CASE REPORT

Rapunzel Syndrome Presenting with Ileo-Ileal Intussusception in A Boy

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ABSTRACT

Trichobezoars are usually reported in young girls with some psychiatric illness and rarely reported in boys. In this manuscript a case of 5 years old boy is reported who presented with signs of peritonitis. Ultrasound abdomen gave a positive donut sign. On exploration, it was revealed to be a case of Rapunzel syndrome with ileo-ileal intussusception.

Key words *Trichobezoar, Rapunzel syndrome, Intussusception, Boy.*

INTRODUCTION:

Trichobezoars are not uncommonly reported in pediatric age group. The term Rapunzel syndrome is used when this bezoar is associated with a long tail. Weight loss, anorexia, failure to thrive, and vomiting are common clinical features in a case of gastric trichobezoar. In some cases they are diagnosed only when complications arise. It is rarely reported in boys.[1-3] Herein a case of Rapunzel syndrome is reported that presented with ileo-ileal intussusception.

CASE REPORT:

A 5-year-old boy was referred with the complaints of abdominal distension, pain, vomiting, and constipation for a week. Past medical history revealed that he had been admitted in a local hospital for prolonged history of low grade fever, weight loss, anorexia, failure to thrive, and occasional emesis and diarrhea. He was put on anti-tuberculous therapy upon the suspicion of abdominal tuberculosis by the treating physician.

On examination the boy was lean with a weight of 10 kg. Abdomen was distended with mild to moderate tenderness all over. On deep palpation a mass was palpable with vague margins in the mid abdomen. Abdominal radiograph revealed two air fluid levels with paucity of gases. Ultrasound of the abdomen delineated donut sign with free fluid in the pelvis.

Patient was optimized and exploratory laparotomy performed. At operation an ileo-ileal intussusception was found. It was reduced manually however the involved small gut was non-viable. There were multiple globular masses within the lumen of small bowel that simulated polyps. Enterotomy was thus performed. The globular masses were part of a trichobezoar tail. The stomach was then palpated for the main bulk of trichobezoar and a gastrotomy was performed followed by retrieval of trichobezoar from stomach and small bowel (Fig I & II). Moreover, the proximal 20cm of jejunum was in u shaped configuration (Fig III). Initially it was tried to mobilize it but there was a great danger of iatrogenic jejunal perforation and since there was no obstruction at that level, therefore, left as such. After resection of gangrenous intestine at the level of mid ileum an ileostomy was fashioned as anastomosis was feared with dehiscence. The parents did admit when enquired after surgery, that their son had a habit of trichophagia at the age of two years but stopped this for the last two years.

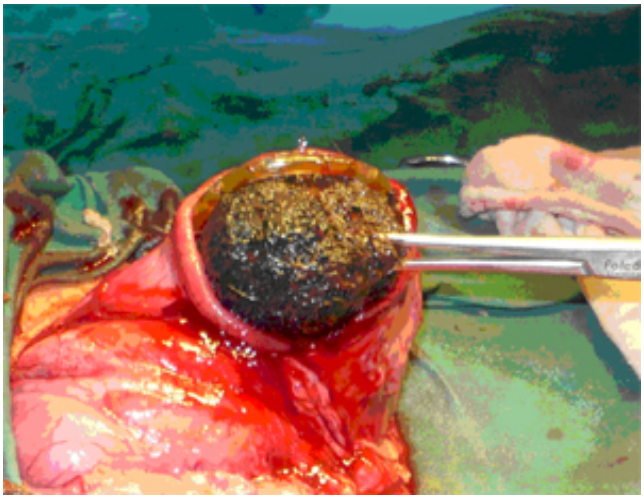

Figure I: Trichobezoar being retrieved from stomach.

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The postoperative recovery remained uneventful. Patient was discharged on 10th postoperative day. The psychiatric evaluation of the patient was normal. The child gained appetite as well as weight in the postoperative period. Ileostomy was then reversed after 6 months of the primary operation.

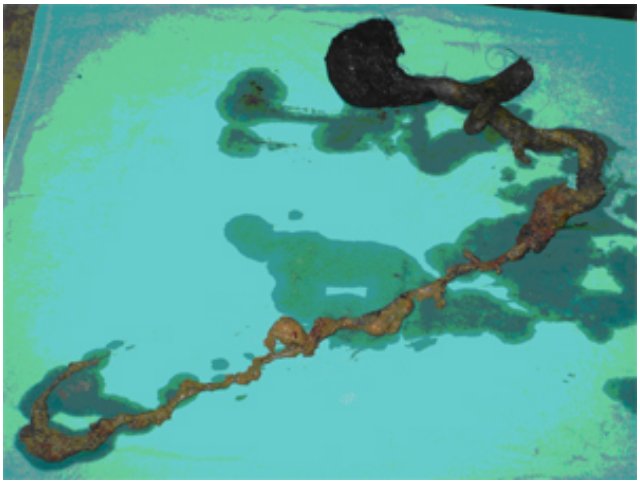

Figure II: About 5 feet long trichobezoar after retrieval from stomach and small bowel.

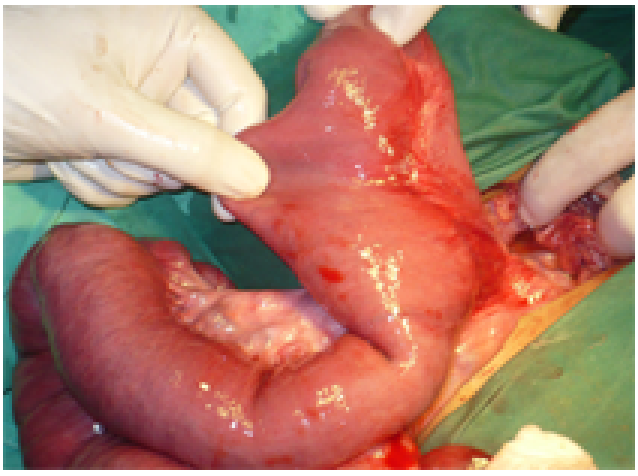

Figure III: Initial 20cm of jejunum was in u-shaped configuration

DISCUSSION:

Any trichobezoar with a long tail as in index case is termed as Rapunzel syndrome. The first description of Rapunzel syndrome came in 1968 by Vaughan et al.[1-3] Rapunzel syndrome is reported mostly in young female patients with some psychiatric problem. It is rarely reported in boys. In a literature review published in 2008 only 28 cases of Rapunzel syndrome were found. Of these 27 were female patients and one boy.[1]

Trichobezoars may remain undiagnosed for years and sometimes present with complications after many years of stopping the habit of trichophagia. Intussusceptions have been observed in 7% of children with trichobezoars.[4-7] In present case the trichobezoar remained undiagnosed rather misdiagnosed for two years and treated with antituberculous drugs without documentation.

In our case the complications of trichobezoars were intestinal gangrene due to ileo-ileal intussusception and pressure necrosis of the mucosa on the mesenteric aspects of the lumen of proximal small bowel that precluded primary anastomosis. Very small bezoars can be retrieved endoscopically; however, surgery remained the mainstay of treatment in cases of complications and large trichobezoars. Postoperative psychiatric evaluation is an essential part of the treatment regimen. In case of failed psychotherapy the recurrence of Rapunzel syndrome may occur though rarely reported.[8,9] In present case the child had no psychiatric illness on evaluation.

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