CASE REPORT

Rapunzel syndrome: A South African variety

J Plaskett,[1] MB ChB, FCS (SA), MMed; **G Chinnery**,[1] MB ChB, FCS (SA), Cert Gastroenterology (SA) Surg; **D Thomson**,[1] MB ChB, FCS (SA), Cert Crit Care (SA) Surg; **S Thomson**,[1] ChM, FRCS (Eng & Edin), FRCP (Edin); **B Dedekind**,[2] MB ChB, FCS (SA); **E Jonas**,[1] MB ChB, MMed, FCS (SA), PhD

[1] *Department of Surgery, Faculty of Health Sciences, Groote Schuur Hospital and University of Cape Town, South Africa*
[2] *Netcare Christiaan Barnard Memorial Hospital, Cape Town, South Africa*

Corresponding author: *E Jonas (eduard.jonas@uct.ac.za)*

Trichobezoars are intraluminal accretions of ingested hair. Rapunzel syndrome is a rare and extreme presentation, with the trichobezoar extending into the small intestine. It is most frequently reported in children and psychiatric patients. We report a South African series of 5 patients who presented with trichobezoars. Each patient was retrospectively reviewed and analysed with regard to background, demographics, clinical presentation, diagnosis, surgical management and complications. Five female patients with a median age of 19 (range 12 - 27) years presented with clinical symptoms, including early satiety, intermittent vomiting with gastric outlet obstruction, abdominal pain and weight loss. The diagnosis was made by endoscopy, abdominal computed tomography (CT) imaging, barium meal examination or plain abdominal radiography. Two patients presented with sealed/contained gastric perforations, and 1 patient with a small-bowel perforation. All 5 bezoars, 2 of which consisted entirely of artificial hair extensions, extended into the jejunum, the longest measuring 1.4 m. All were removed by laparotomy. While trichobezoars are a rare entity, they may present with significant complications, such as obstructions and perforations. In view of the infection risk and considerable size of many of these bezoars, an open removal is probably safer than any minimally invasive attempt.

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Trichotillomania (a compulsive desire to pull out one's own hair) was first described by Hallopeau in 1889, while the first case of trichobezoar was described by Baudamant in 1779.[1] Rapunzel syndrome is a rare form of trichobezoar, reported by Vaughan *et al.*[2] in 1968, and characterised by extension into the small intestine. To date, <30 cases have been reported in the literature.[1] Rapunzel syndrome commonly occurs in young females, who usually have an underlying psychiatric disorder, with only a single reported male patient who ate the hair of his sisters.[3,4] Approximately 10% of patients with trichotillomania also demonstrate trichophagia.[5] The name Rapunzel originates from the Brothers Grimm fairy tale of a 12-year-old princess locked inside a tower, who uses her long, golden hair to permit her young prince to climb up to her window and rescue her.[6]

The term bezoar is believed to be derived from the Arabic *badzehr* or the Persian *panzehr*, meaning antidote. In ancient times, bezoars were believed to have medicinal and magical properties and considered as antidotes to a variety of poisons and diseases.[7] Bezoars are typically either phytobezoars (composed of indigestible vegetable or fruit fibres), trichobezoars (composed of accretions of ingested hair), lactobezoars (composed of milk protein), or pharmacobezoars (composed of concretions of various medications).[8] Trichobezoars form when ingested hair strands become retained in the folds of the gastric mucosa, escaping peristaltic propulsion owing to their slippery surface. More hair becomes enmeshed by peristalsis, forming a ball too large to exit the stomach and causing gastric atony owing to its large size.[9,10] The significant size of a bezoar may eventually reduce the mucosal blood supply to the stomach and proximal small bowel, causing ulceration and perforation. Various criteria have been used in the literature to define Rapunzel syndrome: a gastric trichobezoar with a tail extending up to the ileocaecal junction,[11] a simple trichobezoar with a long tail extending beyond the pylorus,[2] or a bezoar of any size causing obstruction.[1] For the purpose of our case series, Rapunzel syndrome was defined as a symptomatic gastric trichobezoar, with its tail extending into the jejunum for a variable distance, as originally described by Vaughan *et al.*[2]

Risk factors for the formation of gastric bezoars are impaired gastric motility (previous vagotomy or gastroparesis), previous gastric surgery, gastric outlet obstruction, high-fibre diet (especially in the case of phytobezoars) and impaired chewing. Common presenting complaints include abdominal pain, nausea and vomiting, and gastric outlet obstruction. Patients may remain asymptomatic for many years.[1] Less commonly, patients may present with weight loss, anorexia, haematemesis and intussusception.[12,13] Complications include gastric ulceration, perforation with peritonitis,[14-16] obstructive jaundice,[12] acute pancreatitis[17,18] and even death.[19] We describe a retrospective series of 5 patients with trichobezoar, who presented to our unit over a 5-year period.

