CASE REPORT – OPEN ACCESS

International Journal of Surgery Case Reports 40 (2017) 23–26

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Trichobezoar with and without Rapunzel syndrome in paediatric population: A case series from a tertiary care centre of Northern India

Pradeep Kajal[a,*], Namita Bhutani[b], Niharika Tyagi[c], Pratibha Arya[c]

[a] Deptt. of Pediatric Surgery, PGIMS, Rohtak, Haryana, India
[b] Deptt. of Pathology, PGIMS, Rohtak, Haryana, India
[c] Deptt. of Pediatrics, PGIMS, Rohtak, Haryana, India

ARTICLE INFO

Article history:
Received 17 July 2017
Received in revised form 22 August 2017
Accepted 23 August 2017
Available online 8 September 2017

Keywords:
Case series
Trichobezoar
Rapunzel syndrome
Trichotillomania
Trichophagia

ABSTRACT

INTRODUCTION: Trichobezoars are concretions formed by accumulation of hair in stomach. Usually, trichobezoar is confined to the stomach, but rarely it may extend from the stomach to the small intestine and even colon. This is an unusual form called Rapunzel syndrome. Our experience with this rare entity of Rapunzel syndrome and interesting entity of trichobezoar is being presented with review of literature.
PRESENTATION OF CASES: We, at our institute, encountered four cases of trichobezoar in last five years, out of which two were found to be of Rapunzel syndrome. All of these cases were managed successfully by open surgical intervention in view of the very large size of the mass in all the cases.
DISCUSSION: The clinical presentation is highly variable ranging from asymptomatic cases diagnosed incidently to serious gastrointestinal symptoms and complications. Cases of trichobezoar have been reported in literature very infrequently but Rapunzel syndrome is extremely rare and less than 50 cases have been reported in medical literature till date.
CONCLUSION: Trichobezoar leading to Rapunzel syndrome is an extremely rare entity. The clinical presentation is usually vague and non-specific. Treatment is mainly surgical because of delayed presentation in majority of the cases. Psychiatric illness is the usual association.

1. Introduction

A bezoar is the dense mass formed by the non absorbable ingested materials in gastrointestinal tract. Based on their compositions, bezoars are classified into phytobezoars (composed of vegetables or fruit fibres), trichobezoars (balls of hair or hair-like fibres), diospyrobezoars (of persimmon), pharmacobezoar (of pills), lactobezoars (of milk and curd), lithobezoars (fragments of stones) or plasticobezoars (plastic) [1]. Rapunzel syndrome is an extremely rare variant of trichobezoar in which there is a tailed extension of this mass that reaches upto duodenum, jejunum, ileum and even upto colon. It is usually seen in children and young girls predisposed with psychiatric illness like trichotillomania (practice of pulling hair) and trichophagia (habitual ingestion of hair) or some social problem [2,3]. The patients usually present with abdominal pain, nausea, vomiting, early satiety, loss of appetite and even symptoms of intestinal obstruction and peritonitis in advanced stages [4,5]. It is diagnosed incidently 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 removal of the mass either by upper gastrointestinal endoscopy, laparoscopy or by open surgery and treatment of the coexisting psychiatric illness, if any.

2. Clinical presentation

2.1. Case 1 & 2: cases of trichobezoar with Rapunzel syndrome

Two female patients aged 12 years and 4 years presented to our Accident and Emergency department at different time intervals about 1 year apart with complaints of persistent vomiting for last two days and 3 days respectively. The vomiting was non bilious to begin with but then turned to bilious after one day in both the patients. There was history of similar episodes of vomiting off and on for last one and a half years and 6 months respectively and contained whatever the patients had with some semidigested material most of the times. Both the patients gave history of loss of appetite, gradual loss of weight, early satiety and feeling of epigastric fullness. On per abdomen examination, there was a large, firm, mobile, minimally tender lump palpable in left hypochondrium and epigastrium in both the patients (Fig. 1). The patients were admitted and dehydration and dyselectrolytemia were corrected. Nasogas-

