Recurrent Rapunzel syndrome

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ABSTRACT

Rapunzel syndrome is a rare condition that occurs when gastric trichobezoar extends beyond the pylorus and into the bowel. Recurrent Rapunzel syndrome is an extremely rare entity, with only two cases reported in the literature so far. Here, we present a case of recurrent Rapunzel syndrome in a 15-year-old girl. She underwent laparotomy twice in five years to extract trichobezoars of sizes 58 cm and 62 cm. In both instances when Rapunzel syndrome was diagnosed, a tuft of hair extending from the stomach toward the jejunum was observed. The patient underwent psychiatric consultation and supervision after the first laparotomy; however, a lack of follow-up resulted in recurrence. We conclude that complete removal of trichobezoar and psychiatric consultation, coupled with long-term follow-up, are essential to prevent recurrence.

Keywords: bezoars, Rapunzel syndrome, recurrent, trichobezoar

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INTRODUCTION

The first description of Rapunzel syndrome by Vaughan[1] dates back to 1968; since then, 16 more cases have been reported in the literature. Rapunzel syndrome is a rare manifestation of trichobezoar, which occurs when strands of swallowed hair extend beyond the stomach, into the intestine as a tail, and become an intestinal obstruction.[2] Recurrence of this condition is extremely rare, despite it being associated to a psychiatric component. So far, only two cases of recurrent Rapunzel syndrome have been reported in the literature. We report another case here.

CASE REPORT

A ten-year-old girl was admitted with epigastric pain and vomiting for six months in May 2001. On examination, she was anaemic with normal vital signs. A firm, non-tender, mobile mass placed transversely was palpable in the upper abdomen. Her complete blood picture revealed haemoglobin 9.0 gm/dl and white blood cell count 10.1 × 10^9/L (N70, L24, E2, M4). Blood urea, serum creatinine, serum electrolytes, blood sugar, serum amylase and liver function tests were normal. Urine analysis, chest radiograph and electrocardiography were within normal limits. Barium meal study revealed a large filling defect in the stomach that was suggestive of bezoars (Fig. 1). Gastrotomy was performed and a 58-cm-long trichobezoar that completely filled the stomach and extended far into the proximal jejunum was removed (Fig. 2). The patient was discharged on Day 8 after stitch removal, after which she was referred to a psychiatrist for treatment. Psychiatric treatment was mainly in the form of behavioural therapy. Clomipramine was added to the therapy after one month. The patient was followed up for the next six months.

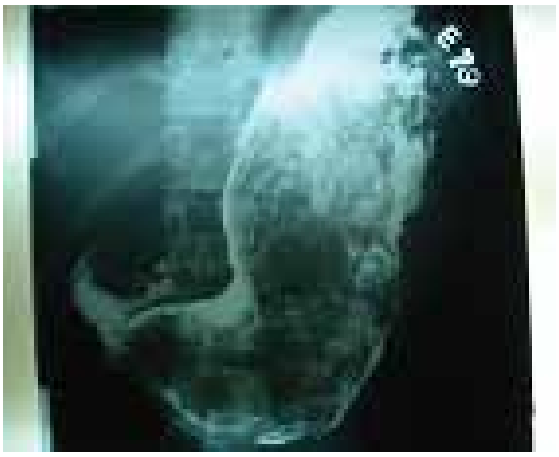

Fig. 1 Barium meal image shows filling defect.

Fig. 2 Photograph shows the trichobezoar extracted at the first laparotomy.

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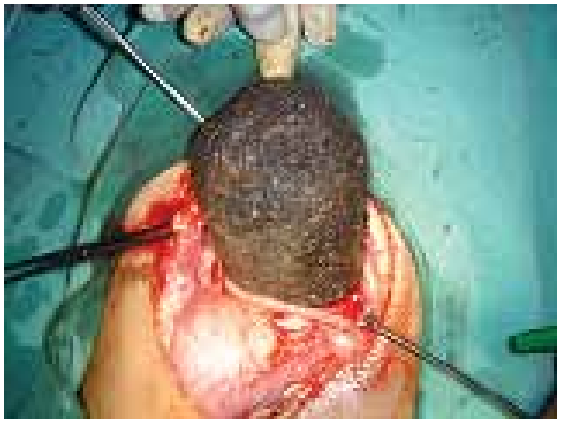

Fig. 3 Photograph shows the trichobezoar being extracted during the second laparotomy.

Fig. 4 Photograph shows the trichobezoar extracted at the second laparotomy.

