DOI: 103126/JNPS.V4113

Case Report

Rapunzel Syndrome: Dual Case Report and Literature Review

Christelle Destinval[1], Olivier Larmure[1], Jean-Louis Lemelle[2]

[1] Doctor, Paediatric Surgery Department, Nancy Regional University Hospital, Vandoeuvre-Les-Nancy, France.
[2] Professor, Paediatric Surgery Department, Nancy Regional University Hospital, Vandoeuvre-Les-Nancy, France.

Article History
Received on - 2023 Sep 24
Accepted on - 2024 Jan 03

Keywords:
Paediatric emergency and acute care; paediatric gastroenterology; paediatric surgery; psychiatric disorder; trichotillomania; trichophagia.

Online Access

DOI: https://doi.org/10.60086/jnps515

Abstract

A 10-year-old and 15-year-old females were referred to the paediatric emergency department. The former complained about acute abdominal pain and faecaloid vomiting, whereas the latter had chronic epigastric pain and iron deficiency anemia. Both patients had a history of trichotillomania and trichophagia. Abdominal CT scan demonstrated a limited oval lesion, molding the stomach. Both girls underwent an exploratory laparotomy with gastrotomy. A gigantic trichobezoar, molding the stomach and expanding to the small bowel was extracted, confirming the diagnosis of Rapunzel syndrome. They were discharged home after 16 and 13 days, respectively. No recurrence occurred.

Introduction

Rapunzel syndrome was first described in 1968.[1] It is a particular trichobezoar, most often found in young women aged under 30 years old,[2] with a history of trichotillomania and trichophagia.[3] Trichotillomania affects 1 in 2000 children, 30% of whom have associated trichophagia but only 1% will require surgical management, in case of trichobezoar.[1] Rapunzel syndrome compounds three characteristics: a gastric trichobezoar, with a tail extending at least to the jejunum, and symptoms of digestive obstruction.[4] Abdominal pain, nausea and vomiting are the main symptoms. On clinical examination, an abdominal palpable mass associated with hair loss may suggest a trichobezoar. Besides, bowel obstruction and peritonitis are complications leading to further investigations.[1] Abdominal X-ray demonstrates amorphous, granular, calcified, with air and solid material in the stomach. Furthermore, diagnosis can be confirmed by an upper gastrointestinal series (UGI).[5] Abdominal CT scan shows expansile, concentric and compressive mass in the stomach, containing air and debris. However, upper digestive endoscopy is the best exam to diagnose, and sometimes cure gastric bezoars.[2] Hence, the trichobezoar can be removed by endoscopy, laparoscopy, or laparotomy, depending on its size.[3] A hundred clinical cases have been reportedly treated by laparotomy, which remains the gold standard.[6] Adding to surgery, psychiatric treatment and psychological support are imperative to prevent recurrency.[1]

Case Presentation

Case 1

A ten-year-old girl, with long lasting history of trichotillomania and trichophagia, was admitted in the Paediatric Emergency Department with acute abdominal pain for a few days. She suffered from anorexia, diffuse abdominal pain and several episodes of fecaloid vomiting. A painful abdominal mass was palpable in the epigastric region. Blood tests showed hemoglobin (Hb) of 13.2 g / dl, leukocytes of 10600 / mm^3 and platelets of 205000 / mm^3. Abdominal CT scan highlighted a bulky trichobezoar molding the whole stomach. (Figure 1)

Correspondence

Christelle Destinval,
Paediatric Surgery Department,
Nancy Regional University Hospital,
Vandoeuvre-Les-Nancy,
France.
Email: christelledestinval@yahoo.fr

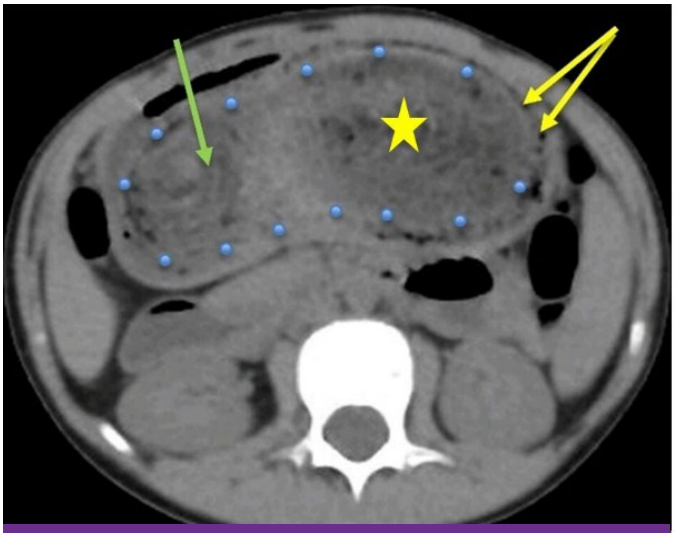

Fig 1: Abdominal CT scan: granular (green arrow), with air (yellow arrows), and solid material molding the stomach (blue dots).