* Corresponding author at: 23/8 FM, Medical Campus, Rohtak, Haryana 124001, India.
E-mail addresses: pkajalsurg@gmail.com, drpradeepkajal@gmail.com (P. Kajal), namitabhutani89@gmail.com (N. Bhutani), ntyagi863@gmail.com (N. Tyagi), pratibhaarya1996@gmail.com (P. Arya).

http://dx.doi.org/10.1016/j.ijscr.2017.08.060

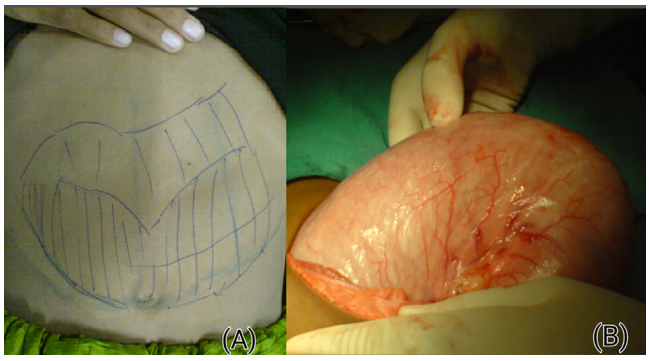


Fig. 1. Peroperative photograph showing a large abdominal lump (marked with pen) reaching upto the umbilicus (A) and the large firm mass in stomach filling its lumen completely (B).

tric tube was inserted in view of vomiting but surprisingly drained very minimal gastic type of effluent in both the patients. Abdominal skiagrams did not reveal any significant air-fluid levels. On abdominal ultrasonography, there was a large mass filling whole of the lumen of the stomach completely and pushing the nasogastric tube to one side. So, a provisional diagnosis of trichobezoar was made though none of the patients or their parents admitted to history of ingestion of hair by the patients. Upper gastrointestinal endoscopy could not be done as it was not available in the emergency set up. So, the patients were taken up for surgery in view of definite surgical pathology, large size of the mass and potential for complications in both the patients. On opening the abdomen by right transverse supraumbilical incision, the lumen of the stomach was found to be completely occupied by a slightly indentable large firm mass (Fig. 1). The stomach was opened on the anterior aspect of the body with electrocautery and its lumen was found to be filled completely by a huge mass made up of hair interspersed with air, undigested food and vegetable material conforming to the shape of the stomach. On trying to remove the mass, it was found to have a tailed hairy extension into duodenum. Therefore, an operative diagnosis of trichobezoar with Rapunzel syndrome was made. The tail was pulled with very mild traction and gentle manipulation of the duodenum and proximal jejunum as it kept on coming out. To our surprise, it was extending upto jejunum in both the patients. Absolute gentleness in handling the hair mass and its long tail resulted in its complete retrieval in both the patients (Figs. 2 and 3). The stomach lumen was irrigated liberally with normal saline and the wounds were closed in layers.

3. Case 3 & 4: cases of trichobezoar without Rapunzel syndrome

Two girls aged 14 years and 8 years respectively presented to our outpatient department at different times with almost similar complaints of pain abdomen, fullness in the mid-upper abdomen, decreased appetite, early satiety, inability to gain weight and easy fatiguability. There was history of episodes of nausea and vomiting off and on, the frequency of which had increased gradually for last few months. On general physical examination, both the patients were having obvious pallor and wrinkling of skin and other signs of malnutrition. On abdominal examination, there was a large lump palpaple in epigastrium and left hypochondrium. On abdominal ultrasonography, the stomach was found to be filled completely with a large intraluminal mass conforming to the shape of the stomach. Both the patients were optimized and taken up for open surgery considering the very large size of the mass. Laparotomy with gastrotomy was done by right transverse supraumbilical incision. The whole lumen of stomach was found to be occupied by a fowl smelling, hard mass of hair interspersed with undigested vegetable matter in both the patients (Figs. 4 and 5). The mass was confined to stomach itself and there was no extension beyond it. So, an operative diagnosis of trichobezoar made and the mass evacuated and stomach repaired in two layers with interrupted silk sutures. The abdominal wound was closed in layers.

