Rapunzel syndrome due to ingested hair extensions: Surgical and psychiatric considerations

Devin C. Flaherty a,*, Francis Aguilar b, Basant Pradhan b, Harsh Grewal a

a Cooper University Hospital, Department of Pediatric Surgery, Three Cooper Plaza, Suite 403, Camden, NJ 08103, USA
b Cooper University Hospital, Department of Psychiatry, Three Cooper Plaza, Suite 307, Camden, NJ 08103, USA
c Rowan University School of Osteopathic Medicine, Department of General Surgery, One Medical Center Drive, Stratford, NJ 08084, USA

A R T I C L E   I N F O

Article history:
Received 3 September 2015
Received in revised form 6 November 2015
Accepted 11 November 2015
Available online 19 November 2015

Keywords:
- Trichotillomania
- Trichophagia
- Trichobezoar
- Obstruction
- Rapunzel syndrome

A B S T R A C T

INTRODUCTION: Rapunzel syndrome is a unique clinical manifestation of trichotillomania and trichophagia. The resulting gastric trichobezoar can be massive and necessitate surgical extraction.

PRESENTATION OF CASE: We present a case involving a 15 year-old female with a known history of trichotillomania. The patient possessed symptoms of nausea and early satiety, and admitted that she had recently consumed a large number of hair extensions. Computed tomography imaging revealed a massive gastric bezoar not amenable to endoscopic extraction. The patient underwent surgical laparotomy, and a large gastric trichobezoar was removed through an anterior gastrotomy. The trichobezoar extended past the pylorus into the duodenum, thus confirming a diagnosis of Rapunzel syndrome.

DISCUSSION: Trichobezoars formed from the consumption of hair extensions is a rare cause of Rapunzel syndrome. Surgical extraction is often necessitated due to the sheer size of the gastric bezoar that results from consuming hair extensions. If not previously established, psychiatric consultation should be pursued in the immediate post-operative course as these patients may require pharmacologic management along with behavioral therapy to avoid further episodes of trichotillomania.

CONCLUSION: This case presents a unique and modern manifestation of Rapunzel syndrome. Surgical treatment most often is required when a patient presents with a massive gastric trichobezoar. Regular post-operative psychiatric follow-up is necessary to prevent recurrent episodes.

© 2015 The Authors. Published by Elsevier Ltd. on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

“Rapunzel, Rapunzel!
let down your hair!”

Jacob and Wilhelm Grimm, “Rapunzel” [1]

1. Introduction

Trichobezoars are collections of hair that can accumulate in the gastrointestinal tract as a result of trichophagia, or mouthing of hair [2]. A trichobezoar extending from the stomach to the small intestine or beyond is termed Rapunzel syndrome. Initially described by Vaughn et al. [3], the diagnosis of Rapunzel syndrome is loosely based on symptoms of gastrointestinal obstruction secondary to suspected gastric trichobezoar with post-pyloric extension of a “tail” distally into the small intestines and confirmed after surgical extraction [4,5]. The term itself is derived from the popular fairy tale, “Rapunzel,” which describes a 12-year-old princess who uses her long hair to help a prince scale the tower wherein she is impris-

oned [1]. Gastric trichobezoar has the ability to cause gastric outlet obstruction, a surgical emergency [6].

