International Surgery Journal
Modh FA. Int Surg J. 2018 Nov;5(11):3764-3766
http://www.ijsurgery.com
pISSN 2349-3305 | eISSN 2349-2902

DOI: http://dx.doi.org/10.18203/2349-2902.isj20184661

Case Report

Rapunzel syndrome: a review of unusual case

Foram Arvindbhai Modh*

Department of General surgery, Surat Municipal Institute of Medical Education and Research, Surat, India

Received: 22 August 2018
Accepted: 27 September 2018

***Correspondence:**
Dr. Foram Arvindbhai Modh,
E-mail: modhforam@yahoo.com

ABSTRACT

A rare condition characterized by trichophagia, or the swallowing of one's own own hair, leading to a solid mass that forms in the gastrointestinal tract. The resulting blockage of the gastrointestinal tract may be fatal. The hair forms a solid mass in the digestive tract known as a trichobezoar. It causes abdominal pain and nausea, but can also present as an asymptomatic abdominal mass, progressing to abdominal obstruction and perforation. It is predominantly found in emotionally disturbed or mentally retarded youngsters. We present the youngest case of Rapunzel syndrome in the Surat Municipal Institute of Medical Education and Research Hospital, Surat, India, a 7-year-old girl who present with abdominal pain and non-tender abdominal mass.

Keywords: Abdominal mass, Trichophagia, Trichobezoar, Rapunzel syndrome

INTRODUCTION

Bezoars are concretions of human or vegetable fibers that accumulate in the gastrointestinal tract. The word "bezoar" comes from the Arabic word "bedzehr" meaning "protecting against a poison".[1] At different times in history, bezoars from animal guts were used as precious stones, antidotes to poison and today as part of traditional Chinese medicine.[2] The first reference to a bezoar in a human was in 1779 during an autopsy of a patient who died from gastric perforation and peritonitis.[3]

The Most common presentation of trichobezoar is small bowel obstruction (SBO) but a rare and sometimes bizarre presentation is reported worldwide.[4] In human, the most common type of bezoar, which is mostly made of hair. However, bezoars can also be made of vegetable or fruit fiber (phytobezoars), milk curd (lactobezoars), or any indigestible materials.

The name "Rapunzel" syndrome comes from the Grimm Brothers' fairy tale of a 12-year-old princess who was shut into a tower with neither stairs nor doors by an enchantress who climbed up the tower's walls with the help of Rapunzel's long tresses.[5] The first reference to a bezoar in a human was in 1779 during an autopsy of a patient who died from gastric perforation and peritonitis.[6]

We present the youngest documented patient with Rapunzel syndrome in the SMIMER Hospital, Surat, India.

CASE REPORT

A 7-year old female, born at 36 weeks gestation with normal labor, presented with a 10-week history of poorly localized abdominal pain. The child appeared normal in behavior. The mother commented on self-hair eating habit since 3 month but not baldness. Her child had early satiety and decreased appetite.no complain of vomiting, fever, constipation and acid reflux. There were no changes in her bowel habits. The child had always been lower end of growth curve with underweight compared to height. Abdominal examination revealed a hard, non-tender, ballotable mass of approximately 8 cm x 6 cm in epigastric region extending in the right upper quadrant.

The rest of the physical examination was normal. Laboratory work-up complete blood count, complete metabolic panel were normal.

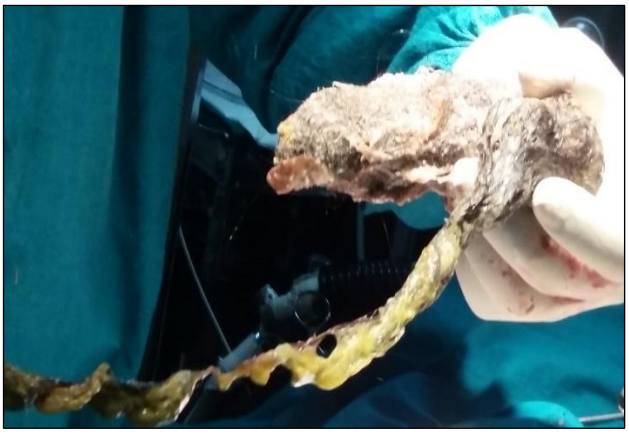

Figure 1: 13cm x 7cm x 5cm mass was found to be a trichobezoar.

