastric mass, and malnutrition, who have a history of trichophagia. We present a case of successful laparotomy removal of a gastro-duodenal trichobezoar in a 4-year-old girl with a history of trichotillophagia. The psychodynamic aspects, clinical manifestations, diagnoses, and therapeutic strategies are discussed.

Keywords: Trichobezoar, rapunzel syndrome, child

ÖZ

Trikobezoar, yutulan kılların midede birikmesi sonucu oluşan nadir bir patolojidir. Mideden ince bağırsağa veya ötesine uzanan alışılmadık bir bezoar formu, Rapunzel sendromu (RS) olarak tanımlanır. Trikobezoarlar tipik olarak karın ağrısı ve mide bulantısına neden olur, ancak aynı zamanda obstrüksiyon ve perforasyona ilerleyen asemptomatik bir abdominal kitle olarak da ortaya çıkabilir. Trikobezoarın neden olduğu RS nadir görülen bir tanıdır. Ağırlıklı olarak emosyonel olarak sıkıntılı veya zihinsel engelli gençlerde görülür. Trikofaji öyküsü olan karın ağrısı, epigastrik kitle ve yetersiz beslenme olan genç kadınlarda bu tanıdan şüphelenilebilir. Biz burada trikotillofaji öyküsü olan ve başarılı bir laparotomi ile çıkarılan gastro duodenal trikobezoarlı 4 yaşında kız olguyu psikodinamik yönler, klinik belirtiler, tanı ve tedavi stratejileri açısından tartışarak sunuyoruz.

Anahtar Kelimeler: Trikobezoar, rapunzel sendromu, çocuk

Introduction

A bezoar is a dense mass formed by the non-absorbable ingested materials in the gastrointestinal tract. Based on their compositions, bezoars are classified into phytobezoars (vegetables or fruit fibers), trichobezoars (hair or hair-like fibers), diospyrobezoars (of persimmon), pharmacobezoar (of pills), lactobezoars (of milk and curd), lithobezoars (fragments of stones) or plasticobezoars (plastic).[1] Usually, trichobezoar is confined to the stomach, but rarely tail of hair may extend beyond the stomach to the small intestine and even up to the colon, it is called Rapunzel syndrome (RS).[2] Trichobezoar leading to RS is an extremely rare entity, with about 90 cases reported in the literature.[1] Less than 40 cases of trichobezoar with RS have been reported in the medical literature.[3] It is usually seen in children and young girls predisposed to psychiatric illnesses like trichotillomania and trichophagia or some problem.[4,5] It is diagnosed incidentally while investigating the patient for these symptoms as the history of coexisting psychiatric illness is usually concealed by the patients and the parents. Treatment is the removal of the mass either by upper gastrointestinal endoscopy or by surgery or treatment of the coexisting psychiatric illness, if any.

Case Report

A 4-year-old girl presented to the pediatric gastroenterology department with a history of abdominal pain and recurrent vomiting for 3-4 months. There was a history of coin ingestion when she was two-years old which was removed by endoscopy. She had a history of loss of appetite, gradual loss of weight, early satiety, and a feeling of epigastric fullness. In abdominal examination, there was a firm, mobile, minimally tender lump palpable in the epigastrium. An abdominal X-ray in erect position did not reveal any significant air-fluid levels. The initial diagnosis of trichobezoar was made once her parents admitted to a history of ingestion of hair by the patient. In endoscopy, a large mass of hair in stomach was observed. Surgery was planned due to the size of the mass and the potential for complications. After the operation, she was referred to a child

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and adolescent psychiatrist for evaluation. Parents and child were assessed together. The parents complained about her irritability, withdrawal, and consuming non-food materials (hair, tiny pieces, etc.). She started kindergarten this year and there was an increase in her frustration, withdrawal, and eating non-food items for 6 months. According to the mother, she had never eaten her hair so far but swallowed hair or tiny pieces she observed on the ground from the time she was ambulatory. Therefore, they applied to a child and adolescent psychiatrist with the same symptoms when she was 1,5 years old. The psychiatrist made some recommendations to the parents about the situation and did not initiate any medication.

During her psychiatric evaluation in the playing room, she was often negative, unable to communicate, and to separate from her parents. Also, she was not interested in the toys. Although she received commands and established eye contact, no reciprocal communication with the clinician was established. Based on the Ankara Developmental Screening Inventory her development was on par with chronological age.

