RAPUNZEL SYNDROME IN A 2 YEAR OLD FEMALE CHILD: A RARE PRESENTATION OF A RARE DISEASE.

Surgery

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ABSTRACT

Rapunzel syndrome is a very rare variant of trichobezoar in which the mass in stomach extends into small intestine and sometimes into the colon. Here we report an extremely rare case of a 55 centimeter long trichobezoar in a female child at an age of 2 years and 4 months. This is the youngest reported case of Rapunzel syndrome.

KEYWORDS

Rapunzel syndrome, bezoar, pediatric surgery, trichobezoar, early presentation.

INTRODUCTION

The name Rapunzel syndrome is derived from the well-known fairy tale by the Grimm brothers. Vaughan et al first reported Rapunzel syndrome and so named it because of the long hair that can extend into the small intestine.[1] Rapunzel syndrome is very uncommon with fewer than 50 reported cases. Rapunzel syndrome is a potential complication of a trichobezoar. It has been defined as a gastric bezoar with a "tail" that can extend through the pylorus all the way to the ileocecal junction.[2] This tail consists of the distal ends of long pieces or fragments of hair, some still attached to the growing mass in the stomach.

It is most commonly found in emotionally disturbed and/or mentally challenged adolescents, usually between the ages of 13 and 20 years. It reflects an underlying psychiatric disorder, not a problem with gastrointestinal motility.[2] Trichotillomania and trichophagia are associated with Rapunzel syndrome and are often indicative of psychosis.[2] Here we report an extremely rare case of a 55 centimeter long trichobezoar in a female child at an age of 2 years and 4 months. This is the youngest reported case of Rapunzel syndrome.

CASE REPORT

A female child of age 2 years and 4 months from a poor family presented with epigastric pain, post prandial emesis, anorexia, loss of weight and early satiety. History elicited from mother revealed that the child had trichotillomania and trichophagia. On general examination, the patient had pallor, generalised wasting and alopecia. Her vitals were normal. On abdominal examination, there was a non-tender, smooth and hard lump in epigastrium which was extending to right hypochondrium. Intussusception and an obstructing mass was found on contrast enhanced CT of abdomen. An open laparotomy was carried out using supraumbilical midline incision. An anterior gastrotomy incision was made and gastric portion of the mass was revealed. As the mass was extending into the small intestine, carefully planned and placed enterotomy incisions facilitated the easy removal without causing significant injury to the bowel. 55 centimeter long trichobezoar was removed (Figure 1 A and B).

Figure A: Ct Of Abdomen Showing Entrapped Gas Bubbles In The Stomach Mass. B: 55 Cm Long Trichobezoar After Its Surgical Removal.

The gastrotomy and enterotomy incisions were closed in 2 layers. The post-operative period was uneventful and the patient recovered well. She was discharged on 12th post-operative day. Referral for a psychiatric consultation was made. She was followed up for 8 months without evidence of recurrence.

DISCUSSION

Bezoar is a hard mass of collected fibres of plant or human origin. Trichobezoar is a condition in which a mass of entangled hair is formed commonly in the stomach. Due to slippery nature of hair, it escapes peristalsis of stomach. Subsequent ingestion of hair leads to formation of a tight and enlarging mass of hair that creates a large and hard obstruction. This mass traps thicker material like food particles that binds the hairs, making its removal difficult.[3] The churning action of the stomach during attempted digestion helps entangle new hair into an already formed mass. Mucus covers the gastric trichobezoar, giving it a shiny surface. The mass is usually black because of the acidic nature of the stomach that denatures the proteins. Patients often have a foul smelling breath odor because of the decomposition and fermentation of fats. Patients often remain asymptomatic for many years and remain so until the bezoar increases in size to the point of obstruction.[2] Personal history, psychiatric disorders in family, previous bezoars as well as physical examination of a palpable mass, halitosis, and patchy hair loss aids in diagnosis. Presentation of trichobezoar is usually late, due to the low index of suspicion by the clinician. Out of 131 cases of trichobezoar, a palpable abdominal mass is present in 87.7%, abdominal pain in 70.2%, nausea and vomiting in 64.9%, weakness and weight loss in 38.1%, constipation or diarrhea in 32%, and hematemesis in 6.1%.[4] Recently, literature indicate that trichobezoars could be the rare cause of jejunal intussusception, appendicitis, nephrotic syndrome, and biliary obstruction.[5]

Diagnosis can be confirmed by endoscopy or contrast radiography. Endoscopy can also be used to remove minor and more proximal trichobezoars. The computed tomography is the most useful diagnostic tool because it reveals the localization of the bowel obstruction. A mottled gas pattern in the mass is reported characterizing the bezoar, and it is supposed to be air bubbles retained within the bezoar.[6]

Despite availability of endoscopy and laparoscopic methods their application as treatment is limited to smaller trichobezoars. Open surgery remains gold standard for full blown Rapunzel syndrome. In a literature review, 100 cases were identified in which conventional laparotomy was successful in the removal of the trichobezoar(s). [7] In the 100 cases, 12% involved 1 or more complications, including perforation of the intestine during removal of the trichobezoar, minor wound infection, pneumonia, paralytic ileus, ileal trichobezoar, and fecal leakage through the lower part of the laparotomy wound.[7]

Psychiatric consultation remains crucial as the recurrence can occur if the behavioural changes are not brought in the patient.

CONCLUSION

A diagnosis of Rapunzel syndrome at early ages is extremely rare and

warrants a strong suspicion in patients presenting with vague symptoms and with a medical history of associated psychiatric conditions. Treating the psychiatric condition at an early stage prevents recurrence of trichobezoar.

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