

Physiology and Anatomy

Elixir Physio. & Anatomy 150 (2021) 55127-55128

Elixir
ISSN: 2229-712X

Rapunzel Syndrome with Intestinal Intussusception: A Case Report

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ARTICLE INFO

Article history:

Received: 16 November 2020;

Received in revised form:

30 December 2020;

Accepted: 9 January 2021;

Keywords

Trichobezoar,
Rapunzel Syndrome,
Intussusception,
Children.

ABSTRACT

Rapunzel syndrome is defined by the presence of a gastric trichobezoar that extends beyond into the small intestine. It is an uncommon pathology, which mainly affects female patients. The association with intussusception is rare. Here we report the case of a 5-year-old girl who presented to the emergency room with abdominal pain and was diagnosed with Rapunzel syndrome complicated by intussusception. She was then operated on with satisfying post-operative results and recovery.

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Introduction

Rapunzel syndrome is a term used to describe gastric trichobezoar that extends into the duodenum and small bowel [1]. Bezoars are an uncommon cause of abdominal pain that can lead, in neglected cases, to complications such as intestinal obstruction, perforation and peritonitis. Jejunojejunal intussusception is a rare cause of small intestine obstruction in patients presenting with Rapunzel syndrome [2]. Here we report one such case that was diagnosed and managed successfully

Case presentation

Headings, or heads, are organizational devices that guide the reader through your paper. There are two types: component heads and text heads.

A 5 years old female patient presented to the emergency department with recently worsening epigastric abdominal pain, vomiting and constipation. She had no medical problems or surgeries. There was no history of fever, hematemesis, melena, or previous such episodes of pain and vomiting. Initial examination revealed no acute distress. Abdominal examination found a mild generalized abdominal tenderness without signs of peritonitis or palpable lump. Routine hematological and biochemical screening was normal.

Contrast enhanced computed tomography of the abdomen showed a large intragastric ovoid, heterogeneous, well-circumscribed mass containing air bubbles. This mass was non enhancing and extended to the duodenum and proximal jejunum. Moreover, there was a target image on the left hypochondrium, suggestive of an intussusception. A small amount of peritoneal effusion was noted. There were no signs of bowel obstruction complications.

The patient underwent a laparotomy. The trichobezoar was removed throughout an anterior gastrotomy and the intussusception was reduced manually. The post-operative period was uneventful.

Discussion

Bezoar is a packed mass of indigested material found trapped in the gastrointestinal tract, mainly in the stomach.

Trichobezoars consist of ingested hair and are mainly seen on young females with psychiatric disorders leading to trichotillomania and trichophagia. Rapunzel syndrome, originally described by Vaughan et al. in 1968, defines a rare form where gastric trichobezoar extends into the small intestines. It was named after a fairytale of a princess called Rapunzel, who let her long hair down from her prison tower to her prince so he can climb and rescue her [1, 3, 4]

Clinical presentation may include a palpable abdominal mass, abdominal pain, nausea, vomiting, weakness, weight loss, anorexia, constipation or diarrhea [2]. Complications may occur, mainly in neglected cases, such as ulceration or perforation of the gastrointestinal tract, small bowel intussusception, obstructive jaundice and pancreatitis [5, 6]

Enhanced CT scan on the abdomen is the modality of choice as it is widely available. It confirms the diagnosis, delineates the extension of a trichobezoar and detects potential complications [1]. Trichobezoar appear on CT as rounded or ovoid, well defined, intraluminal, non-enhanced masses, with a spotted pattern due to air being trapped between hair [7, 8]

Bowel intussusception appears as the classic target sign.

CT scan can also show signs of gastrointestinal tract ischemia, bowel perforation, peritonitis.

Ultrasonography may show a floating heterogeneous mass lying inside the stomach, however its role remains limited [1]

Open surgical removal is the method of choice for large trichobezoar treatment.

Psychiatric support is essential and may take a long time to control the patient behavior and prevent relapses [9].

Conclusion

Rapunzel syndrome presenting with bowel intussusception is a rare condition. Enhanced CT scan helps confirm the diagnosis and guides the management approach.

Conflict of interest

The authors do not declare any conflict of interest.

Author's contributions

All authors contributed to this work. All authors have read and approved the final version of the manuscript.

