Rapunzel syndrome; Report of two cases

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Abstract
Rapunzel syndrome is a rare form of trichobezoar, with less than 50 cases have been reported in literature so far, of which only four cases reported in male child. Bezoars are concretions in the gastrointestinal tract that increase in size by continuous accumulation of non-absorbable food or fibers. Patient commonly present with abdominal pain, vomiting and signs of obstruction. We are reporting a series of two cases (one boy and one girl child) of Rapunzel syndrome with one presented at age of five year.

Keywords: Trichobezoar, Rapunzel syndrome, Broken family, Psychiatric pathology, Male child, Surgery.

Introduction
Rapunzel syndrome was redefined in year 2007 by Naik et al as a trichobezoar with a tail, which extending at least up to the jejunum with symptoms suggesting obstruction[1]. It is a rare form of trichobezoar and mostly reported in young female. There have been less than 50 cases reported in medical literature with four cases were reported in male children to date. To the best of our knowledge we present the fifth male case and one of the youngest cases.

Case Reports
Case 1
A 14-year-old female child presented with a history of intermittent abdominal pain, vomiting and decrease appetite from last 6 months. There was no change in her bowel habits. In family, her parents got divorced when she was 6 years old and since then she was living with her maternal grandmother. She was developmentally appropriate and no other medical or surgical history. The physical examination revealed a firm mass in epigastric region (Fig. 1). Laboratory values revealed hypo-chromic microcytic anemia. CECT abdomen was done before referring to us and was diagnosed as trichobezoar (Fig. 2). Endoscopic removal was tried at the same centre but failed and referred to our centre for further management.

A laparotomy was done with an upper midline vertical incision and a large trichobezoar was removed through gastrotomy wound with a tail extending up to proximal ileum (Fig. 3 & 4). A naso-gastric tube was placed, and baby kept nil per os (NPO) for four post-operative days. Then gradually feed started and post-operative recovery was uneventful. Baby was discharged on 10th day.

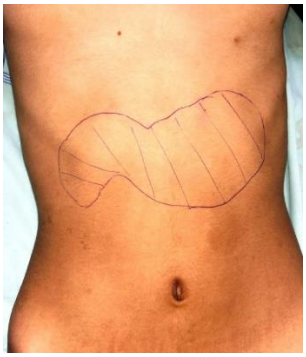

Fig. 1: Firm mass in Epigastric region

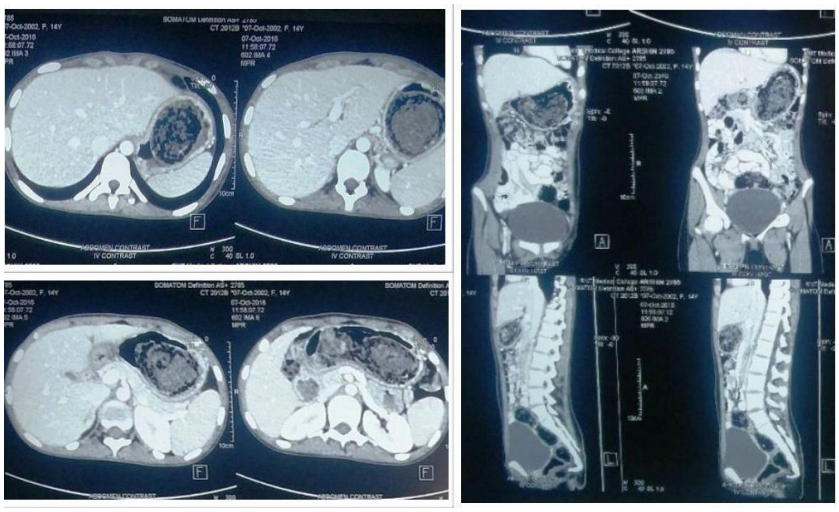

Fig. 2: CECT abdomen- axial, coronal and saggital view showing trichobezoar in stomach cavity