The post operative period was uneventful in all the four patients. On persuading them, all the patients admitted to ingesting their own hair (trichophagia) right from the time they remembered. Therefore, their intensive psychiatric counselling was done and the patients were discharged on 8–10th post operative day in fair health. All of them are on regular follow-up with the operating surgeon, psychiatrist and child psychologist and doing very well in terms of weight gain. There are no complaints or recurrence of previous symptoms.

The clinical summary has been tabulated as follows:

S.No	Name	Age (Yrs)	Sex	CR.No.	Presenting complaints (almost same in all 4 cases)	Extent of the mass	History of psychiatric illness	Procedure	Clinical Progress at Follow up
1	Pooja	12	F	332311	Vomiting, lump abdomen, H/O early satiety, inability to gain weight	Extended to jejunum	Yes	Laparotomy with Gastrotomy with retrieval of mass	Satisfactory
2	Mafia	4	F	496503	Vomiting, lump abdomen, H/O loss of appetite, failure to thrive	Extended to jejunum	Yes	– do-	Satisfactory
3	Meenu	8	F	797371	Pain abdomen inability to gain weight, nausea, epigastric fullness, early satiety	Localised to stomach	Yes	–do-	Satisfactory
4	Anita	14	F	851273	Pain abdomen, epigastric fullness, decreased appetite, early satiety, inability to gain weight and easy fatiguability	Localised to stomach	Yes	-do-	Satisfactory

4. Discussion

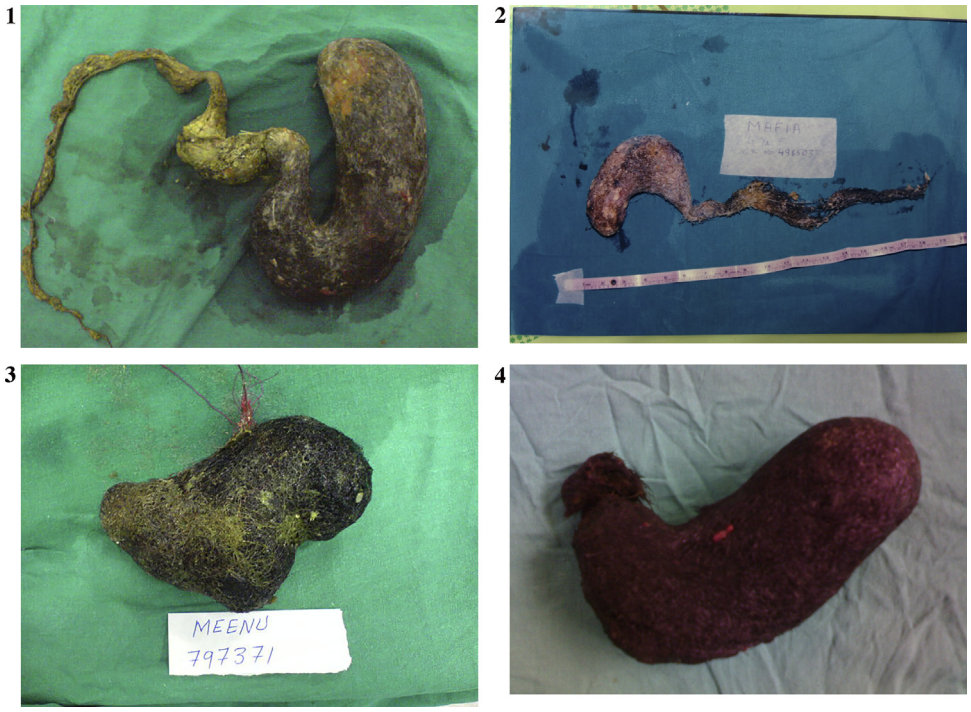
Rapunzel had magnificent long hair, fine as spun gold [6].

