Rapunzel syndrome is not just a mere surgical problem: A case report and review of current management

Obinna Obinwa, David Cooper, Faraz Khan, James M O’Riordan

Obinna Obinwa, David Cooper, Faraz Khan, James M O’Riordan, Department of Surgery, the Adelaide and Meath Hospital, Dublin Incorporating the National Children’s Hospital, Tallaght, Dublin 24, Ireland

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Correspondence to: Obinna Obinwa, MCh, MRCSI, Department of Surgery, the Adelaide and Meath Hospital, Dublin Incorporating the National Children’s Hospital, Tallaght, Dublin 24, Ireland. obinna.obinwa@amnch.ie
Telephone: +353-1-4142211
Fax: +353-1-4142212

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Abstract
Recurrent Rapunzel syndrome (RRS) is a rare clinical presentation with fewer than six cases reported in the PubMed literature. A report of RRS and literature review is presented. A 25-year-old female was admitted to hospital with a 4-wk history of epigastric pain and swelling. She had a known history of trichophagia with a previous admission for Rapunzel syndrome requiring a laparotomy nine years earlier, aged 16. Psychological treatment had been successfully achieved for nine years with outpatient hypnotherapy sessions only, but she defaulted on her last session due to stressors at home. The abdominal examination demonstrated an epigastric mass. Computer tomography scan revealed a large gastric bezoar and features of aspiration pneumonia. The patient underwent emergency open surgical laparotomy for removal as the bezoar could not be removed endoscopically. The bezoar was cast in a shape that mimicked the contours of the stomach and proximal small bowel, hence the diagnosis of RRS. The patient was seen by a psychiatrist and was commenced on Quetiapine before discharge. She continues to attend follow-up.

Key words: Trichobezoars; Rapunzel syndrome; Recurrence; Obsessive compulsive disorders; Case report

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Core tip: There remain to be clear guidelines on the management of trichotillomania associated disorders. Here we report that Rapunzel syndrome requires a comprehensive and long-term psychiatric follow-up as it is not a primary surgical condition. A late relapse of the condition is possible and recognizing this as a clinical possibility can intensify efforts in relapse prevention during the follow-up period. This approach is important in eliminating the need for recurrent surgical
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INTRODUCTION

A bezoar is a collection of foreign material in the gastrointestinal tract. A trichobezoar is a bezoar formed by the ingestion of hair and occurs typically in patients with trichotillomania. The latter is defined as an irresistible desire to pull out one's hair and it has been included in the 2013 Diagnostic Statistical Manual (DSM-5) of the American Psychiatry Association as an obsessive compulsive disorder[1].

Rapunzel syndrome is a rare manifestation of a trichobezoar, which occurs when strands of swallowed hair extend beyond the pylorus of the stomach, into the intestine as a tail[2]. It was first described by Vaughan et al[3] in 1968. Primary or recurrent cases of trichobezoars may lead to complications such as intussusception[4,5], pancreatitis[6] and bile duct dilatation[7,8]. Significant other complications such as gastric perforation[7,8], peritonitis[9], and even death[10] have also been reported. Despite the potential for significant complications and mortality, there is still a lack of any specific and comprehensive guidelines on appropriate postoperative follow-up for patients with Rapunzel syndrome to reduce the risk of recurrence[11].

In this case report, we present a rare case in which Rapunzel syndrome represented nine years following an initial laparotomy. This manuscript is written in accordance with the case report (CARE) guidelines[12]. The clinical management dilemmas in this case, including those accounting for the recurrent Rapunzel syndrome (RRS), have been reported to inform guidelines on appropriate postoperative follow-up of patients with Rapunzel syndrome.

CASE REPORT

A 25-year-old female was admitted to hospital with a 4-wk history of epigastric pain, swelling and early satiety. The symptoms, while initially intermittent, had become more constant and severe over the four days prior to admission. She denied any nausea, vomiting, weight loss or change in bowel habit. She had a known history of trichophagia (compulsive ingestion of hair) with a previous admission for Rapunzel syndrome requiring an anterior gastrotomy nine years earlier, aged 16. She had been referred to a psychiatry service following this episode and was successfully managed with a non-pharmacological treatment strategy in the form of behavioural therapy for nine years. The patient reported finding initial outpatient hypnotherapy sessions very beneficial, however; she admitted to later defaulting on follow-up appointments due to stressors at home.

