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CASE REPORT

The Youngest Case of Rapunzel Syndrome: a variation in presentation

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Abstract:
Rapunzel syndrome is a rare cause of intestinal obstruction caused by a trichobezoar. There have been fewer than 40 cases reported and only one case was reported to occur in a male to date. We present the youngest patient in the literature and only the second male case.

Keywords: Rapunzel syndrome, trichobezoar
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Introduction

Rapunzel syndrome is a rare cause of intestinal obstruction caused by a trichobezoar. There have been fewer than 40 cases reported and only one case was reported to occur in a male to date. We present the youngest patient in the literature and only the second male case.

Case Report

A 2 year-old African American male presented with abdominal pain for one day and bilious vomiting for 2 days. The child had not passed stool for the previous 5 days. His mother reported a history of trichotillomania including ingesting hair from a hair brush since nine months of age. The patient has no previous medical or surgical problems, and a normal developmental history. The physical examination revealed a slightly distended abdomen with hyperactive bowel sounds and a firm mass in the right lower quadrant. There was tenderness to palpation in right lower quadrant and suprapubic area.

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Initial labs revealed a normal complete blood count (CBC) and C-reactive protein (CRP). Electrolyte study was significant for bicarbonate of 17 mmol/l. Abdominal radiograph was consistent with a small bowel obstruction and an abdominal computerized tomography (CT) scan suggested a foreign body in the terminal ileum (Figure 1).

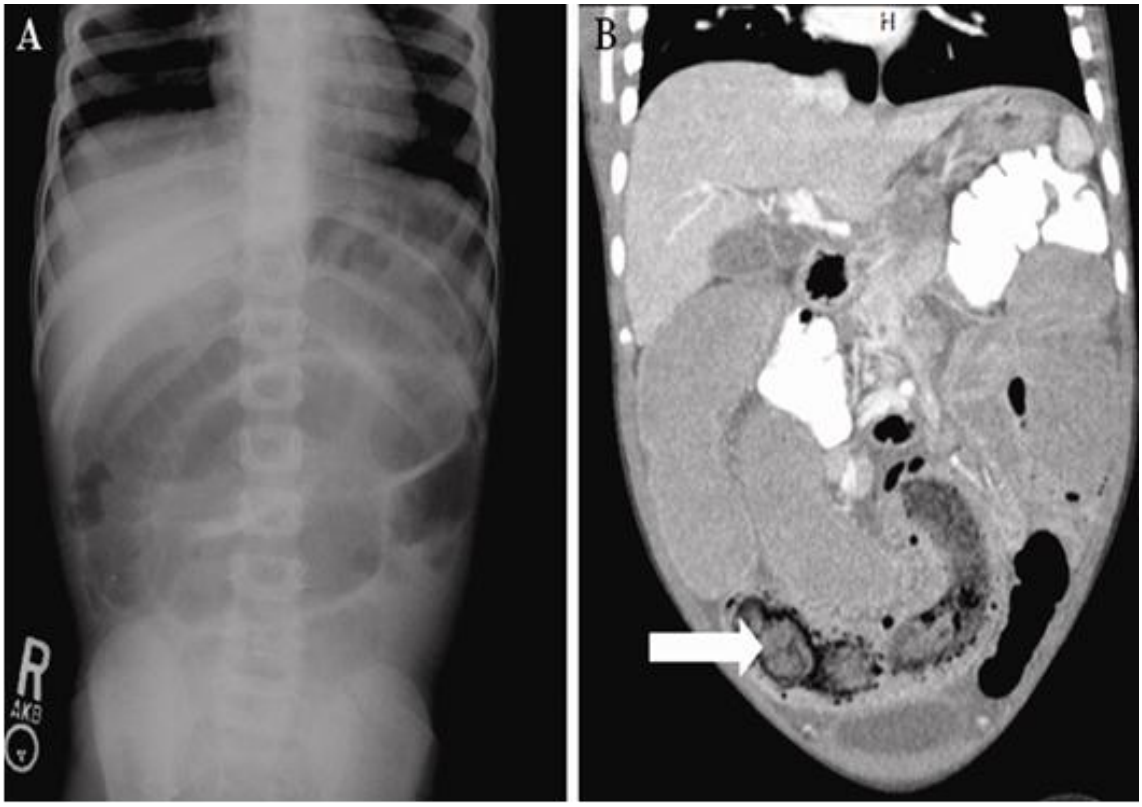

Figure I. Panel A) Abdominal X-ray showing small bowel dilatation suggestive of obstruction Panel B) Abdominal computer tomography showing foreign body in the terminal ileum (white arrow).

