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CASE REPORT

Rapunzel Syndrome – A Rare Form of Trichobezoar: A Case Report

Thayumanavan L, Rajkumar K, Kannan M.

Department of Medical & Surgical Gastroenterology, Vadamalayan Hospitals Madurai, Tamilnadu, India

ABSTRACT

Hair ball in the GI tract is called trichobezoar and the Rapunzel syndrome is a very rare form of trichobezoar found in patients with psychiatric illness in whom the ingested hair extends from the stomach into the small intestine. Trichotillomania (hair pulling) and trichophagia (chewing of hair) are intrinsic to the development of Rapunzel syndrome. A small number of patients have been reported in whom the gastric trichobezoar has a long tail and extends throughout the small bowel to the cecum. This condition, known as the Rapunzel syndrome, occurs almost exclusively in young girls. We report a case Rapunzel syndrome who presented with history of upper GI obstruction and weight loss. Upper GI endoscopy revealed a large trichobezoar occupying fundus of stomach extending along the whole stomach into duodenum beyond its 3rd part. It was treated successfully with surgery followed by psychiatric consultation. We will review the literature on the subject as well. (*J Dig Endosc* 2013;4(1):19-21)

Key Words: Trichobezoar – Trichillotomania – Trichophagia – Psychiatric disorder – Upper GI obstruction – Surgery

Introduction

The origin of name of Rapunzel syndrome is a rare condition caused from eating hair and its name is based after the long-haired girl Rapunzel in a fairy tale almost three centuries ago. The first report of Rapunzel syndrome consisting of two cases was in 1968 by Vaughan *et al.*[1] Among the bezoars seen in the GI tract of animals and humans, phytobezoars are more common.[2] Between 1968-2006, 27 cases of Rapunzel syndrome have been reported.[3] A few other reports have been published since then.[4-6] Rapunzel syndrome represents tirchotillomania and trichophagia that results in the formation of a gastric trichobezoar extending from the fundus of the stomach beyond the duodenum as found in the case recorded here.

Case Report

A young 17 year old girl was evaluated for pain epigastrium, vomiting and weight loss for one year. She has a history of hair pulling from her frontal forehead from a very young age and had been reprimanded by her parents and teachers. Physical examination revealed loss of hair in the frontal forehead with bald patches. Abnormal laboratory investigations included low hemoglobin, 9.2gms/dL; and serum albumin, 2.6g/dL. The other blood biochemistry indices were normal. An upper gastrointestinal (UGI) endoscopy revealed a large trichobezoar extending from the fundus across the pylorus to beyond the 3rd part of duodenum findings compatible with Rapunzel's syndrome (Figures 1, 2 and 3). Surgery was initiated with laparoscopy which was converted to open laparotomy due to a very large intra gastric mass of hair extending to the third part of duodenum. Gastrotomy was performed to facilitate removal of the 60-cm long hair ball extending to the small Intestine in the shape of the stomach (Figure 4). Patient was discharged on the fifth part of day and is on follow up.

Reprints requests and correspondence:
Dr. L. Thayumanavan M.D.,DM.,
Vadamalayan Hospitals, 9-A Chokkikulam
Madurai, Tamilnadu, India- 625002
Email: drthayu2@rediffmail.com