Rapunzel's hair fell to the ground like a rainbow.

It was as strong as a dandelion and as strong as a dog leash [7].

Rapunzel syndrome is named after a girl with long magical hair in the fairy tale written by Grimm Brothers in 1812. Rapunzel lowered her long tresses from the prison tower to the ground to permit her young prince to climb up her window and rescue her [6].

The Rapunzel syndrome was first reported in the literature by Vaughan et al. in 1968 [8].



Figs. 2–5. Photographs of the retrieved specimens showing trichobezoar with large tailed extension (A & B) and confined to the stomach only (C & D).

Bezoar is a melange of swallowed foreign material in gastrointestinal tract, most commonly in stomach. This term 'bezoar' is believed to be arrived from an Arabic word 'badzehr' or Persian word "panzehr" which means antidote. In ancient times, these bezoars from animal gut were anticipated to have medicinal property as antidote to poisons and were also used as precious stones. The most primitive reference on the bezoar was in 12th BC made by Sushruta in India. Charak, a noted Indian physician in the 2nd and 3rd century BC also described the disease in his book Charak Samhita. The first case of bezoar in Western countries was reported in 1779 while doing autopsy of a patient who died of gastric perforation and peritonitis [2,9].

Trichobezoars are formed as a result of accumulation of hair in gastrointestinal tract. Hair being slippery, get trapped in gastric mucosal folds, eluding peristalsis. More and more hair conglumerate and form a stomach shaped mass. Mucus secreted in stomach coats this trichobezoar and gives this a shiny glistening surface and acid secreted in stomach denatures the hair protein which gives black colour to bezoar Fermentation and decomposition of entrapped food especially fat leads to characteristic rancid odour in patient's breath and bezoar [5,10]. Trichobezoars are most commonly seen in females (approximately 90%) of age group 13–19 years with an unrevealed psychiatric disorder. In our series too all the four patients are females. About 50% of these patients are found to be trichophagic. Around 30% of the trichotillomaniac patients indulge in trichophagia (eat their hair) to the extent that requires surgical interventions in removal of trichobezoar [11]. The site of hair pulling is most commonly from the scalp, but can occur from the eyelashes, eyebrows, and pubic area. It can also predispose with the habit of eating threads of clothes. It is important to rule out such history because sometimes signs of trichotillomania are not present.

Very few reports of Rapunzel syndrome are documented in psychiatric literature. An explanation for such disparity is due to the fact that most cases of trichotillomania are referred early to psychiatrist before the development of Rapunzel syndrome [12,13]. Clinical manifestations depend on the bezoar's location and size. Affected patients mostly remain asymptomatic for many years, till the bezoar increases in size to the extent which is responsible to produce clinical symptoms. It is commonly presented as abdominal pain, nausea, vomiting, decreased appetite, early satiety and symptoms due to intestinal obstruction and peritonitis can also be there [5]. Clinical presentation of these patients may be confusing as often they are not forthcoming with a history of trichophagia either due to embarrassment or the unintentional nature of the problem. Trichobezoars should be considered as a differential diagnosis in a young female patient with a mobile epigastric mass. The common complications are anaemia, haemetemesis, gastric mucosal erosion, ulceration, and perforation of the stomach or the small intestine, gastric outlet obstruction, intussusception, obstructive jaundice, protein-losing enteropathy, acute pancreatitis, and death.

Usually, trichobezoar is confined to the stomach, but rarely it may extend from the stomach to the small intestine (even colon). This is an unusual form called Rapunzel syndrome [14]. Less than 40 cases of trichobezoar with Rapunzel syndrome have been reported in medical literature [5]. Bezoars can also be found distally in the gastrointestinal tract without continuity with the stomach due to breakage and distal propulsion.