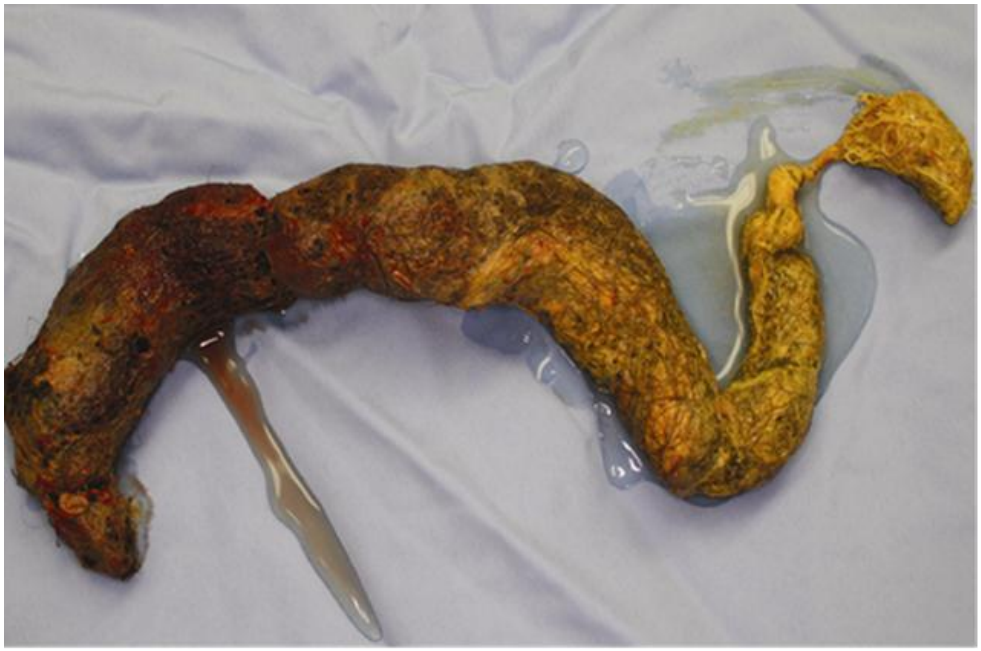

Figure II. showing the 30 cm trichobezoar with a tail.

Exploratory laprotomy demonstrated a firm mass in the terminal ileum. An enterotomy facilitated the removal of a 30 cm cylindrical trichobezoar (Figure 2). He had an uneventful post operative recovery.

Discussion

Trichobezoar is aggregation of swallowed hair within the gastrointestinal tract. It is associated with trichotillomania which is a irresistible impulse-control disorder with recurrent pulling out of their hair to relieve the tension [1]. Rapunzel syndrome is a rare variant of trichobezoar in which gastric trichobezoar extends into the small gut in the form of a long tail. It was first described by Vaughan and colleagues in 1968 [2]. Rapunzel name was derived from the long haired tower-bound character in the Grimm's Fairy Tales [3]. It is commonly presented among females. There are only fewer than 40 cases reported in the literature and this case is the youngest case and only second male case with Rapunzel syndrome [4]. Most of the cases have psychiatric issues but it can also occur among healthy women [5].

This syndrome can present with abdominal pain, nausea, vomiting, early satiety, weight loss, failure to thrive, intussusception, acute bowel obstruction and perforation. On examination they may have alopecia along with or without loss of eye lashes and a palpable mass without any indentation (Lamerton's sign) [6]. Radiographs with barium meal and ultrasound of the abdomen can aid in the diagnosis, but computer tomography (CT) of the abdomen is the modality of choice for the diagnosis. MRI also can be helpful but rarely utilized due to the high cost and time consumption. In the CT the mass will appear as a well-circumscribed lesion, composed of concentric whorls of different densities with pockets of air enmeshed. The smooth surface of hair does not allow propagation of the mass through peristalsis; instead, the hair becomes matted together in a ball. Different treatment options are available to remove the bezoar. But to date laparoscopy or surgical management has been the successful way of removing the trichobezoar, especially in Rapunzel syndrome due to large size and extension into small intestine. Endoscopic removal can be tried in small gastric trichobezoars. Lithotripsy, laser therapy, enzymatic digestion, lavage, prokinetic medications have been utilized with limited effectiveness depending on the size of the mass. In this case we proceeded with surgical approach due to acuity of the condition, size of the mass (30 cm) and the patient's younger age. Prompt removal of the trichobezoar is critical. Most common complication is intestinal obstruction and less frequently perforation leading to peritonitis. Failure to thrive, anemia, hypoalbuminemia, biliary obstruction, pancreatitis, intussusceptions, gastric ulceration have been reported in the literature [7,8].

Conclusion

This patient had a rare cause of intestinal obstruction by a trichobezoar. Thus, this case highlights the need for a high degree of suspicion for intestinal obstruction in pediatric patients with a history of tichophagia.

REFERENCES

1. Duke DC, Keeley ML, Geffken GR, Storch EA. Trichotillomania: A current review.Clin Psychol Rev. 2010;30:181-193.
2. Vaughan ED Jr, Sawyers JL, Scott HW Jr. The Rapunzel syndrome. An unusual complication of intestinal bezoar. Surgery 1968; 63: 339-343
3. Grimm J, Grimm W. Rapunzel. In Household Tales, vol 1. Trans Hunt M. London: George Bell, 1882, pp 50-54.
4. Naik S, Gupta V, Naik S, Rangole A, Chaudhary AK, Jain P, Sharma AK. Rapunzel syndrome reviewed and redefined. Dig Surg. 2007;24:157-161.
5. Coulter R, Antony MT, Bhuta P, Memon MA. Large gastric trichobezoar in a normal healthy woman: case report and review of pertinent literature. South Med J 2005; 98: 1042-1044.
6. Mathai J, Chacko J, Kumar TS, Scott JX, Agarwal I, Varkki S. Rapunzel syndrome: a diagnosis overlooked. Acta Paediatr. 2007;96:135-137.
7. Chogle A, Bonilla S, Browne M, Madonna MB, Parsons W, Donaldson J, Alonso E. Rapunzel syndrome: a rare cause of biliary obstruction. J Pediatr Gastroenterol Nutr. 2010;51:522-3.
8. Western C, Bokhari S, Gould S. Rapunzel syndrome: a case report and review. J Gastrointest Surg. 2008;12:1612-4