Patients and methods

Patients with trichobezoars, who presented to the Upper Gastrointestinal Unit, Groote Schuur Hospital, Cape Town, South Africa, were identified from a prospective registry. The patients' demographic profiles, presenting symptoms, diagnostic findings, treatment and outcomes are reported. The registry has been approved (ref. no. HREC 488/2017) by the Human Research Ethics Committee of the University of Cape Town.

Results

During the study period, 5 patients presented to our unit with trichobezoar (Table 1). All patients were female, with a median age of 19 (range 12 - 27) years. Three patients were of Indian and 2 of African ethnicity. Four individuals were highly functional (3 high-school

Table 1. Patient information

Patient	Age, years	Sex	Presentation	Method of diagnosis	Intra-operative finding	Removal technique	Hair	Complications	Evaluation
Clerk	27	F	Vomiting, weight loss	Endoscopy and CT of the abdomen	Sealed prepyloric perforation, large trichobezoar extending into the jejunum	Laparotomy and gastrotomy	Synthetic	None	Patient denied trichophagia, despite psychiatric intervention
High-school learner	19	F	Abdominal pain, early satiety, epigastric mass	Endoscopy and barium meal	Large trichobezoar extending 1.4 m into the jejunum	Laparotomy and gastrotomy	Synthetic	None	Patient acknowledged 4 years of trichophagia
High-school learner	17	F	Abdominal pain	CT of the abdomen	Contained gastric perforation with massive gastric trichobezoar	Laparotomy and gastrotomy	Natural	None	Patient acknowledged trichophagia in times of stress
University student	23	F	Abdominal distention, abdominal pain	Plain abdominal radiographs	Small-bowel obstruction and perforation from gastric trichobezoar extending into the small bowel	Laparotomy and gastrotomy	Natural	None	Patient acknowledged trichophagia in times of stress
Cerebral palsy patient	12	F	Epigastric mass, anorexia	Endoscopy and CT of the abdomen	Large trichobezoar extending into the jejunum	Laparotomy and gastrotomy	Natural	None	Patient knew she had trichotillomania and trichophagia

F = female; CT = computed tomography.

graduates/students and 1 university student), while the 5th patient was a 12-year-old cerebral palsy child with known trichotillomania. Presenting symptoms included: anorexia and early satiety (n=2), intermittent vomiting with features of gastric outlet obstruction (n=1), abdominal pain (n=3) and weight loss (n=1). Four of the patients had a history of trichotillomania and trichophagia. Diagnosis was made by upper-gastrointestinal (GI) endoscopy in 3 cases (complemented by abdominal computed tomography (CT) and a barium meal), abdominal CT alone in 1 case, and plain abdominal radiography in 1 case (Figs 1 - 3). All patients were managed by laparotomy, gastrotomy and open removal of the bezoar (Figs 4 and 5). In 2 patients there was an intraoperative finding of a sealed,

Fig. 1. Endoscopic examination, indicating a trichobezoar.

Fig. 2. Barium meal examination, showing a gastric trichobezoar with extension into the duodenum.

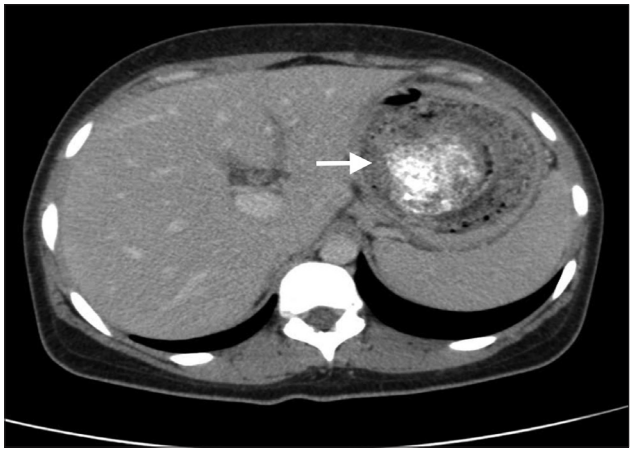

Fig. 3. (A) Computed tomography scan of the abdomen, showing a gastric trichobezoar.