Bezoars have been shown to be the cause of 0.8% of bowel obstructions that were managed laparoscopically [15]. Diagnosis can be easily made with the use of CT scan and endoscopy The most common diagnostic tool used in the literature is a CT scan, with a typical image showing a well-defined intraluminal ovoid heterogeneous mass with interspersed gas.

The gold standard for diagnosis though is upper gastrointestinal endoscopy [15,16]. Successful management and treatment of a trichobezoar demands removal of the mass and prevention of relapse. In the early stages endoscopic removal is possible, if bezoar is limited to small area and is of small size. So, endoscopy usually has a diagnostic role. The hair appears black (despite the normal hair colour) due to denaturing of the hair protein by the acid. Management almost always requires surgical removal. It can be removed by using endoscopic techniques or surgery (laparoscopic approach or open surgery). Choice of method depends on available resources,

technical expertise, location, consistency and size of bezoar. Phytobezoars and lactobezoars are easy to remove using endoscopic techniques because of small size. It is less effective in trichobezoar removal as they are usually of large size. Gorter et al., in a retrospective review of 108 cases of trichobezoar, evaluated the available management options [14]. It was noted that whereas 5% of attempted endoscopic removals were successful, 75% of attempted laparoscopies were successful. However, laparotomy was 100% successful and thus favoured as their management of choice [14].

All of our patients were managed surgically because of delayed presentation in our set up as psychiatric illness is still a taboo in a developing country like India where majority of the population resides in villages. So, open surgical intervention in the form of laparotomy and gastrotomy with retrieval of the mass was done.

Other modalities of treatment like extracorporeal shock wave lithotripsy, laser ignited mini-explosive technique, intragastric enzyme administration (pancreatic lipase, cellulase), and medicaments (like metoclopramide, acetylcysteine) have been reported with varying results. These are required for fragmentation of a large sized bezoar so that the laparotomy becomes easier. Laparoscopy has also been used with limited success. Indications for laparoscopic removal of bezors are small/medium sized bezoars. Advantages include reduced hospital stay and better cosmesis. Disadvantages include more peritoneal contamination as there are more chances of spillage of hair in peritoneal cavity, consumes more time, difficult to examine rest of bowel for additional pieces which may lead to secondary obstruction [5,14]. Laparotomy is frequently chosen treatment as it is 100% effective, rarely complicated and allows a careful examination of the entire gastrointestinal tract [5,14,17].

It is emphasized that the majority of these patients have an underlying psychiatric or social disorder. So, the treatment of coexisting psychiatric illness is of utmost importance and regular follow up is strongly recommended for psychiatric evaluation periodically. All of our patients were having psychiatric illness which was diagnosed only at the time of index admission after finding the presence of trichobezoar incidently for their vague symptoms that too after extensive inquest. A multidisciplinary approach is essential to prevent recurrence of the problem.

5. Conclusion

Trichobezoar leading to Rapunzel syndrome is an extremely rare entity. The clinical presentation is usually vague and non-specific. Treatment is mainly surgical because of delayed presentation in majority of the cases. Psychiatric illness is the usual association.

Therefore, along with removal, treatment should also focus on prevention of recurrence. So, in addition to the acute surgical treatment, parental counselling, neuropsychiatric treatment, follow-up and behavioural therapy should be continued to prevent recurrence.

Conflicts of interest

There is no conflict of interest amongst the authors.

Funding source

There was no source of funding for our research.

Ethical approval

Not applicable as it is a case series.

Consent

We state that the work has been reported in line with the SCARE criteria [18].

The consent has been taken from the parents of the child for publication of this case series.

Author contribution

Pradeep Kajal – Diagnosed and operated upon the patients and did the final editing.

Namita Bhutani – Reviewed the literature and wrote the article.

Niharika Tyagi – Provided the needed articles.

