Trichobezoar causing intussusception: Youngest case of Rapunzel syndrome in a boy in North America

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ABSTRACT

Trichobezoar is a mass of hair retained in the stomach, usually associated with trichotillomania. Large trichobezoars may extend into the small bowel and cause small bowel obstruction, a condition known as Rapunzel syndrome. Here we report, to our knowledge, the youngest North American case of Rapunzel syndrome causing intussusception in a male patient.

1. Case report

An otherwise healthy nine-year-old Native Canadian boy was brought to the emergency department in Northern Ontario with a four-day history of crampy abdominal pain, nausea, and vomiting. He had been having a three month history of chronic abdominal pain and weight loss, but this was never investigated. He did have reduced appetite, but denied any change in bowel movements. He was developmentally appropriate, and had no other medical or surgical history. Family history was unremarkable. On examination, the patient’s abdomen was soft and nontender with normal bowel sounds. Laboratory values did not show any abnormalities. Abdominal radiographs revealed a large mass within the stomach, and dilated loops of small bowel with air fluid levels. A computed tomographic (CT) scan of the abdomen and pelvis was obtained, revealing a large gastric bezoar extending into the duodenum (Fig. 1), and several points of enteroenteric intussusception. The patient was transferred to the London Health Sciences Centre for operative management. In the operating room, after the patient was intubated, esophagogastroduodenoscopy showed a large, hardened mass in the stomach composed of hair, extending beyond the pylorus (Fig. 2). A laparotomy was performed, and the grossly dilated stomach was easily visualized. An 8-cm gastrostomy was performed and the trichobezoar was delivered from the stomach (Fig. 3). We could palpate thick strands of hair in the jejunum and ileum, and noted two perforations in the jejunum caused by the hair strands (Fig. 4), that had been sealed off by mesentery. We resected the perforated jejunum, delivered another trichobezoar through the enterotomy, and completed a primary side to side stapled anastomosis. We then made two more enterotomies in the ileum to deliver a long, thick trichobezoar strand that was causing a long intussusception of the terminal small bowel. The gastrotomy and enterotomies were closed in two layers, no drains were left, and the abdominal fascia and skin were closed. The patient was kept nil per os (npo) for seven days, receiving total parenteral nutrition in the interim, before starting oral feeds. A complete inpatient psychiatric assessment was also performed on him, and did not reveal any abnormalities. No other complications were encountered during the hospital stay, and the boy was discharged home after two-weeks.

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2. Discussion

Trichophagia leading to formation of trichobezoars is a rare condition: the latter occur when hair strands are retained in the folds of the gastric mucosa because their slippery surface prevents propulsion by peristalsis. As more hair is added, peristalsis causes it to be enmeshed until a mass forms and eventually assumes the shape of the stomach. Trichobezoars that extend through the pylorus into the small bowel and cause symptoms of Rapunzel syndrome are even more uncommon. Originally described in 1968 [1], Rapunzel syndrome includes a trichobezoar with a tail that extends at least to the jejunum and causes symptoms suggestive of obstruction. It is particularly rare among males, with only three cases reported among the approximately forty-one cases in the literature [2–4]. Two patients were developmentally delayed, and 1 was a developmentally normal teenager. We believe this case represents the youngest reported case of Rapunzel syndrome in a male patient in North America. On retrospective questioning the patient denied trichophagia but his mother remarked that she had often seen him putting hair-balls in his mouth. Even though the patient had been symptomatic for several months prior to his surgery, his family had not sought medical attention, possibly due to their distance from nearby medical centers in Northern Ontario.

Removal of trichobezoars is typically undertaken endoscopically when feasible, although this approach is rarely successful because of the density and cohesion of the trichobezoar. Laparoscopic approaches have also been undertaken successfully [3,5,6], and have been shown to have fewer complications and reduced length of hospitalization [7]. Because our patient presented with obstructive symptoms secondary to intussusception, a laparotomy had to be performed to examine the bowel, retrieve the distal tail, and resect a portion of jejunum that had been perforated by the hair.

3. Conclusion

We present, to the best of our knowledge, the youngest North American male patient to present with obstruction and...
intussusception due to Rapunzel syndrome. Although endoscopic and laparoscopic approaches have been described, this patient’s clinical presentation with obstruction necessitated a laparotomy.

Consent

Written informed consent was obtained from the patient’s mother for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Conflict of interest

The authors do not have any conflict of interests to declare.

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Appendix A. Supplementary data

Supplementary data related to this article can be found in the online version, at http://dx.doi.org/10.1016/j.epsc.2013.01.001.

References