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Review Article

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Rapunzel Syndrome and the Importance of Early Diagnosis: An Article Review

Ian Caldeira Ruppen[1*], Jamile Diogo de Araujo[2] ,Leandro Hideki Otani[2], Vitor Augusto Olivari do Carmo[1], Isabela Matias Cian[1], Gabriel Botequia Zanatta[1], Yasmin Cavatorta Jannani[1], Felicia Satie Ibuki Otani[2], Gabriel Petermann[3], Lara Beatriz Dallaqua Bitiati[1] Alana Reigota da Costa Rosa[1], Geórgia Verona Cruz[2], Evili Rathier Sandri[1], Marcela Castrequini Guimarães do Vale[1], Rafaela Castrequini Guimarães do Vale[1], Valentina Verona Cruz[4], Larissa da Rosa Piccoli[1], Maria Clara Malheiros Vizzotto[4], Guilherme Albuquerque Nicolau[4], Geovani Almeida Gois[5], Antonio Fabiano Morelli Filho[1], Júlia Alvares Dal' lago[6], Amanda Larissa Zotarelli Pasquali[1], Isabela Viana Veronez[1], Camila Yuri Kakumoto Tsuneto[1]

[1]Centro Universitário Ingá – Uningá, Maringá, PR, Brazil.
[2]Instituto Maringá de Imagem, Maringá, PR, Brazil.
[3]Universidade Anhanguera Uniderp, Brazil.
[4]Faculdade Cesumar - Unicesumar Maringá, Paraná, Brazil.
[5]Universidade Federal do Maranhão, Brazil.
[6]Ulbra- Universidade Luterana do Brasil Canoas- Rio Grande do Sul, Brazil.

***Corresponding author:** Ian Caldeira Ruppen, Centro Universitário Ingá-UNINGÁ, **Email:** Ian2ruppen@gmail.com; **ORCID:** https://orcid.org/0000-0003-1706-1662

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Abstract

Rapunzel Syndrome is a rare condition characterized by the presence of a trichobezoar (accumulation of hair) in the gastrointestinal tract, with an extension that can reach the small intestine and even the colon. This syndrome is particularly common in patients suffering from trichotillomania (compulsion to pull out hair) and trichophagia (ingestion of hair). The lack of early diagnosis can lead to severe complications such as intestinal obstruction, perforation, or hemorrhage. This review examines the importance of early diagnosis for the appropriate management of this rare and potentially fatal condition.

Keywords: *Psychiatry, Trichophagia, Trichobezoar, Rapunzel Syndrome. Pediatrics.*

Introduction

Rapunzel Syndrome is a rare medical condition first described in 1968 by Vaughan et al., characterized by the formation of trichobezoars, masses of hair accumulated in the gastrointestinal tract, which may extend beyond the stomach and reach the small intestine or even the colon. It is a rare and extremely serious condition marked by the formation of gastric trichobezoars (hair accumulations in the stomach) that can extend into the small intestine, potentially causing gastrointestinal obstruction. The disease is generally associated with trichotillomania (the compulsive habit of pulling one's own hair) and trichophagia (the habit of ingesting hair), which often occur in patients with psychiatric disorders. This condition is most commonly observed in young female patients who may suffer from trichotillomania and trichophagia. The syndrome's name refers to the fairy tale character Rapunzel, known for her long hair, which metaphorically alludes to the accumulation of hair extending through the digestive system. The syndrome is more prevalent in young women, and early diagnosis is essential to prevent serious complications, such as intestinal obstructions, perforations, or even sepsis. The literature emphasizes the importance of early identification and rapid intervention, as delayed treatment can lead to invasive surgical procedures and an increased risk of complications. The symptoms of the syndrome can range from vague abdominal pain and weight loss to more acute situations such as intestinal obstruction or perforation. Due to the insidious nature of its development, early diagnosis is critical to prevent fatal complications. However, as it is a rare condition, diagnosis is often delayed, and the syndrome is discovered only when surgical intervention becomes inevitable.

Objectives

The present study aims to highlight the importance of early diagnosis, still infrequent, of Rapunzel Syndrome by describing its clinical characteristics and its main complication, the trichobezoar, to facilitate a more accurate differential diagnosis.

Materials and Methods

A review of articles published in the PUBMED, ScienceDirect, and Scielo databases was conducted to support this study.

