Indian J.Sci.Res. 11 (1): 090-092, 2015 ISSN: 0976-2876 (Print)
ISSN: 2250-0138(Online)

RAPUNZEL SYNDROME IN A SIX YEAR OLD CHILD - A CASE REPORT

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ABSTRACT

Trichobezoar is a tuft of undigested hair mass commonly found in young females with psychiatric disorders. Rapunzel syndrome is a rare condition and occurs when gastric trichobezoar extends beyond the pylorus of the stomach into the small bowel. 28 cases have been reported in English literature till 2012. The diagnosis of trichobezoar may be difficult due to non specific presentation. We present a case of a 6 year old child who came with history of pain abdomen and features of intermittent gastric outlet obstruction. She was diagnosed with Rapunzel syndrome and successfully managed by laparotomy which revealed trichobezoar with a tail measuring 48cms in length extending till the proximal ileum. Child also had transient jejuno-jejunal intussusception.

KEYWORDS: Rapunzel syndrome, Trichobezoar, Bezoar, Trichophagia, Trichotillomania

Bezoars are collection of undigested materials that accumulate to form a mass in the gastrointestinal tract most commonly found in stomach. Various types of bezoars like trichobezoar, phytobezoar, lactobezoar, pharmacobezoar have been reported depending on their compositions. Trichobezoar is a tuft of undigested hair mass commonly found in young females with psychiatric disorders[1] .Rapunzel syndrome is a rare condition and occurs when gastric trichobezoar extends beyond the pylorus into the small bowel. 28 cases have been reported in English literature till 2012[2].

CASE PRESENTATION

A 6 year old female child presented with pain abdomen of 1 month duration for which she approached a family physician and was treated as a case of worm infestation. However, the pain in the epigastric region progressively increased during the last 15 days which was more after having food. The mother also noticed fullness in the epigastric region after taking food during the last 10 days which was followed by pain abdomen and vomiting. Fullness and pain subsided after vomiting. Examination revealed a palpable mass in the epigastrium. There was no definite history of trichophagia on questioning and child had no alopecia. Ultrasonography done elsewhere was suggestive of intussusception. CT abdomen [fig 1] showed a hypoechoic mass lesion in the body of stomach measuring about 8 × 7 cms. Barium meal [fig 2] revealed a filling defect in the lumen of stomach extending into the deuodenum with linear streaks of barium within the filling defect suggestive of a trichobezoar. Endoscopy [fig.3] was done to confirm the diagnosis which showed a trichobezoar with its tail extending into the pylorus. With these findings and repeated questioning, the child's mother admitted that she had a habit of plucking and eating hair with frontal hair loss when she was 2 years old. Endoscopic extraction using a snare was attempted but failed. A laparotomy was performed which showed a transmural ulcer adherent to the anterior abdominal wall. Gastrotomy was done and a gastric trichobezoar of 12 × 5 cms which had taken the shape of the stomach along with 48 cms tail in continuity was extracted [fig 4]. A small segment of tail which was left in the jejunum was removed by enterotomy. Postoperative course was uneventful. Child was started orally on postoperative day 3 and discharged on postoperative day 7 after psychiatric counselling.

DISCUSSION

Trichobezoar is a tuft of hair mass in the digestive tract which is a complication of trichotillomania (plucking of hair) and trichophagia (eating hair). Although about 1 of 2000 children suffer from trichotillomania ,trichophagia is rarely seen and a bezoar does not occur in all children with trichophagia[3]. Stomach is the most common site for a trichobezoar as it is not able to exteriorize hair and other substances out of its lumen because the friction surface is not sufficient for propulsion by peristalsis. Rarely these bezoars can extend beyond dueodenum upto ileocecal junction, a condition termed as RAPUNZEL SYNDROME[1]. It can present with the

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symptoms of pain abdomen, vomiting, perforation, mass per abdomen, intestinal obstruction, intussusception, pancreatitis and ulceration[1]. In our case , the child presented with the symptoms after a period of 4 years from the time of ingestion of hair. The tail of the gastric trichobezoar was about 48 cms in length extending till the terminal jejunum with transient intussusception. Imaging modalities like CT and barium meal helps in establishing diagnosis. Magnetic resonance imaging is less useful compared to a CT as the signal density is low in MRI and is confused with air[4]. Endoscopy can be used as a mode of diagnosis and also for management. Endoscopic removal of trichobezoar has been tried in 40 patients and only 2 were successful according to the literature[5]. In case of Rapunzel syndrome, endoscopic removal should not be tried as it can lead to incomplete removal with left over segments in the small bowel. Surgery is the preferred treatment in most of the cases where a gastrotomy and the enterotomy can be performed for the complete removal of the trichobezoar. In our case a gastrotomy was done and the gastric trichobezoar was removed in continuity with its tail of about 48cms and a small leftover segment was removed by a separate enterotomy. The complications of Rapunzel syndrome ranges from episodes of incomplete pyloric obstruction to complete obstruction of the bowel, ulceration, perforation, pancreatitis, peritonitis and mortality[6]. DeBakey and Oschner reported a surgical mortality of 10.4%[7]. It is mandatory to perform a thorough complete exploration of the small intestine and stomach searching for retained bezoars. Underlying psychological causes have to be treated as a part of management to prevent recurrences[1]. Few cases of recurrences have been reported even after successful surgery.