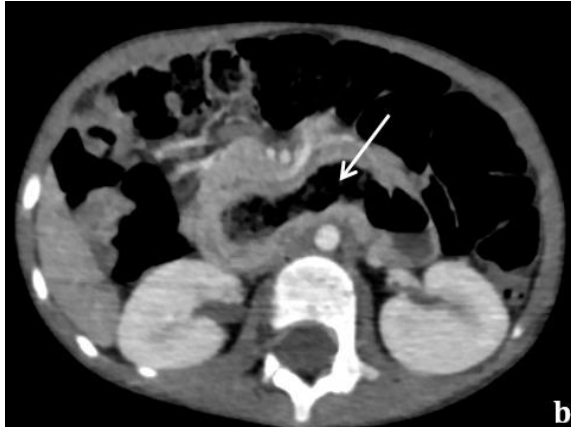
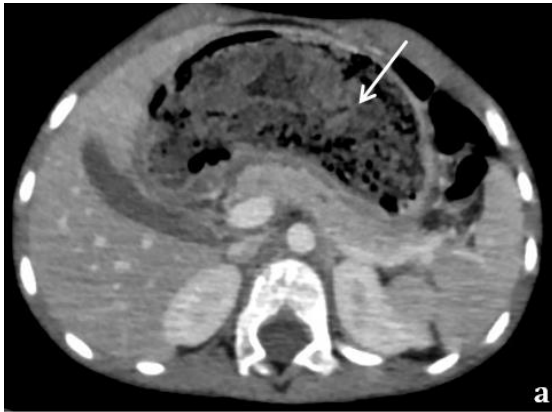


Figure 1. Abdominal CT on axial reconstructions showing gastric trichobezoar (a) extending into the duodenum and jejunum (b) (arrows)

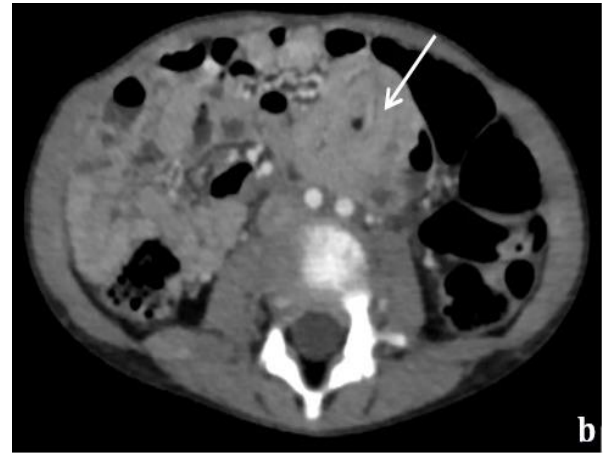
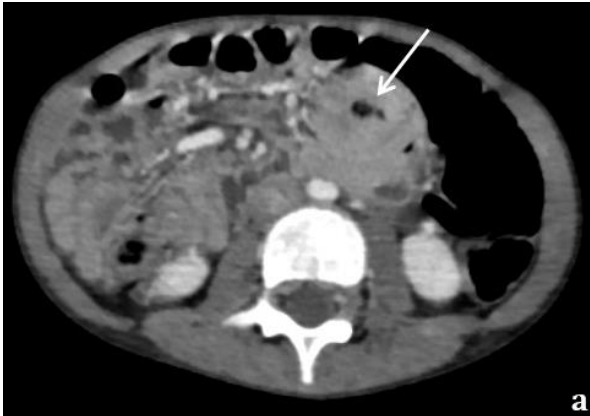


Figure 2. Abdominal CT on axial reconstructions (a, b) showing jejunojejunal intussusception (arrow).

References

- [1] Phillips, M.R., S. Zaheer, and G.T. Drugas, Gastric trichobezoar: case report and literature review. *Mayo Clin Proc*, 1998. 73(7): p. 653-6.
- [2] Naik, S., et al., Rapunzel syndrome reviewed and redefined. *Dig Surg*, 2007. 24(3): p. 157-61.
- [3] Marwah, S., et al., Rapunzel syndrome presenting as jejuno-jejunal intussusception. 2015. 8(4): p. 202-206
- [4] Lesser, R. (1999). *Hansel and Gretel*. Penguin.
- [5] Feldman, M., & BRANDT, L. J. (2011). *Sleisenger and Fordtran's Gastrointestinal and Liver Disease: pathophysiology/diagnosis/management*. v. 1, v. 2. In *Sleisenger and Fordtran's Gastrointestinal and Liver Disease: pathophysiology/diagnosis/management*. v. 1, v. 2 (pp. 1098-1098).
- [6] Gorter, R., et al., Management of trichobezoar: case report and literature review. 2010. 26(5): p. 457-463.
- [7] Ripollés, T., García-Aguayo, J., Martínez, M. J., & Gil, P. (2001). Gastrointestinal bezoars: sonographic and CT characteristics. *American Journal of Roentgenology*, 177(1), 65-69.
- [8] Czerwińska, K., Bekiesińska-Figatowska, M., Brzewski, M., Gogolewski, M., & Wolski, M. (2015). Trichobezoar, rapunzel syndrome, tricho-plaster bezoar—a report of three cases. *Polish Journal of Radiology*, 80, 241.
- [9] Duncan, N. D., Aitken, R., Venugopal, S., West, W., & Carpenter, R. (1994). The Rapunzel syndrome. Report of a case and review of the literature. *The West Indian medical journal*, 43(2), 63-65.