



Case Report

Rapunzel Syndrome: a rare cause of acute small bowel obstruction

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ABSTRACT: The Rapunzel syndrome is a very rare condition where trichobezoar has extended up to the small bowel. Here we are reporting a rare case of Rapunzel syndrome in an adolescent girl with history of trichophagia who presented with small bowel obstruction. Patient underwent exploratory laparotomy and bezoar was removed through gastrotomy. Post-operative course was unremarkable.

KEY WORDS: Rapunzel syndrome; Trichobezoars; Psychotherapy; Gastrotomy

INTRODUCTION

The Rapunzel syndrome is a very rare condition where trichobezoar has extended up to the small bowel. Most common presentation is abdominal lump followed by pain, nausea, and vomiting and weight loss. The laboratory investigations have shown low hemoglobin in approximately 62% cases. Trichobezoars always require surgical removal through gastrotomy as conservative measures are not of much use. Psychotherapy and regular follow up are necessary.

CASE DETAILS

A 14 years old girl was admitted to the hospital with chief complaint of pain in epigastric region since last 1 year and the pain having increased in severity since previous 2 days. The pain was associated with on and off vomiting since previous 2 months; vomiting was bilious since last two days. History of weight loss was present. History of trichophagia was present. On physical examination, she was found to be pale with mild dehydration and had a mild degree of alopecia. On palpation, a lump was palpable in the epigastric region. X- Ray abdomen erect showed sign of small bowel

obstruction (**Figure 1**). USG abdomen showed heterogeneous mass in the epigastrium. All blood investigations were normal except mild anemia. Exploratory laparotomy was planned; on opening the abdomen, mass was found to be present in the stomach, which extended below till the ileum. Gastrotomy was done and mass was delivered through gastrotomy without breaking the extension which was up to the terminal ileum (**Figure 2**). Primary closure of the gastrotomy was done. Postoperative course was unremarkable. Patient was referred to psychiatrist for psychotherapy.



Figure 1: Radiograph showing small bowel obstruction

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Figure 2: Trichobezoar extended up to terminal ileum

DISCUSSION

The word bezoar comes from Arabic badzehr or the Turkish padzehr, meaning antidote. Bezoars are concretion of ingested materials that are unable to exit through pylorus. The classification of bezoars into four varieties, namely, *trichobezoars*, *phytobezoars*, *medicobezoar* and *lactobezoars* is arbitrary and is based on their composition.

The Rapunzel syndrome is very rare condition where trichobezoar has extended up to the small bowel. Till date hardly 30 cases has been reported. The syndrome is named after long haired girl Rapunzel. This condition was originally described by Vaughan et al^{1,2}. Rapunzel was a long haired girl in Grimm's fairy tales who was imprisoned in a tall castle and lowered her hair from a high window to allow her prince to climb up and rescue her. Most of the cases present late. Most common presentation is abdominal lump followed by pain, nausea, and vomiting and weight loss. General examination may reveal alopecia and halitosis. Predisposing conditions for bezoars are either eating habit or pre-existing gastroduodenal pathology like previous surgery or neuropathy. History of trichophagia may be present. Trichophagia is sometimes associated with the

psychological disorder known as trichotillomania (strong desire to pull hair). The laboratory investigations have shown low hemoglobin in approximately 62% cases³. Complications of trichobezoars include obstruction, ulceration, GI bleeding and perforation⁴. Diagnosis of trichobezoars can be made easily on endoscopy or sometimes on plain radiograph. Trichobezoars always require surgical removal through gastrotomy as conservative measures are not of much use. Prophylactic antibiotics should be given as trichobezoars are most of the time putrefied. After removal, attention must be paid towards prevention of recurrence. Prophylactic oral enzyme and prokinetic drugs may help in prevention of recurrence. Psychotherapy and regular follow up are necessary.

In conclusion, high index of suspicion is required for pre-operative diagnosis of Rapunzel syndrome. Complications always required surgical removal. Psychotherapy is an essential part of management of Rapunzel syndrome

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