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Case Report

Unusual trichobezor of stomach and intestines as a manifestation of Rapunzel syndrome: a rare case report

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ABSTRACT

Rapunzel syndrome is a rare form of trichobezoar with accumulation of large amounts of hair extending from stomach to variable portion of small intestine. An 18 year old girl was brought to surgery opd with complaints of vomiting on and off since 3 months. Contrast enhanced computed tomography abdomen showed a bezoar extending from stomach into the duodenum and proximal jejunum suggestive of Rapunzel syndrome with 3rd and 4th part of duodenum along with duodenojejunal junction. On laparotomy, a trichobezoar occupying the entire stomach measured 100 cm in length and 700 grams in weight. Since the mass formed in Rapunzel syndrome is generally too large to be removed endoscopically or laparoscopically, it requires removal by open gastrostomy.

Keywords: Rapunzel syndrome, Trichobezoar, Duodenojejunal junction

INTRODUCTION

Rapunzel is a fictional character with long hair from a German fairy tale penned by the Grimm Brothers first published in 1812.[1] Rapunzel syndrome is a rare form of trichobezoar with accumulation of large amounts of hair extending from stomach to variable portion of small intestine.[2] First case of Rapunzel syndrome was reported in 1968. Herein we report one such case of 18 year old girl with Rapunzel syndrome.

CASE REPORT

An 18 year old girl was brought to surgery outpatient department with complaints of vomiting on and off since 3 months. Vomiting episodes occurred within minutes of taking food but there was some tolerance to liquid diet. There was history of loss of appetite and loss of approximately 10 kg weight in last 3 months. There were no associated complaints of pain abdomen, fever, diarrhea or constipation. On examination, patient appeared malnourished with facial puffiness and bilateral pedal edema. On per abdomen examination; a well-defined, firm, non-tender lump was palpable in the left hypochondrium. Ultrasonography abdomen was suggestive of multifocal jejuno-jejunal and ileo-ileal intussusceptions with omental thickening with mild mesenteric lymphadenopathy with moderate ascites and diffuse stomach wall thickening abdominal Koch's. She had Hb-11.5 gm/dl, total leucocyte count 3200 per cumm, total protein 3.7 gm/dl and albumin 1.8 gm/dl. Contrast-enhanced computed tomography (CECT abdomen showed a bezoar extending from stomach into the duodenum and proximal jejunum suggestive of Rapunzel syndrome with 3rd and 4th part of duodenum along with duodenojejunal junction lying to the right of spine suggestive of bowel malrotation. As such a huge bezoar was not possible to remove endoscopically, patient was taken up for surgery. On laparotomy, a trichobezoar occupying the entire stomach and extending well into the

jejunum was removed by anterior gastrostomy. The trichobezoar measured 100 cm in length and 700 grams in weight. Postoperatively patient recovered well. Psychiatry evaluation was done for trichophagia and patient discharged in a stable condition.

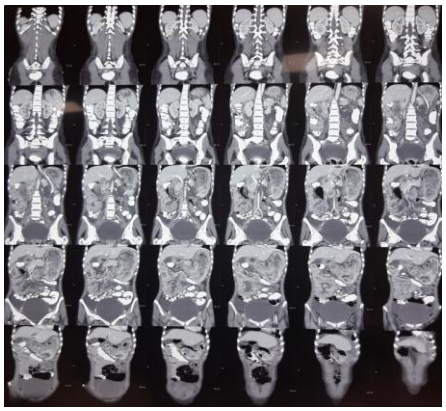

Figure 1: CECT abdomen (coronal sections).

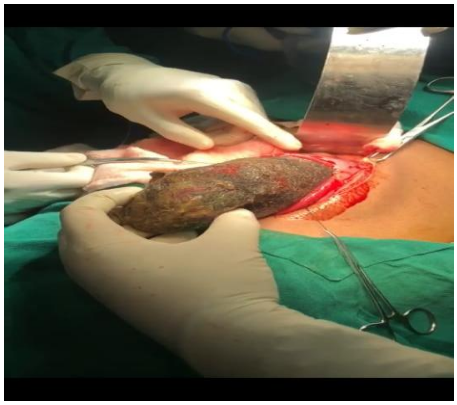

Figure 2: Intraop picture following gastrostomy.

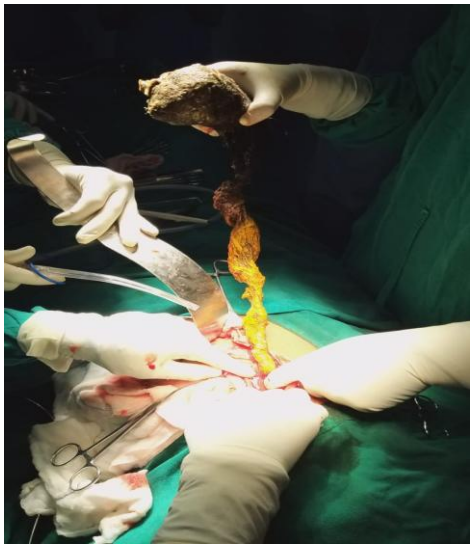

Figure 3: Intraop picture showing a trichobezoar with shape of stomach extending into the duodenum, jejunum and proximal ileum.

Figure 4: Picture of removed trichobezoar.

DISCUSSION

Bezoars are masses of ingested foreign material in the gastrointestinal tract. Trichobezoar are formed when these foreign material are ingested hair. These hair are immune to digestion and because of their smooth slippery surface are not easily propelled by peristaltic movements. As a result, more hair add over time to form a mass occupying most of the stomach. Rapunzel syndrome is a form of trichobezoar with a gastric hair ball and its tail extending into the pylorus and small bowel.[2]

Rapunzel syndrome is mainly seen in females which can be attributed to traditional long hair in females.[3] The presentation varies from mild abdominal pain to signs of perforation and sepsis.[4] On further enquiry, history suggestive of trichotillomania and trichophagia maybe elicited.[5] It is far more common in girls and generally accompanied with psychiatric disorders such as depression, anxiety, body dysmorphic disorders and alcohol and substance abuse.[6]

Since the mass formed in rapunzel syndrome is generally too large to be removed endoscopically or laparoscopically, it requires removal by open gastrostomy.[7] In the postoperative period, patient requires psychiatric evaluation as without it there are high chances of recurrence.

CONCLUSION

Rapunzel syndrome is rare extreme form of trichophagia with more incidences in females of adolescent period. The patient may present with the mild complaints of vomiting only. It can be confirmed with CECT abdomen and requires open gastrostomy for its complete removal. Treatment concludes with psychiatric evaluation and counseling to prevent recurrence.

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