|| ISSN(online): 2589-8698 || ISSN(print): 2589-868X ||
International Journal of Medical and Biomedical Studies
Available Online at www.ijmbs.info
PubMed (National Library of Medicine ID: 101738825)
Index Copernicus Value 2018: 75.71
Case Report Volume 4, Issue 1; January: 2020; Page No. 20-22

A RARE CASE OF RAPUNZEL SYNDROME - A SMALL BOWEL TRICHOBEZOAR

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Article Info: Received 16 December 2019; Accepted 05 January. 2020
DOI: https://doi.org/10.32553/ijmbs.v4i1.847
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Conflict of interest: No conflict of interest.

Abstract
Bezoars are concretions of foreign material in the gastrointestinal tract, mainly the stomach. In humans, the most common type of bezoar is the trichobezoar, is more often found in children and teenage girls. . The most frequent type of bezoar in adults is phytobezoar. A rare manifestation of trichobezoar is Rapunzel syndrome (RS) which occurs when the bezoar extends into the small intestine. An 11 years old girl from low socio economic social status presented to emergency department. . She had a complain of abdominal pain and vomiting before 3 month which was relieved itself after vomiting which contains hair, threads and cloths. Abdominal examination revealed a generalised abdominal distension. Ultrasonography (USG) of abdomen was done. . Trichobezoar was removed by open surgery with longitudinal enterotomy. The mass was occupying 30cm part of terminal ileum up to iliocecal junction with a putrid smell which was removed from enterotomy. Trichobezoars are a bizarre medical problem, and Rapunzel syndrome is an extremely uncommon variety of trichobezoar which occurs when the bezoar extends into the small intestine. The diagnosis of trichobezoar is possible on imaging in a proper clinical setting.
Keywords: Trichobezoar, Rapunzel Syndrome, Enterotomy.

Introduction

Bezoars are concretions of foreign material in the gastrointestinal tract, mainly the stomach. The word “bezoar” comes from the Arabic word “bedzehr” or the Persian word “padzhar,” meaning “protecting against a poison.” The first reference to a bezoar in a human was in 1779 during an autopsy of a patient who died from gastric perforation and peritonitis. Bezoars can be classified in six types: phytobezoars (composed of indigestible plant material), trichobezoars (hairball or hair-like fibres), lithobezoars (fragments of small stones, or gravel stones), pharmacobezoar (tablets or semiliquid masses of drugs), plasticobezoars (plastic), and lactobezoars (inspissated milk) .In humans, the most common type of bezoar is the trichobezoar, is more often found in children and teenage girls. The most frequent type of bezoar in adults is phytobezoar. In contrast to other types of bezoars, trichobezoars are usually observed in individuals linked to pica and other psychiatric conditions, such as emotional disturbance, learning disabilities, and history of neglect or mental retardation. These individuals pull out their own hair and swallow it, processes referred to as trichotillomania and trichophagia.A rare manifestation of trichobezoar is Rapunzel syndrome (RS) which occurs when the bezoar extends into the small intestine . The name “Rapunzel” syndrome comes from the Grimm Brothers’ fairy tale of a 12-year-old princess who was shut into a tower with neither stairs nor doors by an enchantress who climbed up the tower’s walls with the help of Rapunzel’s long tresses.

Case Report:

An 11 years old girl from low socio economic social status presented to emergency dept of the Sir T hospital, Bhavnagar with chief complain of Abdominal distension , bilious vomiting , not passed stool since 3 days. According to her parents, the girl pulled out her own hair and swallowed it, and also swallowed thread and clothes since the age of 5 years. She had a complain of abdominal pain and vomiting before 3 month which was relieved itself after vomiting which contains hair, threads and cloths. Abdominal examination revealed a generalised abdominal distension with generalised tenderness. Guarding and rigidity was also present with hyperactive bowel sound. Her routine blood investigations were with in normal limit. Plain X- ray of abdomen in standing position shows multiple air fluid level with step ladder pattern. Ultrasonography (USG) of abdomen shows moderate free fluid noted in peritoneal cavity with multiple segments of faeces and fluid filled dilated bowel loops in entire abdomen average diameter of 3cm with to and fro peristalsis. Trichobezoar was removed by open surgery with longitudinal enterotomy. The mass was occupying 30cm part of terminal ileum up to iliocecal junction with a putrid smell which was removed from enterotomy. Enterotomy was closed horizontally in double layer using silk 2-0. The child recovered well postoperatively and was

discharged after 10 days of surgery. She was advised to follow up in surgical as well as psychiatric OPD.