2. Presentation of case

A 15 year-old girl experiencing early satiety with intermittent nausea and vomiting was evaluated in the outpatient setting. Four months prior to presentation the patient had been diagnosed with trichotillomania after surgical extraction of a trichobezoar that was obstructing the patient's distal small bowel. The patient did admit she had recently consumed hair extensions, eating them directly from the packaging they came in. An abdominal and pelvis computed tomography scan was obtained and confirmed the diagnosis of gastric bezoar (Fig. 1). As the patient was not clinically or radiographically obstructed, an outpatient upper endoscopy was performed and revealed a massive gastric trichobezoar composed mostly of hair that extended from the gastroesophageal junction to the duodenum. The size of the mass precluded endoscopic extraction, therefore a surgical consultation was pursued. Upon evaluation in the surgical clinic the patient was not in any discomfort. On physical exam her abdomen was distended and a palpable mass extended from her left subcostal margin inferiorly to her umbilicus. Subsequently, the patient underwent
an elective abdominal exploration through a 5-cm upper midline incision. After entering the abdomen and confirming the diagnosis, a 5-cm gastrotomy was performed and the foreign body was visualized (Fig. 2A), grasped, carefully extracted from the stomach and duodenum, and removed from the abdomen through a wound protection device (Fig. 2B). Gross pathologic examination of the specimen revealed a 20 × 12 × 4 cm compressed solid mass composed entirely of black hair extensions. The mass was cast in a shape that mimicked the contours of the stomach and duodenum. Hair was noted to extend 20 cm distally from the body of the specimen, thus establishing the diagnosis of Rapunzel syndrome (Fig. 3). Post-operatively, the patient was without nausea or vomiting and her nasogastric tube was removed on day 1. Her diet was slowly advanced and she was discharged home on post-operative day 4. While in the hospital the psychiatry service did see the patient in consultation. In order to avoid further relapses, close psychiatric follow-up was confirmed by care-coordinators and the psychiatry service with the patient and her guardian upon discharge. In the outpatient setting, the patient was pharmacologically managed with 10 mg of Prozac daily. Attempts were also made to initiate cognitive behavioral therapy.

3. Discussion

The case presented illustrates a novel clinical manifestation of Rapunzel syndrome. Trichobezoar formation through ingestion of hair extensions has previously been reported twice [7,8], and, to our knowledge, consuming hair extensions directly from the packaging has never been reported. A review of 27 cases of Rapunzel syndrome in 2007 by Naik et al. revealed the majority of patients diagnosed are females under the age of 20 [4]. The patient presented does conform to these demographics. In 2013, Fallon et al. reported the largest single institution case series to date regarding trichobezoar. All patients reported were female, and all required surgical laparotomy to accomplish extraction of the trichobezoar [9]. While laparotomy has traditionally been the accepted method of extraction due to the sheer size of these bezoars, laparoscopic techniques have been recently described in the literature [10]. Complications such as obstruction, perforation, intussusception, or giant gastric ulceration may arise if Rapunzel syndrome is left untreated [11–13].

The patient’s psychiatric course was closely followed in the hospital and post-operatively in the outpatient setting. According to the American Psychiatric Association’s Diagnostic and Statistical Manual of Mental Disorders, Fifth Edition (DSM-5), trichotillomania is categorized as an obsessive-compulsive behavior [2,14]. Approximately 0.6–3.6% of the population of the United States suffers from trichotillomania [15,16]. Pharmacologic treatment with agents such as quetiapine may help control the obsessive behavior exhibited by patients who ingest hair extensions [8]. The patient in our case report revealed that prior to consuming the hair extensions she was experiencing many stressors in her home environment. Her parents were separated, and at the time of care she was living with her maternal grandparents. The patient chose to eat hair extensions because it helped temper her anxiety. She denied skin picking, pulling out her own hair, or eating other objects such as dirt or clay. In a similar report of Rapunzel syndrome, Jones et al. described the role of habit-reversal training in the post-operative period through behavioral therapy in the setting of a supportive family environment [8]. Other therapies such as cognitive therapy, and acceptance and commitment therapy can support behavioral therapy in the treatment of this often difficult to manage condition [17].

4. Conclusion

Rapunzel syndrome represents a surgical diagnosis upon extraction of a retained bezoar of hair that extends past the pylorus into the small bowel. Close psychiatric follow-up is mandated to avoid recurrence as trichotillomania is a chronic compulsive behavior.

Conflicts of interest

The authors have no financial and personal relationships with other people or organizations that could inappropriately influence (bias) this submission.

Funding

The authors have no extra or intra-institutional funding to declare.

Ethical approval

None.

Consent

Written informed consent was obtained from the patient’s legal guardian for publication of this case report and its accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Devin C. Flaherty, Harsh Grewal, Francis Aguilar and Basant Pradhan all contributed to the writing and editing of the case report. Devin C. Flaherty and Harsh Grewal were involved in the literature review.
Harsh Grewal and Devin C. Flaherty provided surgical care of the patient during her treatment.
Basant Pradhan and Francis Aguilar have provided long-term psychiatric follow-up.
Surgical images were courtesy of the Pediatric Surgery Department with the patient’s consent.