Discussion

Several studies indicate that Rapunzel Syndrome often goes unnoticed in its early stages, mainly due to the silent nature of the condition and the lack of patient reports regarding hair ingestion. Early symptoms may include abdominal pain, nausea, vomiting, and weight loss, which can be mistaken for other gastrointestinal pathologies. Thus, an early diagnosis requires a combination of careful history-taking, imaging studies such as computed tomography or endoscopy, and attention to clinical signs. In a review of clinical cases, it is noted that patients diagnosed early present a lower need for more invasive surgical interventions, such as laparotomy, and have a better clinical outcome. Furthermore, a multidisciplinary approach, involving psychologists and psychiatrists to treat underlying disorders such as trichotillomania and trichophagia, is crucial for preventing recurrences. This underscores the relevance of early diagnosis not only for immediate treatment but also for the patient's psychosocial rehabilitation. Studies also indicate that delayed diagnosis can lead to the formation of giant trichobezoars, which, upon reaching the small intestine, can result in complete obstruction, causing intense pain, intractable vomiting, and eventually intestinal perforation. These cases may require complex surgical removal and prolonged postoperative care. In contrast, in early diagnoses, removal can be performed endoscopically, with less risk and faster recovery.

The pathogenesis of Rapunzel Syndrome involves continuous ingestion of hair, which is indigestible by the gastrointestinal tract. The resulting trichobezoar typically forms in the stomach, but in cases of Rapunzel Syndrome, it extends beyond the pylorus, entering the small intestine and, in some cases, even the colon. Prolonged hair ingestion leads to the progressive accumulation of this mass, which can cause symptoms ranging from nonspecific abdominal pain and nausea to total intestinal obstruction, perforation, or even internal hemorrhage. Late diagnosis is associated with serious complications such as intestinal obstruction, malabsorption syndrome, anemia, and even gastric perforation. In advanced cases, extensive intestinal resections and emergency interventions may be required. Mortality is rare but possible if not properly treated. Treatment is predominantly surgical, as trichobezoars are often too large to be removed endoscopically in most cases. After surgery, psychiatric follow-up is essential to address underlying conditions such as trichotillomania to prevent recurrence. Cognitive-behavioral therapy and the use of anxiolytic or antidepressant medications may be necessary for treating associated psychiatric conditions.

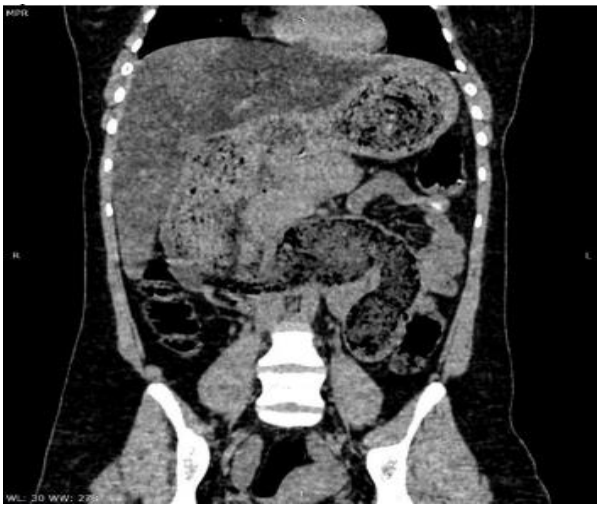

Figure 1: Abdominal Computed Tomography showing intragastric and intestinal bezoar.

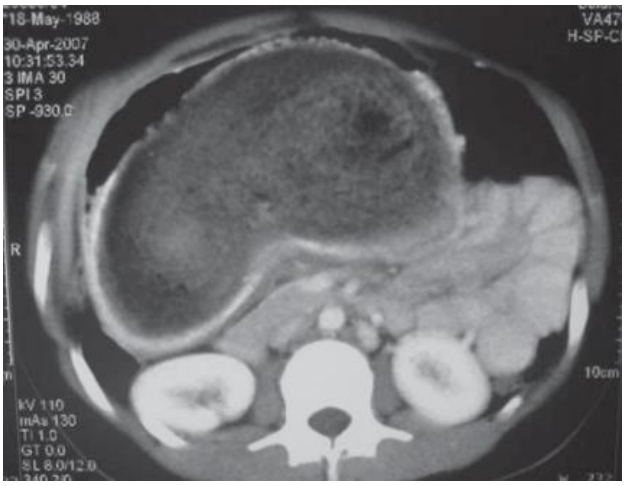

Figure 2: Axial computed tomography scan at the gastric body level demonstrating a "whorled mass" occupying its entire lumen.