Figure 1: Small Bowel Trichobezoare (Found at 30cm segment of terminal ileum up to ileocecal junction)

Figure 2: Removed trichoezoare by enterotomy.

Discussion:

Trichobezoars are most commonly seen in females (approximately 90%) aged between 10 and 19 years. Only in half of these patients, a history of trichophagia is found. Around 30% of the patients with trichotillomania, a psychological condition that involves strong urges to pull hair, will engage in trichophagia. Trichobezoars are formed because of hair being slippery and get retained in gastric folds escaping peristaltic propulsion. More and more hairs accumulate and get enmeshed into a ball and assume the shape of the stomach. Decomposition and fermentation of trapped food often leads to a putrid smell to bezoar and patients breathe. Rapunzel syndrome is a rare form of trichobezoar. Some define it as a gastric trichobezoar with a tail extending up to the jejunum or beyond; and some still define it as a bezoar of any size which can cause intestinal obstruction. Trichobezoars can present with anaemia, abdominal pain, haematemesis, nausea and/or vomiting, bowel obstruction, gastric ulcers, perforation, gastrointestinal bleeding, acute pancreatitis, and obstructive jaundice. The complications of Rapunzel syndrome ranges from attacks of incomplete pyloric obstruction to complete obstruction of the bowel to perforation peritonitis and death. The gold standard for diagnosis of gastric trichobezoar is upper GI endoscopy however, it does not prove the existence of the RS. The abdominal CT scan is the most accurate imaging test concerning the presence of trichobezoars, since it demonstrates heterogeneous, mottled intraluminal mass with low attenuation and air trapping . Furthermore, it can track with great detail the extension of the trichobezoar's tail to the gastrointestinal tract. Abdominal ultrasound and barium meal (with the characteristic honeycomb appearance) can prove to be useful. Successful management and treatment of bezoars requires removal of the mass and prevention of recurrence. Method of removal depends on its consistency, size, and location. In the early stages endoscopic removal is possible, though however it should be reserved for small trichobezoar. Various other methods like extracorporeal shock wave lithotripsy (ESWL), intragastric administration of enzymes (pancreatic lipase, cellulase), and medications (metoclopramide, acetylcysteine) have been reported with varying results. Open surgery still remains the corner stone of large trichobezoar removal especially if it has an extension into the bowel, which might be missed with other methods of treatment. Laparoscopy has been also used with limited success. Recurrences have been reported in the literature after the initial removal of bezoars hence long term psychiatric follow-up is advised. Recurrence of Repunzal Syndrome is extremely rare, with the total number of recurrences reported in the literature, being 3. The most common characteristic of the referred recurrences was that these patients defaulted follow-up after a few months and consequently did not complete the psychiatric treatments. In order to decrease recurrence, patients should receive psychiatric/ psychological support after the surgical treatment. It should be kept in mind that the regular long-term psychiatric follow-up might prevent recurrences, although these are rare. In addition, despite the fact that studies of the pharmacotherapy of the trichotillomania remain inconsistent, recurrence seems to be avoided in some patients after pharmacotherapy.

Conclusion:

Trichobezoars are a bizarre medical problem, and Rapunzel syndrome is an extremely uncommon variety of trichobezoar. The diagnosis of trichobezoar is possible on imaging in a proper clinical setting. However, Rapunzel syndrome is most often an intra operative finding. Even today open surgical removal forms the mainstay of treatment. All patients with trichobezoar should be referred for psychiatric evaluation after surgery to avoid recurrence.

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