

Rapunzel Syndrome as a Rare Presentation of Trichobezoar

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Citation

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Abstract

Rapunzel syndrome is a type of presentation of trichobezoar, an extension of hair fibers into the small bowel. Its clinical presentation is deceptive ranging from abdominal mass to symptoms of obstruction. We report a rare presentation of Rapunzel syndrome in a 16-year-old girl, where the trichobezoar was found as a lump in the abdomen with a history of a psychiatric disorder, trichotillomania, with patches of alopecia. Treatment was by laparotomy with removal of the trichobezoar. In many cases, the diagnosis was made very late in the history of the disease, at a stage where surgery remains the only cure. A multidisciplinary approach including psychiatric evaluation and proper management after surgery should be adopted to prevent recurrence of the disease.

INTRODUCTION

An agglomeration of indigestible organic or inorganic foreign material in the gastrointestinal tract, usually accumulated in the stomach, is called bezoar. Types depending on their composition are: phytobezoars (vegetable matter), pharmacobezoars (medication/drugs), trichobezoars (hair or hair-like fibers) and lactobezoars (milk curd). Trichotillomania (strong urges to pull own hair) and trichophagia are closely related and can lead to formation of a trichobezoar. They predominantly affect females in early childhood or adolescence.^{1,23} Very rarely in cases of the Rapunzel Syndrome, hair extends through the pylorus into the small intestine causing symptoms and signs of partial or complete gastric outlet obstruction.⁴

CASE

A 16-year-old adolescent girl presented with vague abdominal pain of a week's duration. The abdominal pain was generalised and colicky in nature and there was increased tenderness in the peri-umbilical region. The pain was associated with nausea, vomiting and with weight loss. The patient was thin-built and had mild pallor. She had a recent psychiatric history and patchy alopecia as well as a family history of schizophrenia. Examination of the abdomen revealed a large curvilinear hard intra-abdominal mass, occupying the epigastric and left hypochondriac region. Abdominal skiagram and ultrasonography were suggestive of a gastric outlet obstruction associated with an

unusual air shadow in the left hypochondrium in the region of the stomach and duodenojejunal flexure. At laparotomy, the distal part of the stomach, the pylorus, and the duodenum were distended and contained a large mass of dark, foul-smelling hair that had crossed the pylorus and extended into the small bowel up to proximal jejunum. There was no perforation or free fluid noted in the peritoneal cavity. The trichobezoar (Figure 1) was removed via a gastro-enterotomy. Recovery was slow but complete and the patient was eventually discharged after psychiatric evaluation and management with tranquilizers.

Figure 1

Figure 1: The whole mass of hair extracted with its tail extending from pylorus to proximal jejunum.



DISCUSSION

Rapunzel Syndrome is a rare presentation of trichobezoar, involving strands of swallowed hair extending as a tail through the duodenum, beyond the stomach (after 'Rapunzel' – the maiden in the Grimm brothers' fairy tale of 1812 whose long hair flowed out of her prison tower allowing her prince to rescue her).⁴⁵ It was first described by Vaughan in 1968.⁶ It is common in young females usually with mental retardation and an underlying psychiatric disorder; almost half of the patients present with trichophagia.⁷ Its formation begins with small pieces/fibers of hair, which, ingested, gather in the stomach but do not progress any further, and a mass builds up in the stomach which is unable to dislodge it. Sometimes, the aggregate of hair fragments and small pieces that result do pass through into the intestine, thus leading to sequelae such as ulceration, partial or total obstruction, intestinal perforation and peritonitis. Patients are often asymptomatic, but may present with nausea, vomiting, anorexia, weight loss, vague abdominal pain or constipation.⁸⁹ The diagnosis is based on a combination of good history taking, family history of psychiatric disorders, previous bezoars as well as physical findings to look for, a palpable mass, patchy hair loss and halitosis. An abdominal radiograph may show a prominent gastric outline with an intragastric mass, outlined by gas in the distended stomach. A mobile filling defect is seen with barium, and it may show extension into the duodenum. Abdominal ultrasound shows a dense, echogenic rim with sharp, clear posterior shadowing in the epigastrium. Abdominal CT shows a mobile intragastric mass with a mixed density pattern due to the presence of entrapped air and food debris, and it will also delineate the extension of the trichobezoar. In endoscopy, trichobezoars are often found to have a hard, concrete-like appearance and enzymatic oxidation of hair gives them a darkened colour. Treatment may include endoscopic removal but with risk of bowel perforation, so this should be restricted to small trichobezoars only.¹⁰ Chemical dissolution, mechanical fragmentation and laser-ignited mini-explosive technique were used successfully for small bezoars.¹¹ Laparoscopic techniques are also becoming fashionable and large bezoars can be milked into the caecum before removal. However, open surgery still plays a major role in large trichobezoar removal, especially if there is an extension into the bowel, which might be missed or cumbersome with other methods of treatment. Recurrence of Rapunzel Syndrome has been noted and it is seen to occur because the underlying emotional stress trigger was not corrected.¹²¹³ However, treatment and psychological support

of the mental as well as physical disorder is important for prevention of its recurrence.

CONCLUSION

Trichobezoar is an uncommon cause of obstruction to be thought of particularly in young females. It should particularly be borne in mind when the patient is young and female and especially if there is accompanying alopecia circumscripta and a mental abnormality associated with trichophagy. If there is a long history of a gastrointestinal problem in young age with history of trichophagia, early endoscopy is recommended. Radiological investigations may clarify the issue, and the bezoars will sometimes be retrievable by minimally invasive means. A multidisciplinary approach including psychiatric evaluation and proper management after surgery should be adopted to prevent recurrence of the disease.

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