We performed an upper digestive endoscopy which confirmed the diagnosis of trichobezoar, but its removal was impossible. The patient then underwent an exploratory laparotomy. A dilated stomach got a ten-cm-length gastrotomy on its anterior side, allowing to pull off a large trichobezoar molding the whole stomach. The pathognomonic hair tail confirmed the diagnosis of Rapunzel syndrome. The piece, characterized by a tangle of hairs, was sent for histological analysis. Size was 21 x 7 x 6 cm, weight 566 g. (Figure 2)

Fig 1: Rapunzel syndrome: trichobezoar molding the stomach with a tail molding the small bowel (red arrow).

A nasogastric tube was placed for seven days, and the patient was fed again on postoperative day (POD) eight. She was discharged home on POD 13. A three-month follow-up showed no recurrence. Psychiatric management was assigned during and after hospitalization.

Case 2

A 15-year-old female was referred at the Paediatric Emergency Department. She had six months history of epigastric abdominal pain, asthenia, pale skin associated with recent discovery of iron deficiency anemia (Hb 7.5 g / dL and mean corpuscular volume 58.7). On clinical examination, a mobile epigastric mass was found. The patient admitted to pulling and eating her hair for years. An abdominal CT scan showed an intragastric bezoar which size was 118 x 99 x 56 mm. An upper gastrointestinal endoscopy was performed the next day. A large gastric trichobezoar was found but couldn't be removed. (Figure 3)

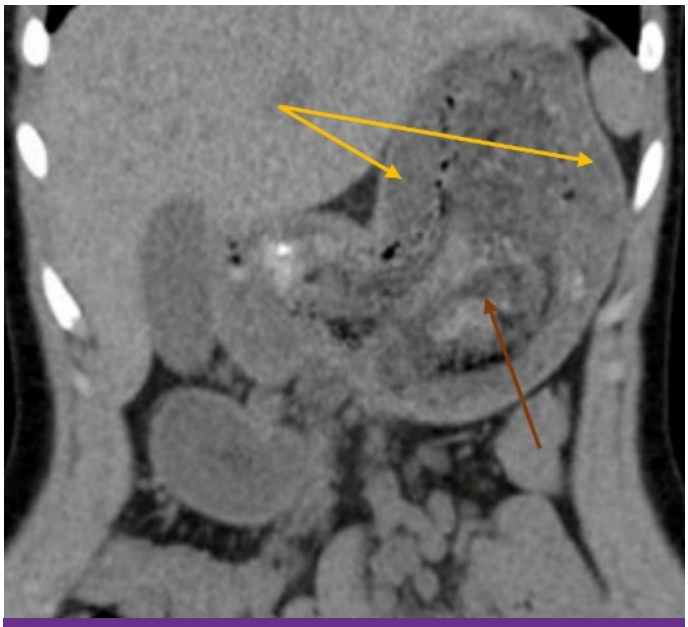

Fig 3: Abdominal CT scan: Frontal view. Stomach with swollen walls (orange arrows), due to inflammation, and containing a heterogenic trichobezoar (brown arrow).

She underwent a laparotomy, gastrotomy and retrieval of the trichobezoar which molded the stomach, the duodenum and had the typical tail. (Figure 4)

Fig 4: Rapunzel syndrome.

She was taken care of in the intensive care unit for two days then, was transferred to the Paediatric Surgery Department for 14 days. Postoperative outcome was a urinary infection treated with antibiotics. Afterwards, she was transferred to a nearer hospital to have a better psychiatric management but the patient was lost to follow-up.

Discussion

Rapunzel syndrome is a gastric trichobezoar with a hair tail which spreads at least through the small bowel with signs of bowel obstruction.[3] A hundred cases have already been reported.[3,6] They were the first two cases occurring in our Department. To highlight the importance of early diagnosis and surgical management, Figure 5 describe the pathophysiology of trichobezoar.

