Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Rapunzel Syndrome in a Young Girl: A Case Report

Zouaki Zakaria[1*], El Biadi Mohamed[1], Slioui Bader[1], Bellasri Salah[1], Hammoune Nabil[1], Atmane Mehdi[1], Mouhsine Abdelilah[1], Gharbi Khalid[2]

[1]Department of Radiology, Military Hospital Avicenne, Marrakech, Morocco
[2]Department of Hepato-Gastroenterology, Military Hospital Avicenne, Marrakech, Morocco

DOI: 10.36347/sjmcr.2023.v11i09.005 | **Received:** 11.07.2023 | **Accepted:** 27.08.2023 | **Published:** 03.09.2023

***Corresponding author:** Zouaki Zakaria
Department of Radiology, Military Hospital Avicenne, Marrakech, Morocco

Abstract **Case Report**

Rapunzel Syndrome is an extremely rare complication of gastric trichobezoars. Here, we report an unusual case of a 16-year-old girl presenting with abdominal pain and vomiting. Clinical examination revealed a tender abdominal mass. Abdominal computed tomography showed a well-defined heterogeneous mass occupying the entire gastric cavity with extension into the third part of the duodenum. A diagnosis of giant trichobezoar was suspected after further questioning revealed trichotillomania, trichophagia, and onychophagia. Endoscopic removal of the trichobezoar was unsuccessful, and only a few hair fibers were retrieved for diagnostic purposes. Subsequently, an exploratory laparotomy was performed. An antral gastrotomy was carried out, and a 25x10x7 cm trichobezoar was extracted without complications. The patient had an uneventful postoperative recovery and was referred to psychiatry. Rapunzel Syndrome is a rare trichobezoar typically found in young women, often associated with an underlying psychiatric disorder. Recurrence is generally infrequent, but long-term psychiatric follow-up is crucial in preventing trichobezoar recurrence.

Keywords: Rapunzel Syndrome, trichobezoar, computed tomography, gastrotomy, psychiatry.

INTRODUCTION

Trichobezoar is a rare condition characterized by the unusual presence of hair, in the form of a solid mass, within the gastrointestinal tract of certain patients with trichotillomania, trichophagia, and learning disorders or psychiatric illnesses [1, 2]. Clinically, most trichobezoars are typically found in young females aged 15 to 20 years due to their nonspecific presentations or even the absence of clinical symptoms in the early stages [3, 4]. Here, we report an unusual case of a 16-year-old girl presented with a giant trichobezoar revealed by abdominal pain.

CASE PRESENTATION

A sixteen-year-old girl presented to our gastroenterology department with intermittent abdominal pain and vomiting for the past six months. It is worth noting that her mother admitted that her daughter had developed an irresistible habit of pulling and swallowing her hair since she was twelve years old, which mainly occurred unconsciously during sleep or when she was alone.

On physical examination, the patient appeared pale and malnourished, with palpation revealing a mobile, firm, and tender mass in the upper left quadrant without signs of complications. Her laboratory workup showed microcytic hypochromic anemia (hemoglobin 9.2 g/dL), hypoalbuminemia (43 g/L), and hypochloremia (1.45 g/L) with a normal blood ionogram.

Computed tomography (CT) revealed massive gastric distension with a well-defined, heterogeneous mass occupying the entire gastric cavity with an extension into the third part of the duodenum (Figure 1). Esophagogastroduodenal endoscopy (EGD) confirmed the presence of a large trichobezoar occupying the entire gastric cavity and extending through the pylorus into the duodenum, with only a few hair fibers visible (Figure 2).

