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Case Report

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RAPUNZEL SYNDROME IN AN 8 YEARS OLD FEMALE CHILD

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ABSTRACT

Rapunzel syndrome is a large gastric trichobezoar with duodenal and jejunul extension, most commonly seen in teenage girls with some psychiatric illness. We are reporting a case of Rapunzel syndrome in an 8 yrs old female child presenting with epigastric mass and gastric outlet obstruction. Due to the rarity we are prompted to report this case.

KEYWORDS: Trichobezoar, Rapunzel, Epigastric Mass, Gastric Outlet Obstruction.

INTRODUCTION

The first reported trichobezoar was described in a 16-year-old boy by Baudamant in 1779.The first case of Rapunzel syndrome was published by Vaughan et al in 1968.[1] It is the rarest type of bezoar most commonly seen in females. In total, around 50 cases of Rapunzel syndrome have been reported in literature so far and only one case is described in male[2] "Rapunzel'' is the heroine in the fairy tale collection by Grimm Brothers with very long, blond hair. Rapunzel is a large trichobezoar that extends beyond the stomach, into the duodenum and jejunum to a variable extent. Bezoars are named according to their components: phytobezoars, trichobezoars, lactobezoars, phamacobezoars, polybezoars, diospyrobezoars, or biliary bezoars. The most common type is the phytobezoar, which is composed of cellulose, hemicelluloses, and other proteins. Trichobezoars, or hairballs, are rare.

CASE REPORT

An 8-year-old girl visited our paediatric surgery outpatient department with epigastric discomfort for 1-2 months. She had history of vomiting containing food particles immediately after intake of food. There was history of nausea, bloating, early satiety, and failure to gain weight. Parents also gave history of lethargy and easy fatiguability. A solid mass was palpable in the epigastric region. The patient had intermittent small hard stools, which included some hair. Her parents described that the patient had habits of hair pulling and chewing for 4-5 years. The child did not show any signs of anxiety, depression, or mental retardation during admission. There were hairless regions on her scalp in bilateral parietal regions. Ultrasonography showed echogenic mass in the stomach and pyloric region with gastric distension. A barium swallow showed intraluminal filling defect in the stomach suggestive of bezoar in gastric lumen. The patient has pallor on general examination. Her height and weight were both in the 25th percentile. Laboratory evaluations revealed haemoglobin of 7.5 g/dL, other laboratory findings, including electrolytes, acid balance, liver function tests, and renal function tests,were within normal limits. Blood transfusion was given to build up haemoglobin. Patient was undertaken for surgery. On exploratory laparotomy, the stomach was dilated. On gastrotomy, there was a large trichobezoar composed of hair bundles occupying the stomach and extending into the duodenum and jejunum. The gastric trichobezoar was removed in toto, alongwith its extension into the duodenum and jejunum. The tip of bezoar was free and was not adherent to jejunum. Gastrostomy was closed into layers and subsequently abdomen was also closed.

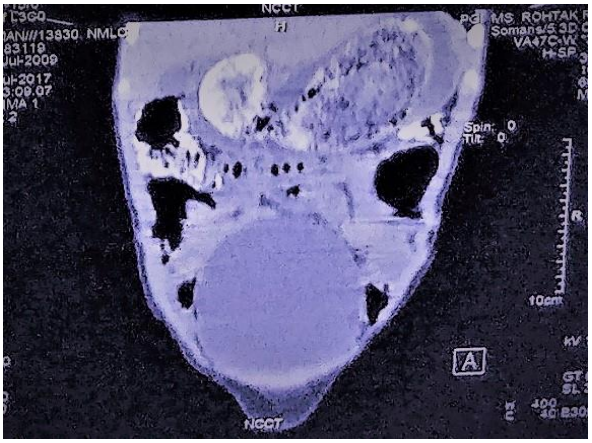

Fig. 1.

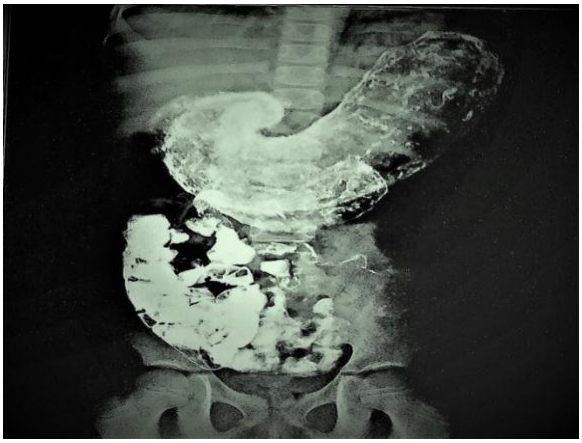

Fig. 2.

fig 3 trichobezoar along with extension up to jejunum

Fig 1 CECT film: non adherent spindle shape mass in the stomach, with the contrast mean surrounding coating and infiltrating trichobezoar extending into duodenum, Fig 2: Barium Swallow: shows radiolucent shadow occupying lumen in the stomach suggestive of trichobezoar, Fig 3: Gross specimen: large trichobezoar with tail like extension.

Patient was kept on intravenous fluids and antibiotics for 3 days and started orally after 3 days after passing stools. Stitches were removed on 10th postoperative day. The child had psychiatric evaluation in the hospital. There was only history of hair plugging and eating. There was no psychiatric component. Child personality development and mental examination were normal. Treatment was given to the family to change the child habit. She was advised regular follow-up. Biopsy report was correlating with trichobezoar.

DISCUSSION

The Rapunzel syndrome occurs predominantly in females, mostly between 5-19 years of age, and they have some associated psychiatric illness. The symptoms depend on the size of trichobezoar. Common presentation is abdominal mass, and upper gastric outlet obstruction. In literature, the complications like bleeding, perforation, ulceration and perforation or pancreatitis have been described.[3] History of trichotillomania and trichophagia along with weight loss, loss of hair, is usually present. In most of the cases, the history is prolonged upto 3-4 years, as it takes time for the hair ball to become significantly large to result in symptoms. In our present case, there was history of trichotillomania and trichophagia with hair loss. So far, only 3 cases of Rapunzel syndrome resulting in mortality have been reported.[2] The diagnosis is based on history, clinical features, ultrasonography, computerised tomography of abdomen and barium meal follow through. The mass is seen as a space occupying radio lucent mass in the stomach and duodenum.

The treatment of choice is surgical extraction of the trichobezoar in toto through gastrotomy, as recommended by Uroz et al.[4] The distal end of trichobezoar may lie free in jejunum or may be adherent to its wall at single or multiple sites. The distal end should be pulled gently so as to avoid perforation of the jejunum if it is adherent to the jejunum. Solitary or multiple jejunotomies are recommended when it is stuck to the jejunum as recommended by Duncan and Singla et

al.[5,6] In the index case, the tip was lying free in jejunum. Minimally invasive techniques are laparoscopy and endoscopy.[7,8] However, successful treatment of a Rapunzel syndrome by endoscopy or laparoscopy alone has not been reported in literature, as they are large in size and difficult to fragment. So, the treatment of choice is surgical removal.

CONCLUSION

The diagnosis of Rapunzel syndrome requires a high degree of suspicion and should be promptly managed surgically along with psychiatric evaluation.

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