Ultrasonography report suggesting of over distended stomach and content shows posterior acoustic shadow. Rest bowel loops grossly normal. Gastrostomy was performed the next day and the mass removed in one piece. The 13cm x 7cm x 5cm mass was found to be a trichobezoar with a tapering tail extending into small bowel and was perfect cast of the stomach, pylorus and duodenal bulb (figure 1). The patient was discharged home 5 days later, having recovered without complications. Psychiatric follow-up was arranged, where she showed improvement in behavior, which is key role to prevent recurrence.

DISCUSSION

The term bezoar comes from the Arabic "badzehr", meaning antidote. Although there are not many articles in the literature concerning bezoars, many are presumed to go unreported. The etiology of bezoar formation is usually the ingestion of indigestible materials, especially by mentally retarded or psychiatric patients. Gastric dysmotility may be another factor.[5] Most cases involve trichobezoars in young females with trichotillomania, reported in India. Other rare forms include phytobezoars (vegetable fibers), lactobezoars (milk products), pharmacobezoars (drugs) or diospyrobezoars (persimmon fibers). Cotton bezoars are also rare, resulting from the ingestion of strings unraveled from clothes, especially by mentally retarded patients. The strings coil up and mix with mucus and food particles to form a gastric bezoar, and the long strings extend down to the duodenum and intestines, leading to Rapunzel syndrome.[6]

Trichotillomania results in highly variable patterns of hair loss, ranging from small undetectable patches of hair loss to total baldness and because of ingested hair accumulation, it may present with different kinds of GI problems. Not all cases of trichotillomania have trichophagia, also all of patients with trichophagia will not have trichobezoars.[7] The mass of a bezoar can lead to epigastric pain due to distention and ulceration, early satiety and loss of weight. In cases of Rapunzel syndrome, a tail present in the intestines can trigger peristaltic movements, resulting in colicky abdominal pain. Gastrointestinal obstruction, bleeding and perforation are rare complications.[8] Any suspicion of the above symptoms, especially in mentally retarded or psychiatric patients, should alert physicians to perform an endoscopy, as complications can be life-threatening. The hyperammonemia coma state in the patient was due to portal vein thrombosis in a non-cirrhotic condition, and episodic type B hepatic encephalopathy has been described in portal-systemic shunts.[9]

Affected patients occasionally remain asymptomatic for many years. Symptoms develop as the bezoar increases in size to the point of obstruction. Not surprisingly, most of the cases have been reported in countries where women traditionally have long hair. The most common presenting signs are abdominal pain, nausea and vomiting, obstruction, and peritonitis. Less commonly, patients have presented with weight loss, anorexia, hematemesis and intussusception. Complications by a large eroding or obstructing bezoar additionally include gastric ulceration, obstructive jaundice, acute pancreatitis and gastric emphysema.[10,11] Other malabsorption-related complications include protein-losing enteropathy, iron deficiency, and megaloblastic anemia.[12]

Treatment for symptomatic gatorboard is usually surgical intervention but some advocate primary non-surgical attempts such as transendoscopic fragmentation and extraction or transendoscopic lytic enzyme administration although the success of these methods depends on the size of trichobezoar and its duration of formation. Older trichobezoar is much harder to treat with transendoscopic lytic enzyme administration. Surgical treatment of bezoars is their removal, either by enterotomy or by endoscopy, if possible.

Large bezoars can be difficult to remove in a one session endoscopy, so fragmentations and removal may be attempted. Enzymatic fragmentation with acetylcysteine may be helpful during endoscopic removal.[8] Phytobezoars are more sensitive to pharmacological degradation than trichobezoars. Cellulase, papain, acetylcysteine and carbonated beverages are reported pharmacotherapies for bezoars.[13]

Case reports of children with trichobezoars or Rapunzel syndrome are rare, and many links the trichophagia to early childhood neglect or abuse, psychiatric conditions, mental retardation or bereavement.[14,15] Although studies of the pharmacotherapy of trichotillomania remain inconsistent, some patients seem to respond to fluoxetine or other serotonin reuptake inhibitors.[16] Parental counseling is also a regular part of treatment to prevent recurrence. The patient's long-term prognosis is excellent

if behavioral therapy is used to control trichophagia, and psychological/psychiatric follow-up is maintained.