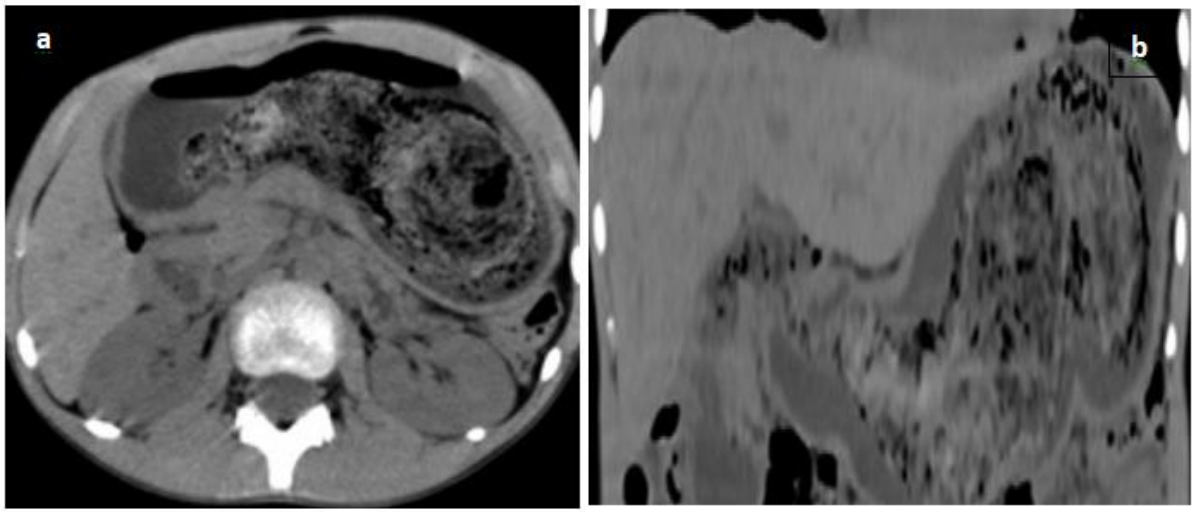

Figure 1: Axial (a) and coronal (b) CT scan sections showing the extension of a trichobezoar from the stomach to the second part of the duodenum

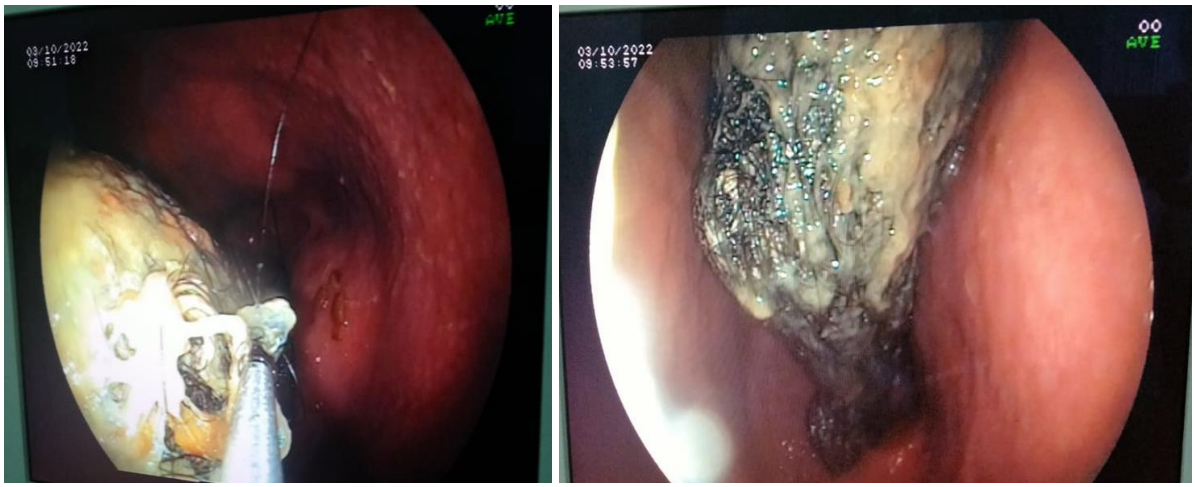

Figure 2: Presence of a large trichobezoar during esophagogastroduodenal endoscopy (EGD)

The patient underwent surgical intervention, and an upper midline incision was made to perform an antral gastrotomy. A giant trichobezoar was identified and removed (Figure 3). There was a long tail of hair extending through the pylorus. The postoperative course was uneventful, and psychiatric management was arranged after her discharge.