CONCLUSION

Trichobezoar should be considered as a differential diagnosis in a female patient presenting with non specific abdominal complaints. Endoscopy as a preoperative management should be done to visualise extension of trichobezoar into the small bowel. In case of Rapunzel syndrome removal of the mass by endoscopy can be incomplete because of fragmentation and laparotomy is considered as the preferred mode of treatment for complete extraction of the trichobezoar along with its tail.

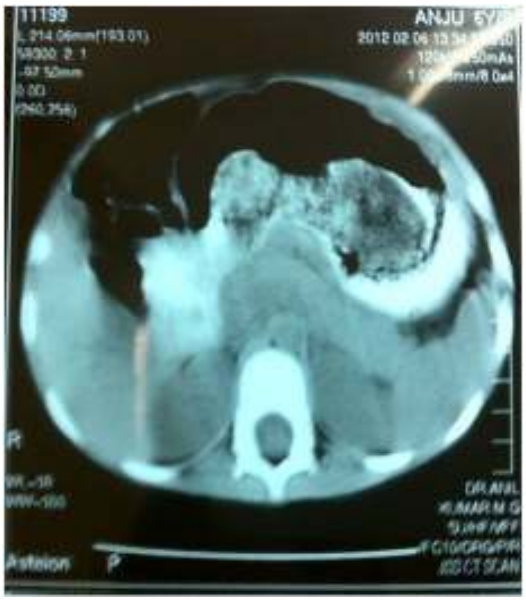

Figure 1

CT Abdomen : An ill defined Soft tissue density mass interspread heterogenously with oral contrast material noted in the lumen of the stomach extending into pylorus of stomach.

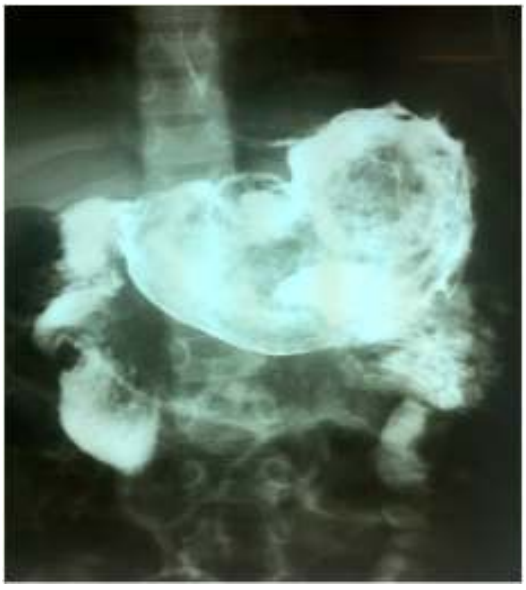

Figure 2

Barium Swallow: contrast X ray of stomach showing irregular tuft of hair extending into the pylorus of the stomach.

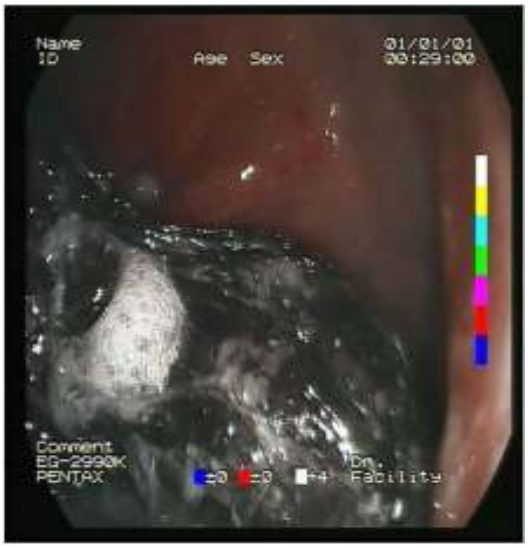

Figure3

Endoscopy : Tuft of hair mass seen in the lumen of stomach on endoscopy

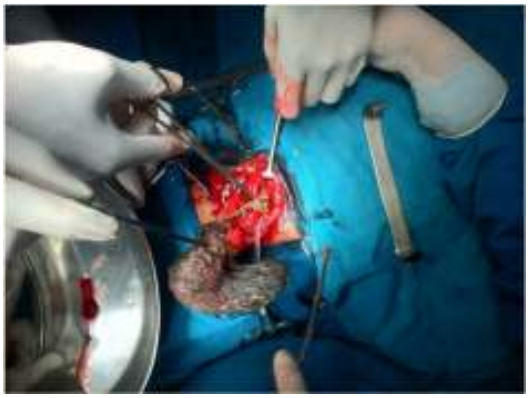

Figure4

Intra Operative image while extracting the specimen

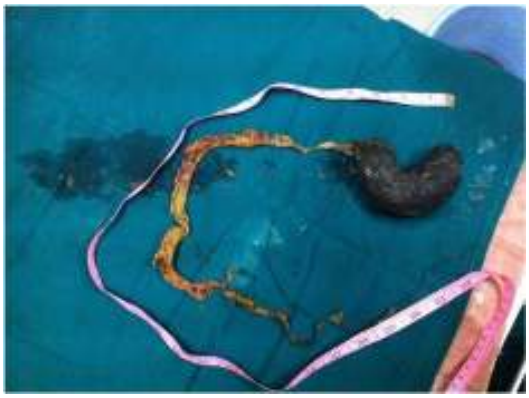

Figure5

Trichobezoar specimen with 48cms tail extending beyond the pylorus

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