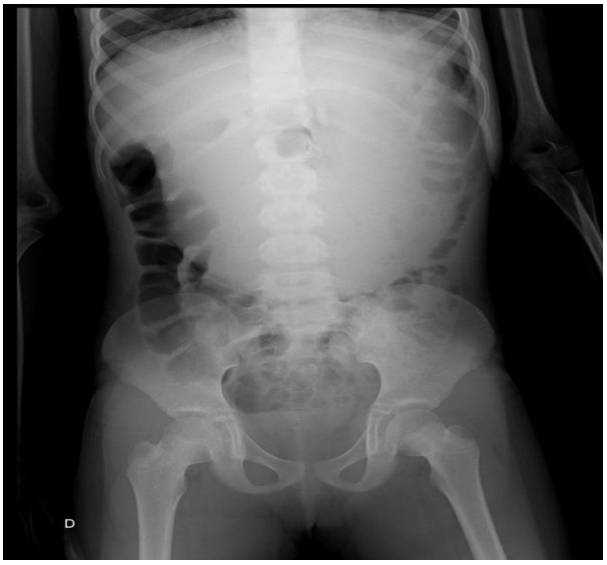

Figure 3: Panoramic radiography of a contrasted stomach. Gastric distension with loss of normal mucosal relief definition, little contrast opacification, with linear images enveloped by the contrast medium within its interior.

Figure 4: Surgical specimen post-removal of the trichobezoar

Conclusion

Although rare, Rapunzel Syndrome is a medical condition that can have serious consequences if not diagnosed early. Continuous hair ingestion leads to the formation of trichobezoars that can cause a variety of gastrointestinal symptoms, often leading to the need for emergency surgical intervention. Early diagnosis, through clinical suspicion based on patient history and confirmation by imaging exams, can prevent serious complications. After surgical treatment, psychiatric follow-up is crucial to prevent recurrence, as trichotillomania and trichophagia are frequently associated with this condition. Awareness of this rare syndrome is essential to reduce diagnostic delays and improve patient outcomes.

References

1. VAUGHAN, E. D. et al. The Rapunzel syndrome: an unusual complication of intestinal bezoar. *Journal of Pediatric Surgery*, v. 4, n. 5, p. 517-524, 1968.
2. NAIK, S. et al. Rapunzel syndrome reviewed and redefined. *Digestive Surgery*, v. 24, n. 3, p. 157-161, 2007.
3. SEHGAL, V. N. et al. Trichotillomania with tricophagia leading to trichobezoar (Rapunzel syndrome). *International Journal of Dermatology*, v. 49, n. 10, p. 1198-1200, 2010.
4. McKEOWN, D. J. et al. The Rapunzel syndrome: a case report and review of the literature. *Irish Journal of Medical Science*, v. 175, n. 1, p. 69-71, 2006.
5. PHILLIPS, M. R. et al. Gastric trichobezoar: Case report and literature review. *Mayo Clinic Proceedings*, v. 61, n. 8, p. 637-641, 1986.
6. GONUGUNTLA, V.; JOSHI, D. D. Rapunzel syndrome: a comprehensive review of an unusual case of trichobezoar. *Clinical Medicine & Research*, v. 7, n. 3, p. 99-102, 2009.
7. IFTIKHAR, P. M. et al. Rapunzel syndrome in a young girl: A case report and review of the literature. *Case Reports in Surgery*, v. 2018, 2018, p. 1-4.
8. MOHITE, P. N. et al. Rapunzel syndrome: a review of the literature and report of a case. *Saudi Journal of Gastroenterology*, v. 16, n. 3, p. 209-212, 2010.
9. GORTER, R. R. et al. Management of trichobezoar: case report and literature review. *Pediatric Surgery International*, v. 26, n. 5, p. 457-463, 2010.
10. ALTINTOPRAK, F. et al. Rapunzel syndrome: an unexpected cause of acute appendicitis. *Journal of Emergency Medicine*, v. 43, n. 6, p. e395-e397, 2012.
11. HASHMANI, N. et al. A rare cause of gastric outlet obstruction: Rapunzel syndrome. *Cureus*, v. 11, n. 10, e5971, 2019.
12. TIWARI, K. et al. A case report of Rapunzel syndrome: A rare form of trichobezoar extending into the small intestine. *Journal of Clinical and Diagnostic Research*, v. 10, n. 7, p. PD05-PD06, 2016.