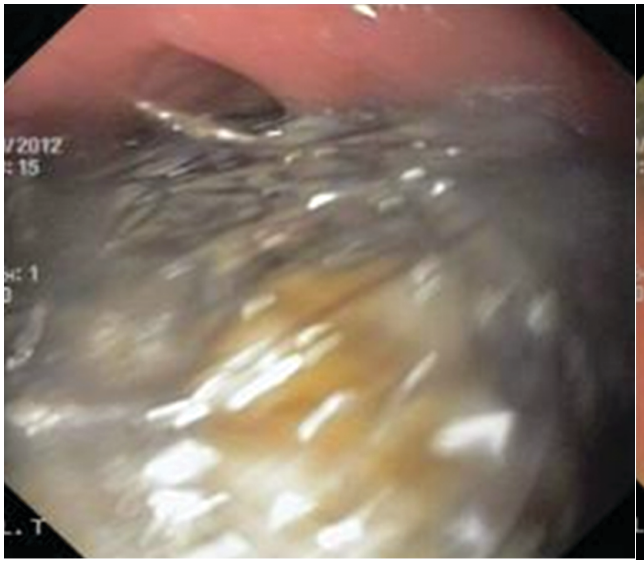

Figure 1: Hair ball in the fundus of stomach

Figure 2: Hair ball extending beyond the pylorous

Figure 3: Hair ball extending deep in the duodenum

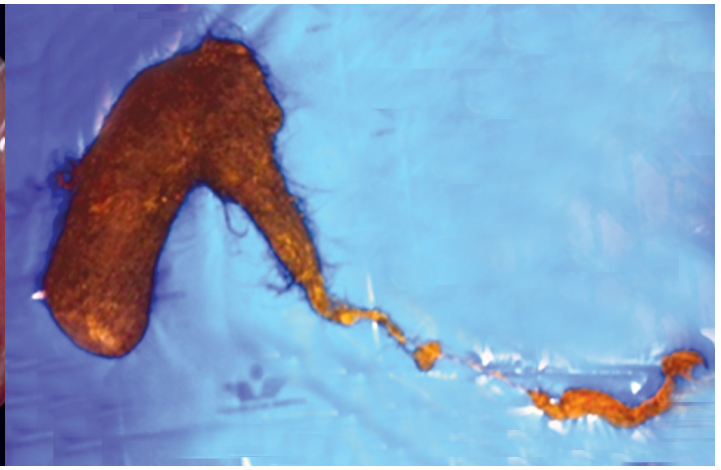

Figure 4: Operated Specimen

Discussion

Rapunzel syndrome occurs in young ladies with psychiatric disorders.[3,7] Plucking of hair from the frontal region and its chewing is often available in the patient's history and also evidenced by small bald patches in frontal scalp.[8] Bezoars grow by the continuing ingestion of food rich in cellulose and other indigestible materials such as hair, cotton, and tissue paper, matted together by protein, mucus, and pectin. Properties of the specific ingested material and some degree of gastric dysfunction also contribute bezoar formation.

It was recognized in 1938 that most patients with bezoars had undergone gastric surgery, implying that there are underlying anatomic and functional abnormalities.[2] More recent studies have shown that 70 to 94 percent of patients have a history of gastric surgery and 54 to 80 percent have undergone vagotomy and pyloroplasty.[9,10] It was initially thought that the most common functional abnormality found in patients with bezoars was delayed gastric emptying. However, some studies have found that many patients have normal or accelerated gastric emptying. In one series gastric emptying was studied in 10 patients who presented with bezoars 1 to 20 years after some type of gastric surgery and in operated patients without bezoars,[11] There were no differences between the two groups in gastric emptying of Tc 99m-labeled solids at 45, 75, and 105 minutes. This implies that the pathogenesis of bezoar formation is more complex than initially thought and involves other factors such as alterations in the production of acid, pepsin, and mucus, and impairments in the grinding mechanism and the inter-digestive migrating motor complex.[12]

The diagnosis was confirmed by endoscopy which showed the hair ball extending beyond the 3rd part of duodenum. Bowel invagination is seen in 7% of Rapunzel syndrome[3] but was not seen in our case. Obstruction to the bowel is the most common clinical manifestation.[4] Necrosis,

perforation, bleeding and intussusceptions have been reported.[13]

Mechanical fragmentation, chemical substances to dissolve small trichobezoars are options apart from surgery or endoscopic removal.[5,6] Surgery and removal of the long trichochobezoar through gastrostomy is the procedure described as standard of care. We attempted laparoscopic removal of bezoar but its enormous length and size prevented its removal and laparotomy was performed to remove the hair ball .

Psychiatric assessment will form an integral form of treatment as recurrences have been described.[8] Follow-up is needed and usually the fear of repeat surgery prevents the patients from swallowing the hair again.

Conclusions

Rapunzel syndrome is an extremely rare presentation of trichobezoar seen in young patients detected by gastroscopy, ultrasound and CT. Complications like vomiting, anemia, bowel obstruction, perforation and necrosis are seen. But death is rare. Surgery is the standard of care for extraction of large trichobezoar in Rupenzel syndrome. Psychiatric consultation is essential for prevention of recurrence.

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