

Multiple Small Bowel Perforations in a Patient with Rapunzel Syndrome

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ABSTRACT

A twelve year old, mentally challenged female, weighing 17 kilogram with the habit of trichophagia and trichotillomania presented with colicky abdominal pain for the last fifteen days. On examination of abdomen generalized tenderness was present. Ultrasound was suggestive of intussusception. At laparotomy trichobezoar mixed with phytobezoar found in stomach with tail up to proximal ileum. There were multiple perforations found in the jejunum and proximal ileum along their length in linear fashion. All perforations were repaired. Postoperative course was uneventful.

Key words Trichophagia, Trichotillomania, Phytobezoar, Rapunzel syndrome.

INTRODUCTION:

Trichophagia (eating of hair) and trichotillomania (pulling of hairs) are commonly reported in mentally challenged patients.¹ Trichobezoar are not uncommon in such patients.² Its frequency is reported as 1% in all cases of trichophagia.³ Rapunzel syndrome, in which tail of the bezoar is found in a variable length in small bowel, is an uncommon psychological disorder in association with trichobezoar.⁴ These patients are mostly females usually presenting in adolescence. Intestinal perforation in these patients is rarely reported. This case report describes management of one such patient.

CASE REPORT:

A 12-year old female patient weighing 17 kg, who was mentally challenged, presented to ER with the complaints of severe abdominal pain for the last 15 days. There was a history of trichophagia and trichotillomania. She was a neglected child. Her parents were not alive. She had fifteen siblings of whom ten died during the first month after delivery. She was living with her grandmother with other three sisters and one brother.

On clinical examination patient looked unwell though hemodynamically stable. Abdomen was mildly tender on palpation. Investigations showed leukocytosis of

13,500 mm³ and hemoglobin of 10.5 gm/dl. Serum electrolytes, urea and creatinine were in normal range. X-ray abdomen in erect posture showed few air-fluid levels with no free air. Ultrasound abdomen suggested a mass lesion measuring 2.5cm x3cm in right iliac region favoring intussusception.

The patient was diagnosed as having sub acute intestinal obstruction and kept under observation but she did not improve. During her stay in the ward abdominal pain worsened with generalized tenderness. Laparotomy was then performed. At exploration a large mass was found in stomach and a smaller one in proximal ileum (Fig. I). Both were connected to each other with a rope like structure. Multiple perforations and devitalized areas of gut wall were found in jejunum and ileum in linear fashion near mesenteric border, starting two centimeters distal to duodeno-jejunal junction up to proximal ileum (Fig. II). The mass on gastrotomy was found to be a trichobezoar mixed with phytobezoar with tail up to proximal ileum. As the whole length of jejunum and proximal ileum of about 110cm was involved, a plan to repair all perforations was made. Margins of the perforations were freshened and closed with polyglycolic sutures in extra mucosal fashion. A total of 22 perforations were repaired. Postoperatively patient was monitored in Surgical ICU and given parenteral nutrition from peripheral route. Except for superficial wound infection, postoperative course remained uneventful. At discharge counseling was done with the family and patient was referred for psychiatric treatment and follow up.

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Fig I: Trichobezoar tail removed from the small bowel.



Fig II: Linear perforations in the jejunum and ileum.

DISCUSSION:

The final diagnosis in the case managed was Rapunzel syndrome due to a mixed bezoar composed of hairs and vegetable residue, with multiple small bowel perforations in a mentally challenged female. It is a psychiatric disorder, found more commonly in females. Bezoars are concretions of human hairs or vegetable fibers that accumulate in the gastrointestinal tract. The most common type of bezoar is the trichobezoar. They are also found in people without mental disorders.

Clinical presentation in such cases may be vague. In those with psychiatric disorders it may not be noticed even when disease is advanced as noted in present cases. Trichophagia though often noticed by care providers but often its gravity is not appreciated. Pulling of one's own hair cannot go unnoticed. Such symptoms must raise suspicion of trichobezoar.

Intestinal obstruction and perforations in patients with Rapunzel syndrome are reported though

infrequently.^{5,6,7} In the index case the perforations were different from what is reported in literature. These were multiple over considerable length of the intestine. As there was chance of small gut syndrome due to removal of almost whole of the jejunum, decision to repair them primarily was made and it was found correct. The postoperative outcome was uneventful. These patients also need psychological and social support. This must be ensured as chances of recurrent trichobezoar and Rapunzel syndrome are always there.⁸

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