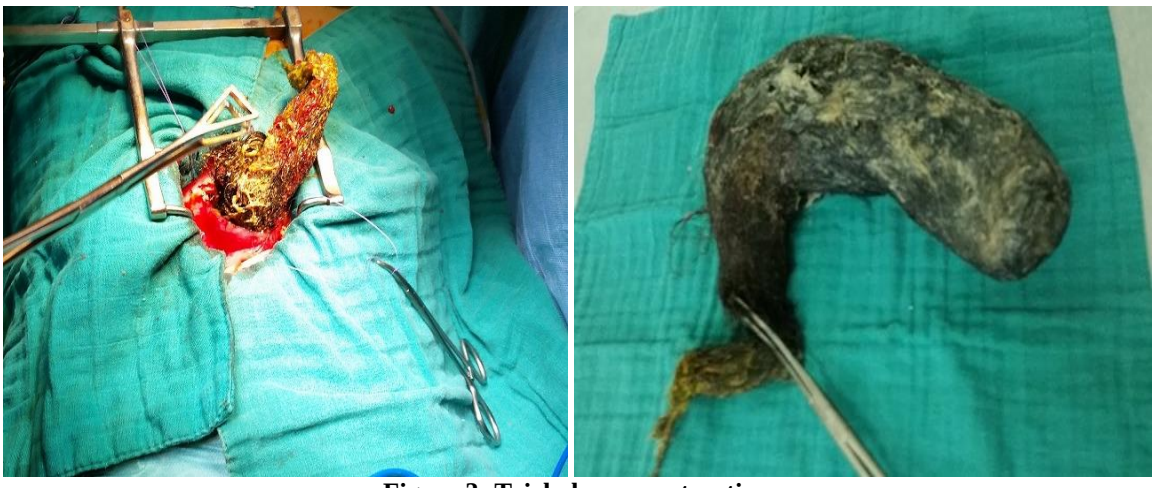

Figure 3: Trichobezoar extraction

DISCUSSION

A bezoar is an agglomeration of food or foreign materials that accumulate and remain in the gastrointestinal tract. They have been classified into four types: phytobezoars (caused by vegetables), trichobezoars (caused by hair), lactobezoars (caused by curdled milk), and miscellaneous (caused by medications, tissue paper, shellac, tar, sand, or mushrooms) [5]. In 1779, Baudamant described the first case of trichobezoar [6].

It is a rare condition, with females being more affected (90% of cases), and the age of onset is in 80% of cases below 30 years, with a peak incidence between 10 and 19 years [7]. It usually occurs in emotionally disturbed or intellectually challenged young individuals who have an irresistible urge to pull out and swallow their hair, known as trichotillomania and trichophagia [8].

Trichobezoars are inefficiently propelled by peristalsis due to their smooth surface and undigested keratinous substance. As a result, hair becomes entangled in a ball and is retained in the folds of the upper digestive tract, continuing to grow in size and weight, leading to devastating complications such as gastric ulcers, perforation, or even death [9].

Rapunzel Syndrome is encountered as a rare complication of trichobezoar formation in which the gastric bezoar extends into the duodenum and small intestine, which may increase the risk of complications such as obstruction and perforation.

Less than a hundred cases of Rapunzel Syndrome have been reported in the literature since its initial description in 1968 [10-12]. Clinically, patients frequently present with nonspecific symptoms and signs, including loss of appetite, weight loss, vomiting, and abdominal pain [13, 14].

During clinical examination, a well-defined, smooth, firm, and mobile abdominal mass in the epigastric region is found in 85% of patients. Anemia and hypoalbuminemia have also been described [15, 16].

The diagnosis relies on EGD, which remains the examination of choice, allowing visualization of hair tangles pathognomonic of trichobezoar. In some cases, EGD may have therapeutic value by enabling the endoscopic extraction of small trichobezoars [17]. However, due to its size, this extraction is often impossible, as in the case of our patient, and any attempt carries a risk of serious esophageal injury.

Abdominal X-ray without preparation may show a dense or heterogeneous rounded mass with or without calcification projecting onto the gastric area [18]. Abdominal ultrasound can diagnose the condition in only 25% of cases, visualizing a superficial, hyperechoic, curvilinear band with a clear posterior acoustic shadow [19]. Upper gastrointestinal contrast study reveals a mobile intraluminal gastric gap with convex borders, which may extend into the duodenum [6]. Small bowel follow-through completes the exploration of the intestine to look for continuous distal extension or detached fragments [1]. Abdominal CT scan may show a variable-sized mass, heterogeneous, occupying almost the entire gastric lumen, and composed of multiple concentric circles of different densities distributed in onion bulb-like patterns. Two pathognomonic and consistent signs are the presence of tiny air bubbles scattered within the mass and its lack of attachment to the gastric wall [20].

Various therapeutic modalities have been proposed to treat trichobezoars. Non-surgical extraction through endoscopy or dissolution with papain syrup often fails and may lead to serious complications [21]. Surgical options have been modified with the advent of laparoscopy, and its effectiveness in combination with a small laparotomy has been reported [22]. However, the size of the mass limits this approach in this case. We chose a relatively small antral gastrotomy through a small midline incision [23]. Additionally, psychiatric management is often necessary for patients [7].

CONCLUSION

Rapunzel Syndrome is a rare trichobezoar, commonly found in young women, often with an underlying psychiatric disorder. Management requires gastrotomy. Psychiatric evaluation and long-term follow-up are recommended as a regular part of treatment to prevent recurrences.

REFERENCES

1. Al-Wadan, A. H., Al-Absi, M., Al-Saadi, A. S., & Abdoulgafour, M. (2006). Syndrome de Raiponce. *Med saoudien J*, 27, 1912-1914.
2. Balik, E., Ulman, I., Taneli, C., & Demircan, M. (1993). Le syndrome de Raiponce: rapport de cas et revue de la littérature. *Eur J Pediatr Surg*, 3, 171-173.
3. Dalshaug, G. B., Wainer, S., & Hollaar, G. L. (1999). Le syndrome de Raiponce (trichobézoard) provoquant une invagination atypique chez un enfant: un rapport de cas. *J Pediatr Surg*, 34, 479-480.
4. Dindyal, S., Bhuva, N. J., Dindyal, S., Ramdass, M., & Narayansingh, V. (2008). Trichobezoar présentant le « signe de virgule » dans le syndrome de Raiponce: un rapport de cas et une revue de la littérature. *Affaires J*, 1, 286.
5. Ibuowo, A. A., Saad, A., & Okonkwo, T. (2008). Trichobézoar gastrique géant chez une jeune femelle. *International journal of surgery (Londres, Angleterre)*, *6*(6), E4-6.
6. Naik, S., Gupta, V., Naik, S., Rangole, A., Chaudhary, A. K., Jain, P., & Sharma, A. K. (2007). Rapunzel syndrome reviewed and redefined. *Digestive surgery*, *24*(3), 157-161.