Guarantor
Harsh Grewal.

References

1. J.L.C. Grimm, W.C. Grimm, Grimm's Fairy Tales, Wordsworth Editions Limited, Hertfordshire, 1993.
2. American Psychiatric A, Force DSMT. Diagnostic and statistical manual of mental disorders: DSM-5. 2013; Available from: http://dsm.psychiatryonline.org/book.aspx?bookid=556.
3. E.D. Vaughan Jr., L. Sawyers, H.W. Scott Jr., The Rapunzel syndrome. An unusual complication of intestinal bezoar, Surgery 63 (2) (1968) 339–343.
4. S. Naik, V. Gupta, S. Naik, A. Rangole, A.K. Chaudhary, P. Jain, et al., Rapunzel syndrome reviewed and redefined, Dig. Surg. 24 (3) (2007) 157–161.
5. A.R. Jensen, C.T. Trankiem, S. Lebovitch, H. Grewal, Gastric outlet obstruction secondary to a large trichobezoar, J. Pediatr. Surg. 40 (8) (2005) 1364–1365.
6. K. Salaam, J. Carr, H. Grewal, E. Sholevar, D. Baron, Untreated trichotillomania and trichophagia: surgical emergency in a teenage girl, Psychosomatics 46 (4) (2005) 362–366.
7. W.T. Kwong, D. Kalmaz, A modern form of Rapunzel syndrome: trichobezoar composed of synthetic hair extensions, Clin. Gastroenterol. Hepatol. 12 (5) (2014) A33–A34.
8. G.C. Jones, K. Coutinho, D. Anjaria, N. Hussain, R. Dholakia, Treatment of recurrent Rapunzel syndrome and trichotillomania: case report and literature review, Psychosomatics 51 (5) (2010) 443–446.
9. S.C. Fallon, B.J. Slater, E.L. Larimer, M.L. Brandt, M.E. Lopez, The surgical management of Rapunzel syndrome: a case series and literature review, J. Pediatr. Surg. 48 (4) (2013) 830–834.
10. S.N. Jatal, N.P. Jamadar, B. Jadhav, S. Siddiqui, S.B. Ingle, Extremely unusual case of gastrointestinal trichobezoar, World J. Clin. Cases 3 (5) (2015) 466–469.
11. N. Pul, M. Pul, The Rapunzel syndrome (trichobezoar) causing gastric perforation in a child: a case report, Eur. J. Pediatr. 155 (1) (1996) 18–19.
12. S. Marwah, S. Pandey, A. Raj, M.S. Jangra, H. Sharma, Rapunzel syndrome presenting as jejuno-jejunal intussusception, Clin. J. Gastroenterol. 8 (4) (2015) 202–206.
13. A. Athanasiou, A. Michalinos, D. Moris, E. Spartalis, N. Dimitrokaliss, V. Kaminitis, et al., Rapunzel syndrome: a rare presentation with giant gastric ulcer, Case Rep. Med. 2014 (2014), 267319.
14. American Psychiatric Association: Diagnostic and Statistical Manual of mental Disorders. 2013.
15. B.W. Woods, D.C. Houghton, Diagnosis, evaluation, and management of trichotillomania, Psychiatric Clin. North Am. 37 (3) (2014) 301–317.
16. G.A. Christenson, R.L. Pyle, J.E. Mitchell, Estimated lifetime prevalence of trichotillomania in college students, J. Clin. Psychiatry. 52 (10) (1991) 415–417.
17. I. Snorras, G.S. Berlin, H.J. Lee, Optimizing psychological interventions for trichotillomania [hair-pulling disorder]; an update on current empirical status, Psychol. Res. Behav. Manag. 8 (2015) 105–113.

Open Access
This article is published Open Access at sciencedirect.com. It is distributed under the IJSCR Supplemental terms and conditions, which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.