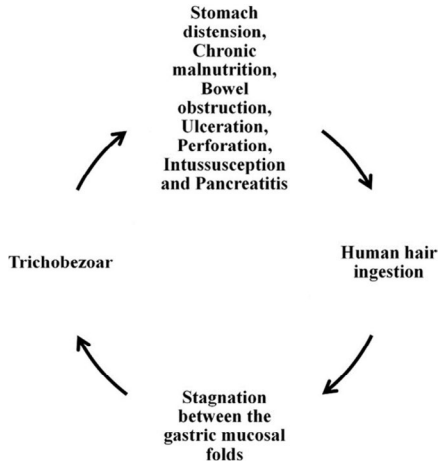

Fig 5: Pathophysiology of a trichobezoar.

Risk factors of trichobezoar are mental disorders, depression, anorexia,[6] and gastric surgery.[5] Clinical signs are abdominal pain (37%), nausea and vomiting (33%), intestinal obstruction (25%),[3] early satiety, and weight loss.[5] In our cases, the second patient suffered from anemia.

Although X-ray exams and upper gastrointestinal studies with contrast medium used to help diagnosis in the past, nowadays abdominal CT scan is the most relevant exam, and demonstrates a heterogenous intragastric mass, with air and debris, which can reach the small or the large bowel. Moreover, adding contrast medium may highlight ulceration or perforation.[2,5,7] Finally, human hair ingestion stagnates between the gastric mucosal folds, causing stomach distension, chronic malnutrition, bowel obstruction, ulceration, perforation, intussusception and pancreatitis. Though an endoscopic approach may confirm diagnosis, it is not always successful (only in 5% of cases).[3] In our cases, none of the trichobezoars could be endoscopically removed because of their huge size. Consequently, exploratory laparotomy with gastrotomy and / or enterotomy is the gold standard to remove the trichobezoar.[1,3]

Nevertheless, no healing can be achieved without psychiatric management. A panel of tools can be of use such as antidepressants and behavioral therapy. The first patient went on with psychological therapy and is fine but the second one did not come to the follow-up consultation. Recurrence depends upon the efficacy of psychiatric treatments.[1]

Conclusion

Rapunzel syndrome is a rare type of trichobezoar. Paediatricans should consider this diagnosis in young females suffering from abdominal pain, nausea, vomiting, anemia, associated with psychiatric disorders or trichotillomania. UGI endoscopy and Abdominal CT scan leads to diagnosis. But surgery is the treatment and gastrotomy is essential to remove the hair mass. No healing can be hoped without psychiatric management.

References

1. Pinheiro Martins de Almeida e Souza G, Eugênio de Sousa Lima M, Portela Muniz A, Albuquerque Landim J, César Gadelha de Abreu Filho A. Rapunzel syndrome with anal protusion of trichobezoar. J Pediatr Surg Case Rep. 2019 Nov 1.
DOI: 10.1016/j.epsc.2019.101312

2. Isberner RK, Carlos Antônio da Silva C, Bruno Lorenzo S, Gustavo Becker P, Renan O. Giant Gastric Trichobezoar: A Case Report and Literature Review. Radiol Bras, vol. 43, no. 1, 2010, pp. 63-65.
DOI: 10.1590/S0100-39842010000100015

3. Lyons D. Large gastric trichobezoar causing failure to thrive and iron deficiency anaemia in an adolescent girl: a case report emphasising the imaging findings and review of the literature. BJR Case Rep. 2019 Jan 11;5(2):20180080 DOI: 10.1259/bjrcr.20180080

4. Naik S, Gupta V, Naik S, Rangole A, Chaudhary AK, Jain P, et al. Rapunzel Syndrome Reviewed and Redefined. Dig Surg. 2007;24(3):157-61. DOI: 10.1159/000102098 PMID: 17476105

5. Phillips MR, Zaheer S, Drugas GT. Gastric trichobezoar: case report and literature review. Mayo Clin Proc. 1998 Jul;73(7):653-6. DOI: 10.1016/S0025-6196(11)64889-1 PMID: 9663194

6. Gorter RR, Kneepkens CMF, Mattens ECJL, Aronson DC, Heij HA. Management of trichobezoar: case report and literature review. Pediatr Surg Int. 2010 May;26(5):457-63. DOI: 10.1007/s00383-010-2570-0 PMID: 20213124 PMCID: PMC2856853

7. Hamid M, Chaoui Y, Mountasser M, Sabbah F, Raiss M, Hrora A, et al. Giant gastric trichobezoar in a young female with Rapunzel syndrome: case report. Pan Afr Med J. 2017 Aug 4;27:252. DOI: 10.11604/pamj.2017.27.252.9110 PMID: 29187921 PMCID: PMC5660300