On examination, she was anaemic, and her abdomen was distended with an upper midline laparotomy scar visible, consistent with her previous surgery. A firm abdominal mass, which extended from her left subcostal region to her umbilicus, was palpable. At this point, the differential diagnosis included an enlarged spleen, recurrence of the trichobezoar or Rapunzel syndrome. Her blood work revealed a microcytic hypochromic anaemia with a haemoglobin level of 9.1 g/dL. Blood urea, creatinine, electrolytes, blood glucose, serum amylase and liver function tests were normal. An abdominal CT showed a grossly distended stomach and pylorus filled with debris (Figure 1), with infiltrates within the right lower lung lobe. Following this, the patient consented to the removal of the foreign body under general anaesthesia (Figure 2).

The patient was brought to the theatre, intubated, and under general anaesthesia, a diagnostic upper gastrointestinal endoscopy was performed. The endoscopy showed that the stomach and pylorus were filled with a large mass of hair (Figure 3). The greater curvature of the stomach was also ulcerated. The high density of the hair conglomerate precluded successful endoscopic extraction, and surgical exploration was performed through a 7-cm upper midline incision. The adhesions from her previous surgery were divided and an Alexis® O Wound Protector (Applied Medical, United States) was used to protect the wound. A gastrostomy (5 cm) was made in the anterior stomach away from the pylorus. The foreign body was visualised, grasped, and carefully extracted from the stomach. The trichobezoar weighed 850 g and was cast in a shape that mimicked the contours of the stomach and proximal small bowel, hence the diagnosis of RRS (Figure 4). The gastrostomy was closed in two layers, and this was followed by...
fascial and skin closure.

Postoperatively, the patient received analgesia and was kept nil by mouth for three days. She received intravenous fluids and proton pump inhibitors during this period. The postoperative period was complicated by a chest infection on day 2. The infection necessitated chest physiotherapy and an extended duration of prophylactic antibiotics to a full 7-d course. Of note, the chest infection was apparent at the time of preoperative diagnosis. The nasogastric tube was removed on day 3, and her diet was slowly re-introduced. She had resumed full diet by day 5 and was also commenced on haematinics. She was reviewed by a psychiatrist and was started on Quetiapine 25 mg daily before discharge on day 12. Outpatient follow-up for further management of her mood symptoms and cognitive behavioural therapy was arranged.

**DISCUSSION**

Rapunzel syndrome is not a primary surgical condition. Treating the underlying trichotillomania is critical in preventing a relapse, but this can be challenging in clinical practice. Clinical dilemmas and valuable lessons learned from the management of this rare case of recurrence are described herein.

Firstly, laparotomy is the recommended approach of choice for removal of the trichobezoar in Rapunzel syndrome\(^{[11,13]}\). Enzymatic degradation, pharmacotherapy, endoscopic fragmentation and laparoscopy have been shown to be ineffective in these cases as the tail often extends into the jejunum\(^{[13,14]}\). The
The major drawback of the open surgical technique is the high incidence of postoperative infection[13]. However, the chest infection in this case report was arguably present due to aspiration at the time of diagnosis and was evident on the preoperative CT imaging. The site of incision, the wound protection technique, and the outlined postoperative care all limited the morbidities in this case. The pre-morbid anaemia and gastric ulceration were also well managed using haematinics and proton pump inhibitors.

Secondly, this case showed that cognitive behaviour therapy in the form of exposure and response prevention, although useful in the initial management may become limited in the long-term prevention of relapse of trichotillomania. For this behavioural therapy to be effective, there needs to be a comprehensive home support network with family or friends also monitoring treatment compliance at home[6,15]. Randomised control trials have shown that patients who respond to psychotherapy might still be stigmatized or be socially rejected[16]. Such stigmatization and rejection may lead to depression, the latter has been described as an independent predictor of quality of life deficits in patients with trichotillomania[17]. The involvement by the family helps to reinforce treatment and facilitates early detection of relapse. Despite these efforts in the management of our case, the presence of home stressors was subtle and was undetected in the outpatient setting. The result was a delay in diagnosis of a relapse, an emergency presentation and morbidity at presentation.

