Rapunzel syndrome complicated with pancreatitis, intussusception and intestinal perforation

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Accepted 26 June 2022

SUMMARY

We report a case of a 4-year-old girl with limited financial resources, a background history marked by chronic abdominal discomfort and a positive *Helicobacter pylori* stool antigen test. The child presented with pallor, striking epigastric pain, nausea and vomiting. Blood tests reported high serum lipase levels. Investigations showed proof of nodular gastritis, intussusception and mild acute pancreatitis. The surgical procedure revealed Rapunzel syndrome complicated with intussusception and intestinal perforation, successfully treated. The postoperative course went uncomplicated.

BACKGROUND

Rapunzel syndrome is uncommon in children and can easily pass by undiagnosed, especially when associated with other diseases. Our case highlights how more than one complication can occur at the same time, making the early diagnosis crucial.

We present to the best of our knowledge the first case of a 4-year-old girl who presented with abdominal pain, and later found out that not only Rapunzel syndrome was the explanation for the course of her symptoms but also the combination of complications made of multiple diseases.

This underlines the importance of looking for eating disorders in children, and however rare Rapunzel syndrome and its complications are, it needs to be included in the differential diagnosis of chronic abdominal pain as well as the fact that it can be related to other diseases such as intussusception, pancreatitis and intestinal perforation.

CASE PRESENTATION

A 4-year-old girl, living in an impoverished area with no mental retardation, presented with a 3-month history of chronic abdominal discomfort mildly relieved by common medications, weight loss, and *Helicobacter pylori*-positive stool antigen test treated with antibiotics and Omeprazole. The non-amelioration of her clinical course and the sudden urge episodes of vivid epigastric pain had made the parents come to our emergency department. The pain was associated with nausea and occasional vomiting worsened by meals. On examination, the patient was asthenic and pale. She was afebrile, her weight and height were 11 kg (−1 SD) and 96 cm (−1 SD), respectively, her pulse rate was 110/min, her respiratory rate was 25/min and her blood pressure was 110/70 mm Hg. Her abdomen was mildly distended with rebound tenderness in the epigastric and periumbilical area and no palpable mass.

INVESTIGATIONS

Laboratory findings showed mild microcytic, hypochromic anaemia and iron deficiency. Her serum lipase levels were raised at 789 IU/L (normal up to 67 IU/L) but electrolyte concentrations, urea nitrogen, serum bilirubin, liver enzyme concentrations and serum albumin were normal. Acute pancreatitis is diagnosed based on clinical and biological criteria.

An abdominal ultrasound done showed an echogenic mass within the stomach and pylorus region, but also a target sign (peripheral hypoechoic ring) with a pseudo-kidney sign (central echogenicity) consistent with intussusception. No signs of peritonitis were seen.

An abdominal CT scan was ordered and revealed a heterogeneous gastric mass, thick gastric and intestinal wall (figure 1A,B), signs of mild acute pancreatitis (figure 1C) and ileocolic intussusception (figure 1D), with no sign of occlusion or perforation.

Upper endoscopy was necessary to rule out malignancy and decided to aid in the diagnosis of a trichobezoar inside the stomach (figure 2A,B). Nodular gastritis was also present.

Backward looking for further history, the parents informed us that the child used to pull her hair (trichotillomania) and sometimes eat it (trichophagia). Aside from this, they also mentioned frequent swallowing of pieces of sponge. This history made clear the series of events.

Hence, Rapunzel syndrome complicated with pancreatitis and intussusception was diagnosed.

DIFFERENTIAL DIAGNOSIS

Based on the ultrasound and CT findings, the diagnosis of a complicated tumour (probably a lymphoma) was done. It was only after the upper endoscopy was done that the trichobezoar was found out, highlighting the importance of looking for a history of eating disorder and putting a bezoar in our differential diagnosis.