Pratibha Arya – Developed the images and managed the patients post-operatively.

Guarantor

Kamal N. Rattan.

References

[1] C.A. Santiago Sánchez, P. Garau Díaz, H.L. Lugo Vicente, Trichobezoar in a11-year old girl: a case report, Boletín Asoc. Médica Puerto Rico. 88 (March (1–3)) (1996) 8–11.
[2] S. Naik, V. Gupta, S. Naik, et al., Rapunzel syndrome reviewed and redefined, Dig. Surg. 24 (3) (2007) 157–161.
[3] E. Pérez, J.R. Sántana, G. García, et al., Perforaciéngástrica en adulto por trichobezoar (síndrome de Rapunzel), Cirugía Espaóola 78 (4) (2005) 268–270.
[4] R.R. Gorter, C.M. Kneepkens, E.C. Mattens, D.C. Aronson, H.A. Heij, Management of trichobezoar: case report and literature review, Pediatr. Surg. Int. 26 (5) (2010) 457–463.
[5] V. Gonuguntla, D.-D. Joshi, Rapunzel syndrome: a comprehensive review of an unusual case of trichobezoar, Clin. Med. Res. 7 (3) (2009 Sep) 99–102.
[6] Grimm Brothers: Rapunzel Translated by R. Godwin-Jones, Richmond, Virginia Common Wealth University Department of Foreign Language (1994–1999).
[7] A. Sexton, Rapunzel, in Transformations, Oxford University Press, London, 1972.
[8] E.D. Vaughn Jr., J.L. Sawyers, H.W. Scott Jr., The Rapunzel syndrome-an unusual complication of intestinal bezoar, Surgery 63 (1968) 339–343.
[9] R.S. Williams, The fascinating history of bezoars, Med. J. Aust. 145 (1986) 613–614.
[10] R.M. Zent, C.C. Cothren, E.E. Moore, Gastric trichobezoar and Rapunzel syndrome, J. Am. Colloids Surg. 199 (2004) 990. Ed by a large duodenal diverticular bezoar, World J. Gastroenterol. 18 (October (38)) (2012) 5485–5488.
[11] A.S. Frey, M. McKee, R.A. King, A. Martin, Hair apparent: Rapunzel syndrome, Am. J. Psychiatry 162 (2005) 242–248.
[12] S. Tiago, M. Nuno, A. João, V. Carla, M. Gonçalo, N. Joana, Trichophagia and trichobezoar, Clin. Pract. Epidemiol. Mental Health 8 (2012) 43–45.
[13] C. Bouwer, D.J. Stein, Trichobezoars in trichotillomania: case report and literature review, Psychosom. Med. 60 (1998) 658–660.
[14] R.R. Gorter, C.M. Kneepkens, E.C. Mattens, D.C. Aronson, H.A. Heij, Management of trichobezoar: case report and literature review, Pediatr. Surg. Int. 26 (5) (2010) 457–463.
[15] J.D. Fraser, C.M. Leys, S.D. St Peter, Laparoscopic removal of a gastrictrichobezoar in a pediatric patient, J. Laparoendosc. Adv. Surg. Tech. A 19 (6) (2009) 835–837.
[16] P.N. Mohite, A.B. Gohil, H.B. Wala, M.A. Vaza, Rapunzel syndrome complicated with gastric perforation diagnosed on operation table, J. Gastrointest. Surg. 12 (2008) 2240–2242.
[17] E.C.G. Tudor, M.C. Clark, Laparoscopic-assisted removal of gastric trichobezoar; a novel technique to reduce operative complications and time, J. Pediatr. Surg. 48 (March (3)) (2013) e13–e15.
[18] R.A. Agha, A.J. Fowler, A. Saetta, I. Barai, S. Rajmohan, D.P. Orgill, for the SCARE Group, The SCARE statement: consensus-based surgical case report guidelines, Int. J. Surg. 34 (2016) 180–186.