Thirdly, a comprehensive and long-term psychiatric follow-up is needed in all cases as late relapse is possible. An ideal psychiatric follow-up approach is one which can early detect relapse, or highlight those who require closer monitoring and more aggressive treatment.

### Table 1 Management of cases of recurrent Rapunzel syndrome in the literature

| Ref.          | Year published | Age (S1) | Psychiatric management | Age (S2) | Psychiatric management | Interval (recurrence) | Reason for recurrence |
|---------------|----------------|----------|------------------------|----------|------------------------|-----------------------|-----------------------|
| Memon et al[2] | 2003           | 10       | Advised treatment of her emotional disturbances | 12       | Supervised psychiatric treatment[2] | 2 yr                  | Unresolved emotional stress factor (ignored psychiatric treatment, continued to eat hairs of females neighbours) |
| Eryilmaz et al[20] | 2004          | 12       | Psychiatric treatment[2] | 19       | Supervised treatment with family counselling | 7 yr                  | Underlying depressive personality disorder |
| Morales-Fuentes et al[22] | 2010         | 16       | No treatment mentioned | 22       | Psychiatric treatment[2] | 6 yr                  | Inadequate initial treatment Obsessive disorder Pleasure feeling of how the hair scraped the throat |
| Jones et al[24] | 2010           | 35       | No treatment mentioned | 37       | Quetiapine Habit reversal training with family and neighbours involvement | 2 yr                  | Inadequate initial treatment |
| Tiwary et al[18] | 2011           | 10       | Behavioural therapy Clomipramine after 1 mo Follow-up × 6/12 | 15       | Supervised psychiatric treatment[2] | 5 yr                  | Lack of psych follow-up Defaulted after 6/12 |
| Current study | 2016           | 16       | Behavioural therapy | 25       | Supervised behavioural therapy Quetiapine 25 mg | 9 yr                  | Defaulted follow-up due to stressors at home |

All cases involved female patients; Details not specified. S1: First surgical intervention; S2: Second surgical intervention.

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Figure 3 Gastroscopy showing the obstructing trichobezoar.

Figure 4 The fully extracted giant gastric trichobezoar with a tail.
Furthermore, patients who are on pharmacological therapy should be monitored by a psychiatrist. Continued surveillance by carers for adverse events while on medication is also advisable. Close monitoring is especially important during the times of adjustment of dosage regimens. Adjunctive investigations such as biannual abdominal imaging during the follow-up period has been proposed by some authors, while others advocate routine ultrasound or upper GI endoscopy at 6, 12 and 24 mo. The use of trichotillomania severity scales as a way of assessing treatment response may prove useful in the future. All these proposals are however yet to be universally adopted in clinical practice.

Currently, there are no Food and Drug Administration approved treatments for trichotillomania, which makes it difficult for clinicians to select an appropriate therapeutic plan. When effective, long-term treatment with an SSRI may be a reasonable first-line option to prevent relapse. Clobazam, quetiapine or augmenting an SSRI with an atypical antipsychotic have been used for treatment-resistant cases. However, all cases in which a drug treatment is considered should be referred to a psychiatrist who then makes a decision on the appropriate therapy. In this case report, quetiapine was recommended. Furthermore, patients on drug treatment should be carefully monitored as treatment may be associated with psychiatric comorbidity and suicidal ideation in later life. It is clear that new targets are warranted to ensure a clinically supported effective pharmacological approach to treat this condition.

Recurrence of Rapunzel syndrome is extremely rare and fewer than six cases have been reported in the PubMed database. Management of the condition can be challenging even in experienced hands. Our patient did well on cognitive therapy alone for nine years without any issues, and this justified the continued non-pharmacological management in the first instance. As mentioned earlier, pharmacological treatment may be limited and is not without risks, but this had to be instituted following the relapse. So far, the cases of recurrence have been recorded in females with variable times of between two and nine years between the initial surgical treatment and presentation with relapse (Table 1). Our review of the management also showed that RRS occurs when the underlying psychological trigger is under-diagnosed or treated. With specific reference to the index case report, it was principally due to an inadequate supervision by carers and subsequent failure of the patient to attend follow-up sessions.

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 RRS 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.

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