TREATMENT

The girl underwent abdominal surgery, a medial supraumbilical incision was made and a dilated stomach was visualised. A 10-centimetre incision on the anterior surface of the stomach was done that led to the extraction of the brownish trichobezoar in one piece along with the long tail extending as far as the proximal colon, making a long multiple

To cite: Abourazzak S, Jerrar IO, Lakhdar Idrissi M, *et al*. *BMJ Case Rep* 2022;**15**:e247005. doi:10.1136/bcr-2021-247005

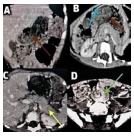

Figure 1 Coronal and axial contrast-enhaced CT scans (A,B) showing a heterogeneous mass (trichobezoar) inside the stomach (red arrow), and a gastric wall thickening (blue arrow), (C) showing a mild acute pancreatitis (yellow arrow) and (D) showing an image of bowel-into-bowel configuration typical of intussusception (green arrow).

intussusception, which was removed manually. In addition, a small jejunal perforation was found and was successfully treated (figure 3A,B). The other organs were macroscopically normal.

OUTCOME AND FOLLOW-UP

The postoperative course was normal with regression of the patient's serum lipase concentration. The child was discharged 1 week after having a psychiatric evaluation. On follow-up a month later, full blood count, electrolyte concentration and serum lipase levels were normal.

DISCUSSION

Abdominal pain is one of the most common presentations in the paediatric population.[1] Rapunzel syndrome is an extremely rare condition that is portrayed by the expansion of a trichobezoar, which is a foreign gastric formation due to gulped hair, from the stomach to the intestine or beyond.[2 3] Trichotillomania is an unusual urge to pull hair and is frequently associated with chewing and swallowing it (trichophagia).[4] Trichobezoars are an uncommon condition of bezoars (undigested material) constituted from a hair-ball mass coated with mucus and are often seen in girls with a history of psychiatric illness.[2–5] Rapunzel syndrome is an even rarer syndrome that is characterised by the extension of a trichobezoar from the stomach to the intestine or beyond.[2 3] This syndrome was first named after the Grimm Brothers' fairy tale in 1812 of the maiden Rapunzel with the long tresses who lowered her long hair from the prison tower to help a prince climb up and rescue her.[2] It was only in 1968 that it was first reported by Vaughan *et al* due to the long hair trapped in the gastrointestinal tract similar to the fairy tale.[4]

While abdominal pain is a frequent symptom and can be associated with several diseases, Rapunzel syndrome can be the cause. It can easily pass undiagnosed or misdiagnosed, and usually requires several investigations. Even if estimated that only 1% of patients with trichophagia develop a trichobezoar, a patient's history of trichophagia can certainly help the physician pose the diagnosis.[6] Also, patients with trichobezoars need a psychological evaluation and follow-up. Mental disorders, obsessive-compulsive disorder, abuse, and anorexia nervosa can cause trichotillomania, trichophagia, or pica due to consumption of non-nutritional substances and thus lead to iron deficiency anaemia, as in this case.[2 7 8] Unfortunately, it tends to be forgotten and is only asked retrospectively after making the diagnosis by an investigation.

Figure 2 (A,B) Endoscopic view of the trichobezoar.

Figure 3 (A) Trichobezoar removed in one piece during surgery. (B) Surgical specimen showing the trichobezoar and its long tail.

The majority of cases are seen in patients between 5 and 23 years old, so this case of a 4-year-old girl is a relatively early presentation.[7]

Clinically, it can remain asymptomatic for a long period as the bezoar enlarges in size and only appears later after a complication. Symptoms vary from abdominal pain (46.66%), nausea and vomiting (44.44%), to obstruction (20%), abdominal distention (8.88%) and weight loss (8.88%).[7] In this patient, abdominal pain and distention, nausea and vomiting were present. Large bezoars can be the source of multiple complications such as obstructive jaundice, gastrointestinal bleeding, acute pancreatitis, protein-losing enteropathy, perforation, appendicitis, peritonitis and intussusception.[2 3]

Other cases have been cited in literature combining more than one complication, such as a trichobezoar presenting with intussusception, and intestinal and biliary perforation,[5] or Rapunzel syndrome with cholangitis and pancreatitis,[7] especially that pancreatitis caused by obstruction of ampulla of Vater by the bezoar itself has been reported in only four cases of Rapunzel syndrome thus far and can be the only manifestation.[7 9]