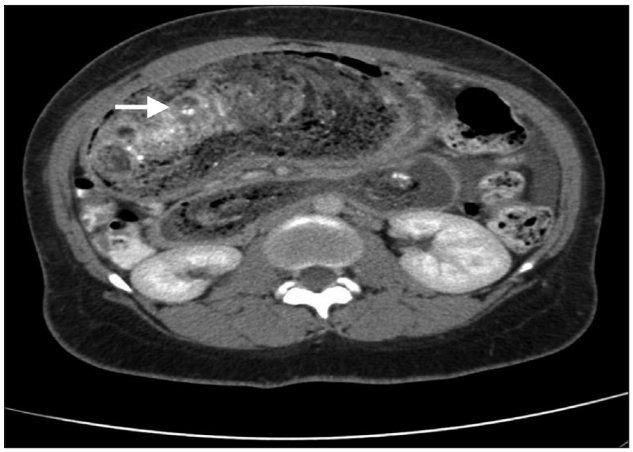

Fig. 3. (B) Computed tomography scan of the abdomen, showing a gastric trichobezoar extending into the jejunum.

contained gastric perforation, and a jejunal perforation was found in 1 patient. All 5 bezoars extended into the jejunum, the longest measuring 1.4 m; 2 consisted entirely of artificial hair extensions. All patients had an uncomplicated postoperative course and 4 were referred for psychiatric evaluation.

Discussion

We present the joint largest series of patients with Rapunzel syndrome. Fallon *et al.*[20] described a series of 7 trichobezoars, 5 of which had features of Rapunzel syndrome. Our series also includes 2 patients in whom the bezoar consisted entirely of artificial hair extensions. Only 2 other cases of trichobezoar composed of synthetic hair have been described.[21,22]

Trichobezoar and Rapunzel syndrome should be considered as a differential diagnosis in young females who present with nonspecific symptoms, such as epigastric pain, fatigue and weight loss, and an epigastric mass. A thorough history regarding any psychiatric comorbidity, as well as a history of trichotillomania/trichophagia, should be sought, although most patients deny this, even when specifically asked.[1] Severe halitosis or patchy alopecia may be an indication of the condition. Physical examination may occasionally reveal a palpable abdominal mass. Plain abdominal radiographs often serve as the initial imaging modality, showing either an inhomogeneous mass or filling defect in the region of the stomach. A diagnosis based solely on plain radiography can be made in 50 - 75% of patients with small-bowel obstruction,[23] while only

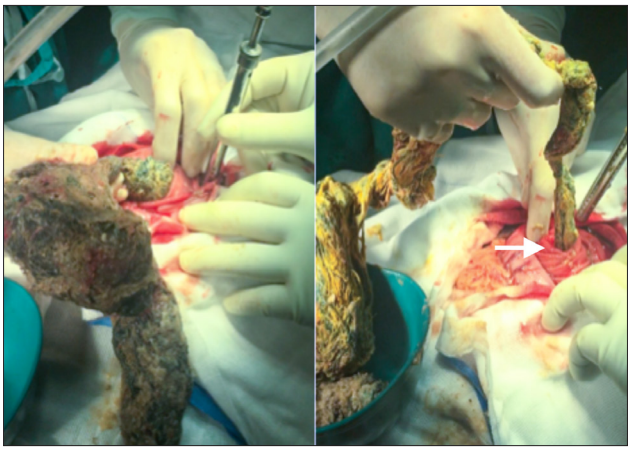

Fig. 4. Removal of a bezoar via open gastrotomy.

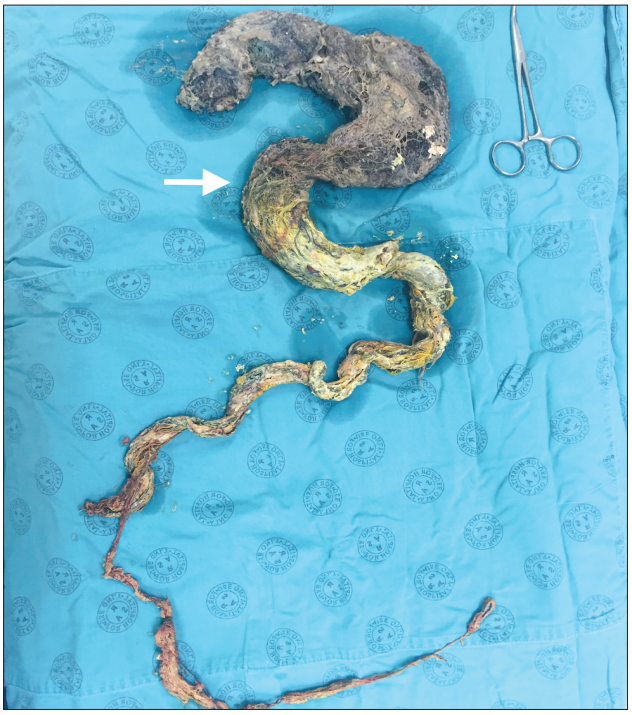

Fig. 5. Removal of a 1.4 m trichobezoar.