Five years later, the patient was again admitted with a large upper abdominal mass associated with nausea, vomiting and loss of appetite. On physical examination, a firm, non-tender, mobile intra-abdominal lump was noted in the upper abdomen. Barium meal study and ultrasonography (US) of the abdomen revealed a large intraluminal mass in the stomach, extending distally into the jejunum. A clean acoustic shadow with a solid mass in the stomach confirmed the diagnosis of trichobezoar. Haematological and biochemical parameters were normal, except for the haemoglobin level (6.5 gm/dl). The patient had been anaemic at both presentations due to reduced nutritional intake caused by vomiting and loss of appetite. After preoperative blood transfusion, exploration was done through the previous abdominal scar. The stomach was found to be distended with a mobile lump. Gastrotomy was performed and a large, 62-cm mass of hair was removed (Figs. 3 & 4). The postoperative period was uneventful, and the patient had a smooth recovery. Oral dietary intake was started on Day 5, and stitches were removed on Day 7. The patient was referred to a psychiatrist after discharge on Day 8, and supervised treatment was administered.

DISCUSSION

To date, only two cases of recurrent Rapunzel syndrome have been described in the literature.[2,3] Rapunzel syndrome is an extensive form of gastric trichobezoar that extends into the small bowel. The description of the first case of human trichobezoar dates back to 1779, and the nomenclature seems to resemble the magical properties of these masses.[4] This symptom occurs most commonly in females under the age of 30, the youngest reported case being a six-month-old infant.[5] Our patient was ten years old at initial detection and 15 years old when recurrent Rapunzel syndrome was confirmed.

There are four types of bezoars described according to their composition: trichobezoars, phytobezoars, lactobezoars and miscellaneous.[6] Patients usually present with nausea, vomiting, abdominal pain, abdominal mass, weight loss, anaemia, jaundice, pancreatitis, bowel obstruction or intussusception. Slow-growing bezoars, if left untreated for a long time, may lead to obstruction, haemorrhage, ulceration, intussusceptions and perforations in the corresponding part of the bowel.[8] If the duodenum is affected, pancreatitis or obstructive jaundice may occur.[9] Sometimes, the fragmented part of the trichobezoar may break down and migrate distal to the ileocaecal valve, causing colonic bezoars.

Trichobezoar is commonly identified as a secondary symptom in young girls with psychiatric disorders.[7] Therefore, the diagnosis should be considered in patients who have a history of emotional disorders (e.g. trichotillomania). Plain radiograph films are not useful for diagnosing trichobezoar; instead, US revealing solid intraluminal mass in the stomach with clean acoustic shadow is a diagnostic indication of trichobezoar.[10] In addition, US can detect calcified mass, neuroblastoma, aneurysm, abscess and faecal material. Thus, the detection rate of trichobezoars by US has been reported to be around 88%.

Depending on the extension of the trichobezoars, contrast-enhanced computed tomography (CT) imaging, which is used to diagnose 97% of bezoars,[10] may also be used as a detection tool. Trichobezoars manifest as free-floating filling defects in the stomach, duodenum or jejunum.[10] Therefore, diagnostic tools such as US, CT and upper gastrointestinal (GI) endoscopy along with contrast radiography should be used to establish the diagnosis of Rapunzel syndrome.[8,10,11] We made the current diagnosis based on the patient's history, clinical examination and results of the barium meal study.

Small trichobezoars may be extracted by endoscopic fragmentation, vigorous lavage enzymatic therapy or a combination of these approaches.[12] Endoscopic removal after fragmentation using water pick, laser, extracorporeal shock-wave lithotripsy or by enzymatically dissolving the mass is possible in small-sized bezoars.[13,14] Another option for removing small bezoars is with laparoscopy through a small incision.[13] Large bezoars (> 20 cm) usually require removal through open surgery, as none of the above techniques are effective.[11] In Rapunzel syndrome and large bezoars, gastrotomy is essential for surgical removal, and enterotomy is required if impacted extension is present.[15] Routine upper GI endoscopy or US may be used to detect recurrence during follow-up.

Recurrent Rapunzel syndrome results if the psychiatric component is ignored. Our patient defaulted follow-up after six months, and hence did not complete the psychiatric treatment. This is similar to the first case of recurrent Rapunzel syndrome reported by Memon et al,[2] where recurrence was due to the discontinuation of psychiatric treatment. Another case of recurrent Rapunzel syndrome reported in the literature was that of a 19-year-old girl with depressive personality disorder, whose syndrome recurred after seven years.[3] Based on the three available reports, counselling and treatment for trichotillomania by a psychiatrist is integral to the management of Rapunzel syndrome. Cognitive behavioural therapy or psychotherapy is often used for treatment, as there are no formal guidelines to treat childhood trichotillomania.[16] However, it is reported that there is no difference in responses to psychotherapy, behavioural therapy, clomipramine or fluoxetine and that none of these treatments provide significant or lasting improvement of symptoms.[17] In the absence of convincing evidence for the effectiveness of medication in childhood trichotillomania, many clinicians prefer habit-reversal therapy as the initial, most benign intervention, with subsequent addition of a selective serotonin reuptake inhibitor or clomipramine, especially if comorbid depression, anxiety or obsessive compulsive disorder is present.[17]

In conclusion, recurrence in Rapunzel syndrome is an extremely rare manifestation, but may be anticipated if emotional disorder of the patient is not given due consideration and managed properly through psychiatric counselling and medications.

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