7. Ousadden, A., Mazaz, K., Mellouki, I., & Taleb, K. A. (2004, May). Le trichobézoard gastrique: une observation. In *Annales de chirurgie* (Vol. 129, No. 4, pp. 237-240). Elsevier Masson.
8. Vaughan, E. D., Jr, Sawyers, J. L., & Scott, H. W., Jr. (1968. Le syndrome de Raiponce. Une complication inhabituelle du bézoard intestinal. *Chirurgie*, *63*(2), 339-343.
9. Marique, L., Wirtz, M., Henkens, A., Delchambre, E., Rezaï, M., & Venet, C. (2017). Perforation gastrique due au trichobézoar géant chez un enfant de 13 ans. *J Gastrointest Surg*, 21, 1093-1094
10. Vila, S., García, C., Piscoya, A., De Los Ríos, R., Pinto, J. L., HuertaMercado, J., & Bussalleu, A. (2009). Giant gastroduodenal trichobezoar: Rapunzel syndrome. *Official journal of the American College of Gastroenterology| ACG*, *104*(11), 2864-2865.
11. Frey, A. S., McKee, M., King, R. A., & Martin, A. (2005). Cheveux apparents: syndrome de Raiponce. *Am J Psychiatrie*, 162, 242-248.
12. Gonuguntla, V., & Joshi, D. D. (2009). Syndrome de Raiponce: un examen complet d'un cas inhabituel de trichobézoard. *Clin Med Res*, 7, 99-102.
13. Mehta, P., & Bhutiani, R. (2009). Le syndrome de Raiponce: est-ce un problème asiatique ? (rapport de cas et revue de la littérature), *Eur J Gastroenterol Hepatol*, 21, 937-940.
14. Middleton, E., Macksey, L. F., & Phillips, J. D. (2012). Syndrome de Raiponce chez un patient pédiatrique: un rapport de cas. *AANA J*, 80, 115-119.
15. Nour, I., Abd Alatef, M., Megahed, A., Yahia, S., Wahba, Y., & Shabaan, A. E. (2019). Syndrome de Raiponce (trichobézoar gastrique), une présentation rare avec œdème généralisé: rapport de cas et revue de la littérature. *Paediatr Int Santé de l'enfant*, 39, 76-78.
16. Ersoy, Y. E. A. F., Ayan, F., & Ersan, Y. (2009). Bézoards gastro-intestinaux: trente-cinq ans d'expérience. *Acta Chir Belg*, *109*(2), 198-203.
17. Qureshi, N. H., Morris, K., & McDevitt, B. (1992). Trichobezoar–a condition to think of in case of mobile abdominal mass. *Ir Med J*, *85*(2), 74.
18. Hafsa, C., Golli, M., Mekki, M., Kriaa, S., Belguith, M., Nouri, A., & Gannouni, A. (2005). Trichobézoard géant chez l'enfant. Place de l'échographie et du transit œsogastroduodénal. *Journal de pédiatrie et de puériculture*, *18*(1), 28-32.
19. Newman, B., & Girdany, B. R. (1990). Gastric trichobezoars-sonographic and computed tomographic appearance. *Pediatric Radiology*, *20*, 526-527.
20. Palanivelu, C., Rangarajan, M., Senthilkumar, R., & Madankumar, M. V. (2007). Trichobézoards dans l'estomac et l'iléon et leur élimination assistée par laparoscopie: un cas bizarre. *Singapour Med J*, 48, e37-e39.
21. Tayyem, R., Ilyas, I., Smith, I., & Pickford, I. (2010). Syndrome de Raiponce et perforation gastrique. *Ann R Coll Surg Engl*, 92, W27-W28.
22. Western, C., Bokhari, S., & Gould, S. (2008). Rapunzel syndrome: a case report and review. *Journal of Gastrointestinal Surgery*, *12*, 1612-1614.
23. Ben Cheikh, A., Gorincour, G., Dugougeat-Pilleul, F., Dupuis, S., Basset, T., & Pracros, J. P. (2004). Trichobézoar gastrique chez un adolescent présentant une anémie: résultats de l'échographie et de l'IRM, *J Radiol*, 85, 411-413.