CONCLUSION

In conclusion, this case report is relevant as it clearly describes important clinical lessons learned from the psychological and surgical management of a case of Rapunzel syndrome which, to our knowledge, represents the longest published interval between initial treatment and presentation with relapse of the condition.

The key message is that although surgery is the initial treatment, a comprehensive and long-term postoperative psychiatric follow-up is needed in patients with Rapunzel Syndrome as a late relapse is possible. Multidisciplinary health care teams headed by a psychiatrist as well as family support play a key role in the prevention of recurrence. It is hoped that our shared experience will inform the management of similar cases.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Dart RC. Medical toxicology. 3rd edition, Philadelphia: Lippincott, Williams and Wilkins; 2004:1301.
2. Wang C, Zhao X, Mao S, Wang Y, Cui X. Management of SAH with traditional Chinese medicine in China. Neurol Res. 2006;28:436-44.
3. Vaughan ED, Sawyers JL, Scott HW. The Rapunzel syndrome. An unusual complication of intestinal bezoar. Surgery. 1968;63:339-43.
4. Ousadden A, Mazaz K, Mellouki I, Taleb KA. Gastric trichobezoar: one case report. Ann Chir. Philadelphia: Lippincott, Williams and Wilkins, 2004;129: 237-40.
5. Grimm Brothers: Rapunzel. Translated by Godwin-Jones R. Richmond, Virginia Commonwealth University Department of Foreign Languages, 1994-1999.
6. Naik S, Gupta V, Naik S, Rangole A, Chaudhary AK, Jain P. Rapunzel syndrome reviewed and redefined. Dig Surg. 2007;24:157-61.
7. Singh G, Mitra SK. Gastric perforation secondary to recurrent trichobezoar. Indian J Pediatr. 1996;63(5):689-91.
8. Goldstein SS, Lewis JH, Rothstein R. Intestinal obstruction due to bezoars. Am J Gastroenterol. 1984;79:313-8.
9. Jiledar Singh G, Mitra SK. Gastric perforation secondary to recurrent trichobezoar. Indian J Pediatr. 1996;63:689-91.
10. Klipfel AA, Kessler E, Schein M. Rapunzel syndrome causing gastric emphysema and small bowel obstruction. Surg. 2003;133:120-1.
11. Wang YG, Seitz U, Li ZL, Soehendra N, Qiao XA. Endoscopic management of huge bezoars. Endoscopy. 1998;30:371-4.
12. Ferenci P, Lockwood AH, Mullen KD, Tarter RE, Weissenborn K, Blei AT. Hepatic encephalopathy: definition, nomenclature, diagnosis, and quantification: final report of the working party at the 11th World Congress of Gastroenterol, Vienna. Hepatol. 2002;35:716-21.
13. Walker-Renard P. Update on the medicinal management of phytobezoars. Am J Gastroenterol. 1993;88:1663-6.
14. Gockel I, Gaedertz C, Hain HJ, Winckelmann U, Albani MI, Lorenz D. The Rapunzel syndrome: rare manifestation of a trichobezoar of the upper gastrointestinal tract. Chinurg. 2003;74:753-6.
15. Sharma NL, Sharma RC, Mahajan VK, Sharma RC, Chauhan D, Sharma AK. Trichotillomania and trichophagia leading to trichobezoar. J Dermatol. 2000;27:24-6.
16. Swedo SE, Leonard HL, Rapoport JL, Lenane MC, Goldberger EL, Cheslow DL. A double-blind comparison of clomipramine and desipramine in the treatment of trichotillomania (hair pulling). N Engl J Med. 1989;321:497-501.

Cite this article as: Modh FA. Rapunzel syndrome: a review of unusual case. Int Surg J 2018;5:3764-6.