While radiological investigations such as ultrasound, CT scan or dye study can help narrow the diagnosis, upper gastrointestinal endoscopy is, in most cases, performed to confirm the diagnosis. This gold-standard tool may provide direct visualisation and even therapeutic intervention in some cases.[6]

Treatment depends on multiple factors; size and location are one of them. Small trichobezoars can be removed endoscopically in contrast with large ones.[10] Large bezoars removed by endoscopy have shown to be related to more risks as fragmentation of the bezoar is nearly impossible, making surgery the ideal choice for large ones.[7] Rapunzel syndrome is another indication for surgery so that the long tail of hair can be removed. In this case, the exploration intraoperatively confirmed the radiological findings. Although laparoscopic removal can be done, the rarity of trichobezoars nevertheless, the Rapunzel syndrome, its complications and the lack of surgical experience in these cases may make the laparotomy a more reasonable choice.[2]

Even if recurrence is extremely rare, deep, comprehensive and psychiatric consultations are needed in all cases to understand and treat the patients. Treatment can be done with behaviour

Learning points

- Rapunzel syndrome is a rare type of trichobezoar and can be complicated with pancreatitis, intussusception and intestinal perforation.
- This clinical situation may need an urgent surgical intervention.
- The need to look for a history of trichophagia and other eating disorders should be considered when diagnosing young girls with chronic abdominal pain and iron deficiency anaemia.
- Psychological counselling plays a major role in the prevention of recurrence.

therapy and pharmacotherapy such as tricyclic antidepressants and selective serotonin reuptake inhibitors in some recurrent or resistant cases.[3 10]

Acknowledgements The authors are grateful to Dr Issam Taha, Professor Meryem Boubou, Professor Youssef Bouabdellah, Professor Khalid Khattala, Professor Moustapha Harrandou and Dr Soukaina Driouch.

Contributors IOJ, SA, MLI and MH contributed equally to the design and implementation of the case report, to the analysis of the results and to the writing of the manuscript.

Funding The authors have not declared a specific grant for this research from any funding agency in the public, commercial or not-for-profit sectors.

Competing interests None declared.

Patient consent for publication Parental/guardian consent obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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REFERENCES

1 Lynch KA, Feola PG, Guenther E. Gastric Trichobezoar: an important cause of abdominal pain presenting to the pediatric emergency department. *Pediatr Emerg Care* 2003;19:343–7.
2 Blejc Novak A, Zupančič Živa, Plut D, *et al*. Rapunzel syndrome: a rare form of Trichobezoar in the stomach with some extension into the small intestine. *Acta Dermatovenerol Alp Pannonica Adriat* 2018;27:155–7.
3 Khanna K, Tandon S, Yadav DK, *et al*. Rapunzel syndrome: a tail too long to tell! *BMJ Case Rep* 2018;5:bcr-2018-224756.
4 Lopes LR, Oliveira PSP, Pracucho EM, *et al*. The rapunzel syndrome: an unusual Trichobezoar presentation. *Case Rep Med* 2010;2010:1–3.
5 Baheti AD, Otjen JP, Phillips GS. A hairy situation: Trichobezoar presenting with intussusception, and intestinal and biliary perforation in a child. *Radiol Case Rep* 2017;12:42–4.
6 Gonuguntla V, Joshi D-D. Rapunzel syndrome: a comprehensive review of an unusual case of Trichobezoar. *Clin Med Res* 2009;7:99–102.
7 Vellaisamy R, Iyer S, Chandramohan SM, *et al*. Rapunzel syndrome with cholangitis and pancreatitis - A rare case report. *Open Med* 2020;15:1137–42.
8 Cannalire G, Conti L, Celoni M, *et al*. Rapunzel syndrome: an infrequent cause of severe iron deficiency anemia and abdominal pain presenting to the pediatric emergency department. *BMC Pediatr* 2018;18:125.
9 Dayasiri K, Rodrigues A, Lee A. Rapunzel syndrome presenting as acute pancreatitis, hypoproteinaemia and subsequent distal intestinal obstruction. *J Pediatr Surg Case Rep* 2020;59:101507.
10 Wang Z, Cao F, Liu D, *et al*. The diagnosis and treatment of Rapunzel syndrome. *Acta Radiol Open* 2016;5:205846011562766.