10 - 18% of bezoars are recognised on radiographic features alone.[24,25] A barium meal may demonstrate a filling defect in the stomach and confirm the diagnosis. Both ultrasound and CT scans have been shown to be reliable methods for diagnosing trichobezoars and distinguishing them from other possible causes, particularly when an abdominal mass is palpated. Abdominal ultrasound scanning might demonstrate increased echogenicity, with a marked acoustic shadow due to intermixed hair and food, and CT imaging might show a low attenuating heterogenous mass containing trapped air, with oral contrast most prominent at its margins.[26,27] In a retrospective analysis of 17 patients with GI bezoars by Ripollés *et al.*,[24] sonography detected only 25% of gastric bezoars, while CT imaging indicated the preoperative diagnosis in all patients. Upper GI endoscopy, however, remains the gold standard for diagnosis and also allows for treatment options in selected cases of small trichobezoars.[28,29]

As the literature comprises small series and case studies, and there is a paucity of randomised data, it is difficult to draw conclusions regarding the optimal management of this rare group of patients.

The goals of bezoar treatment are the removal of the bezoar and the prevention of recurrence by addressing the underlying psychiatric/emotional cause.[29] Unlike phytobezoars, trichobezoars are often resistant to enzymatic dissolution.[8] Endoscopic management of the trichobezoar is rarely a definitive treatment, as the bezoars are usually too large for dissolution and retrieval, and fragmentation is often impossible owing to size, density and hardness, even with specialised bezotomes and bezotriptors.[28,30] Endoscopic fragmentation can also lead to distal migration and small-bowel obstruction.[20] In the case of Rapunzel syndrome, complete removal without breakage and distal migration emphasise the importance of adequate exposure, as well as the limitations of endoscopic management. Furthermore, a recent review by Gorter *et al.*[30] showed that only 5% of trichobezoars were amenable to complete endoscopic removal. Endoscopic management was not attempted in any of our patients because of the large size of the bezoar and anticipated difficulty with complete removal.

The optimal surgical management of trichobezoar remains controversial and divided between laparoscopic and open approaches, most often dictated by local expertise and the size of the bezoar.[30] The first laparoscopic removal of a trichobezoar was reported by Nirasawa *et al.*[31] in 1998. Since then, fewer than 10 other reports of attempted laparoscopic removal have been published, usually restricted to trichobezoars without extension. A novel laparoscopic-assisted technique using a wound retractor has also been described to improve access and reduce operative time and complications.[32] The laparoscopic approach remains an attractive option, warranting further consideration with regard to its appropriate use in managing this rare pathological condition.

The surgical management of Rapunzel syndrome, however, appears more clearly defined, as the bezoar extends beyond the pylorus and because of the risk of incomplete removal. Careful manipulation of the jejunum, to ensure complete removal of the tail without perforation or breakage, further highlights the limitations of the laparoscopic approach. Increased operative time, risk of intra-abdominal spillage and necessity of extended incisions (for bezoar removal) are additional arguments made by opponents of this approach.[30] All of our patients were successfully treated with a laparotomy. This is comparable with the findings of Fallon *et al.*[20] and Gorter *et al.*,[30] demonstrating a 99% and 100% success rate with laparotomy, respectively. Our complete lack of postoperative complications is also in line with the benign postoperative course reported in the series by Fallon *et al.*[20] However, there have recently been 3 reports of successful laparoscopic removal of trichobezoars in paediatric patients, 2 of whom had post-pyloric extension.[33-35] Psychiatric assessment and long-term behavioural therapy should be considered a standard component of treatment.[30,36]

Conclusion

Although Rapunzel syndrome is a very rare entity, our series suggests that its occurrence could be higher than expected among patients with trichobezoars. The finding of a contained perforation in 60% of patients emphasises the potential for serious complications. Preoperative diagnosis requires a thorough history, complemented by radiological and endoscopic investigations. Laparotomy and open removal by gastrotomy and/or enterotomy seem to be the most frequently reported approaches, with the highest success rate and lowest complication rate. The case for laparoscopic surgical management remains compelling in appropriate cases, where it can achieve complete removal. Further experience in the form of larger series, however, is required to establish its precise role. Psychiatric consultation should be considered in all patients to prevent recurrence.

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