According to her mother and teachers, she had not been observed eating her hair in school. Also, she never participated in group activities; instead, she used to play with one of her schoolmates only, and was found shy by her teachers in general. When investigating the stress factors; it became clear that the mother gave birth eight months ago and her communication with the patient was reduced during pregnancy and the post-partum period which also coincided with the patient's initiation of kindergarten.

After the operation, when her requests were not met, she was frustrated and said to her parents ''if you do not do what I want, I will eat hair again''. The symptoms were thought to arise due to problematic relationship with the mother. Because of depressive symptoms the mother reported, she was referred to an adult psychiatrist for a detailed evaluation while treatment for her daughter was initiated in our department. Low dose risperidone (0.25 mg/day) was started along with play therapy sessions. Monthly follow-up of the patient continues at our outpatient clinic.

Discussion

Very few reports of RS among children are documented in the psychiatric literature. This may be due to early referral of most cases of trichotillomania and trichophagia to child and adolescent psychiatrists prior to development of RS.[6] Trichobezoars are most commonly seen in females (approximately 90.0%) in the age group 13-19 years with an unrevealed psychiatric disorder. About 50.0% of these patients are found to be trichophagic. Classically, trichobezoars develop because of trichophagia in mental retardation and psychiatric disorders.[7] However, our patient's intelligence level was found normal. Most trichobezoar cases are accompanied by trichotillomania, a rare psychiatric disorder, which is usually characterized by the urge to pull the hair. Therefore, recent studies suggest that bezoar cases should be evaluated from a psychiatric perspective.[8] The clinical evaluation of our case did not reveal trichotillomania. Trichobezoar is usually seen in adolescent girls and young women. It is associated with psychiatric conditions accompanied by emotional trauma.[8] In 78 cases with pediatric RS, only 5 cases were found to be under 4 years of age.[9] Trichophagia, on the other hand, maybe seen as a clinical reflection of pica. Pica is generally observed in the presence of iron deficiency anemia and childhood neglect.[6] Iron deficiency anemia was not detected in our study. However, considering factors such as sibling birth and mother's depressive appearance, it was thought that the patient could not receive adequate parental warmth.

Since trichobezoars are often associated with underlying psychiatric disorders, the evaluation of these patients by psychiatrists is critical. Considering the risk of recurrence of trichobezoar, medical treatment and behavioral therapies for these patients should be arranged appropriately and their follow-up should be performed at close intervals.

Ethics

Informed Consent: We obtained a written consent form from the patient and her parents.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: L.C., Concept: S.A.S., L.C., A.U.Ç., Design: S.A.S., L.C., A.U.Ç., Data Collection or Processing: S.A.S., L.C., A.U.Ç., Analysis or Interpretation: S.A.S., L.C., A.U.Ç., Literature Search: S.A.S., L.C., A.U.Ç., Writing: S.A.S.

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References

1. Sehgal VN, Srivastava G. Trichotillomania +/- trichobezoar: revisited. J Eur Acad Dermatol Venereol. 2006;20:911-915.
2. Nirasawa Y, Mori T, Ito Y, Tanaka H, Seki N, Atomi Y. Laparoscopic removal of a large gastric trichobezoar. J Pediatr Surg. 1998;33:663-665.
3. Iwamuro M, Okada H, Matsueda K, Inaba T, Kusumoto C, Imagawa A, Yamamoto K. Review of the diagnosis and management of gastrointestinal bezoars. World J Gastrointest Endosc. 2015;7:336-345.
4. Ripolles T, Garcia-Aguayo J, Martinez MJ, Gill P. Gastrointestinal bezoars: sono-graphic and CT characteristics. AJR Am J Roentgenol. 2001;177:65-69.
5. Alsafwah S, Alzein M. Small bowel obst-ruction due to trichobezoar: Role upper endoscopy in diagnosis. Gastrointest Endosc. 2000;52:784-786.
6. Islek A, Sayar E, Yılmaz A, Boneval C, Artan R. A rare outcome of iron deficiency and pica: Rapunzel syndrome in a 5-year-old child. Turk J Gastroenterol. 2014;25:100-102.
7. Gonuguntla V, Joshi DD. Rapunzel syndrome: a comprehensive review of an unusual case of trichobezoar. Clin Med Res. 2009;7:99-102.
8. Grant JE, Odlaug BL. Clinical characteristics of trichotillomania with trichophagia. Compr Psychiatry. 2008;49:579-584.
9. Phillips MR, Zaheer S, Drugas GT. Gastric trichobezoar: case report and literature review. Mayo Clin Proc. 1998;73:653-656.