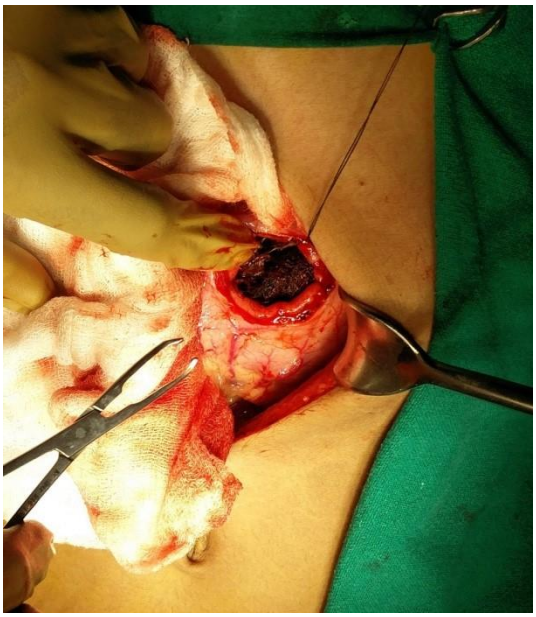

Fig. 3: Intra operative visible trichobezoar through gastrotomy wound

Fig. 4: Gastric trichobezoar with a 50cm long tail

Case 2

A five-year-old male child presented in emergency with abdominal pain for 3 days, bilious vomiting and abdominal distention for 2 days. In family history, mother died when baby was 6 months old and father is a chronic alcoholic. On physical examination baby had sign of malnutrition with an alopecic patch at left side of scalp (Fig. 5), tachycardia with distended abdomen and a firm mass palpable in the right lower abdomen with marked tenderness. Abdominal radiograph showing multiple air fluid level with dilated small bowel loops with slightly dense radio-opaque shadow, round to oblong shaped, shows lucent areas interspersed in it suggestive of air trapped in the lesion (Fig. 6). Laboratory work-up revealed anemia with increased leukocyte counts.

Baby was taken up for laparotomy and a 45 cm cylindrical trichobezoar removed through enterotomy wound (Fig. 7). He had an uneventful post-operative recovery.

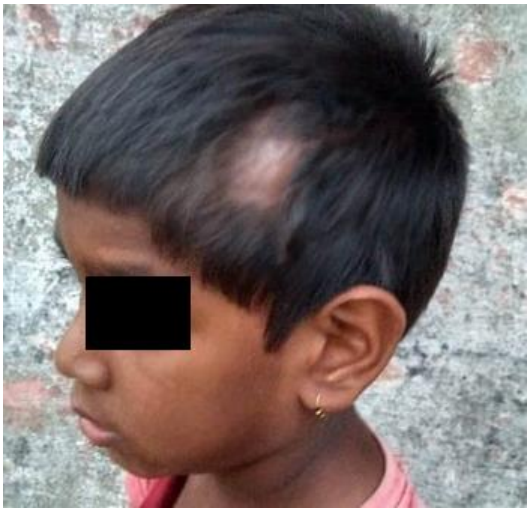

Fig. 5: Alopecic patch left side of scalp

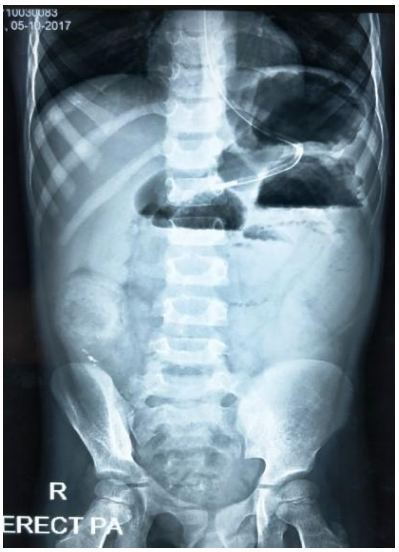

Fig. 6: X-Ray abdomen showing multiple air fluid level (broad arrow) with slightly dense radiopaque shadow, round to oblong shaped, shows lucent areas interspersed in it suggestive of air trapped in the lesion (small arrow)

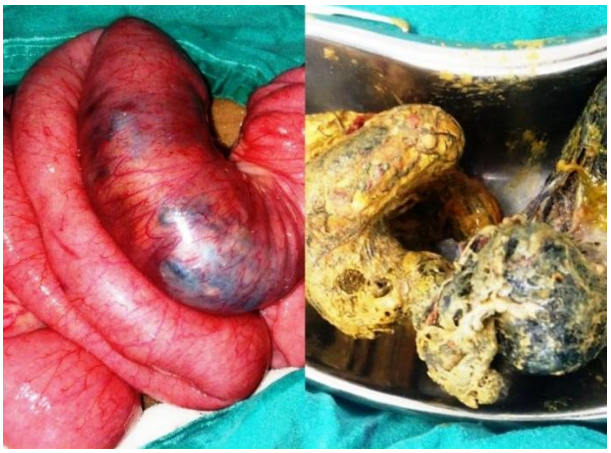

Fig. 7: Right side bowel part where trichobezoar present and left side extracted specimen

Discussion

Rapunzel syndrome was originally described by Vaughan et al.[2] in 1968 but definitive clinical criteria to label as Rapunzel syndrome was redefined in 2007 by Naik et al. According to him cases to be a Rapunzel syndrome are: (1) a trichobezoar with a tail; (2) extension of tail at least to the jejunum, and (3) symptoms suggestive of obstruction.

Bezoars are concretions in the gastrointestinal tract that increase in size by continuous accumulation of non-absorbable food or fibers.[3] Bezoars include trichobezoar (hair), phytobezoar (vegetable material), lactobezoar (milk products), pharmacobezoar (medicines), and plastobezoar (plastic materials). Rapunzel syndrome is a rare form of trichobezoar and fewer than 50 cases have been reported till date. Most of the cases have been reported in countries where women traditionally have long hair and more than 30% of cases has been reported in India may be because of same reason.[4] This disease was usually known to be occur in teen ager girl child and only one case was reported in the male child till 2010, but in the recent trends more number of cases reported in male child recent year as shown in table 1.

Table 1: Rapunzel syndrome: reported cases in male child till date

Author	Year	Age (in years)	Sex
Hirugade et al.[5]	2001	4	Male
Parachuri et al.[6]	2011	2	Male
Anantha et al.[7]	2013	9	Male
Singh et al.[8]	2013	3	Male
Jhanwar et al (present study)	2018	5	Male

The most common presenting features are abdominal pain, vomiting, obstruction and peritonitis. Less commonly patient also presented with weight loss, anorexia, hematemesis and intussusception.

The management of a bezoar needs to encompass removal of mass and prevention of recurrence. Depending on its consistency, size and location, bezoar removal may occur via endoscopy or surgery. Endoscopy or medical therapy may be effective in small size trichobezoar[9] but for condition like Rapunzel syndrome surgery still is the gold standard. As in first case of our series, endoscopic removal was tried before referring to us, but failed most probably because of large size. In these cases, trial of endoscopic removal may cause complication like perforation[7]. Many of these patient's report having parental discontent, bereavement, or other family problems as also observed in both the cases of our series. So a long term psychiatric follow-up as well as parental or spouse counseling is also advised as a regular part of treatment to prevent recurrence.

Conclusion

Rapunzel syndrome is a rare form of trichobezoar with increased incidence in male child. Any child with history of broken family presented with recurrent abdominal pain then possibility of trichobezoar must be considered in differential diagnosis. Surgery stills the corner stone in the management of Rapunzel syndrome. Many of these patients have psychiatric pathology so counseling by a psychiatrist is an important part of management to prevent recurrence.

Abbreviations: Fig- Figure, CECT- Contrast Enhanced Computed Tomography

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