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Case Report

'Rapunzel Syndrome' Trichobezoar In A 11year-Old Girl - A Case Report

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ABSTRACT

Bezoars are masses, which are commonly encountered in patients with psychiatric problems, formed by the accumulation of intraluminal non digestible substances that can lead to obstruction of the stomach and the small intestine. Rapunzel syndrome is the extension of the bezoars down to the duodenum, jejunum and colon at times, which is a rare condition. In this case report, an 11-year-old girl with trichotillomania is reported, who was admitted in our hospital with complaints of nausea, vomiting and pain abdomen. Laparotomy was done and a giant trichobezoar was identified, which completely filled the stomach, a long tail of hair extending through the pylorus up to distal transverse colon causing intussusceptions of terminal ileum was found and removed successfully. Postoperative period was uneventful Patient was discharged after six days. The parents were advised to continue the follow up in the psychiatry department..

Key words: Rapunzel, trichobezoar, intussusceptions, trichotillomania.

INTRODUCTION

Trichobezoars, commonly occur in with psychiatric disturbances patient (trichotillomania) a habit disorder, who and swallow their own Trichobezoars have been described in literature and they comprise 55% of all bezoars. In very rare cases the" Rapunzel Syndrome," hair extends through the pylorus into the small bowel causing symptom and sign of partial or complete gastric outlet [1] obstruction. In this case report psychodynamic aspects. clinical manifestations, diagnosis and therapeutic strategies of 11 year old girl having trichobezoar is discussed

CASE REPORT

11 year old girl was referred to our paediatric surgery clinic, with a history of abdominal pain, distension, weight loss and vomiting. This history was on and off for almost one year and treated in different hospitals as a case of gastritis and parasitic infestation. Despite the treatment, she was not improving; in fact she was getting worse. In our paediatric surgery clinic, the parents gave doubtful history of hair chewing by their daughter. Abdominal palpation revealed an ill-defined mass around 3 cm ×4 cm occupying the upper half of the abdomen in epigastrium extending to left hypochondrium, the mass was non tender and was firm in consistency.

Ultrasonography was done which showed a mass filling entire stomach. Girl was posted for surgery through upper mid line incision the abdomen was opened and a huge Trichobezoar was identified which took the shape of the stomach. (fig-1)







Fig-2

There was a long tail of hair extending through the pylorus, extending up to the distal transverse colon (fig-2) whichwas causing intussusceptions of terminal ileum. By this feature the diagnosis was clear of a Rapunzel syndrome. A distal enterotomy was performed and the

remaining part of hair tail was dislodged gently. Ileo-ileal intussception was manually reduced simultaneously. Both gastrotomy and enterotomy in the gastric wall and small bowel were closed with continuous sutures (polyglycolic acid). The patient had an uneventful postoperative course and was discharged after six days. The parents were advised to continue the visit in the psychiatry department for the follow up of trichotillomania.

DISCUSSION

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. [2] The postulated reason for formation in the stomach is that hair is indigestible and due to its smooth nature cannot be propelled with peristalsis and over time forms a bezoar within the stomach. Extension of the bezoar from the stomach into the jejunum or further on is referred to as "Rapunzel syndrome," first described by Vaughan Jr. et al. in 1968, [3] Patients may remain asymptomatic for many years until the trichobezoar grows to the point of obstruction. The typical presentation of trichobezoars and Rapunzel syndrome includes palpable abdominal mass, chronic abdominal pain, nausea vomiting and constipation. Some patients present with weight loss, anorexia, hematemesis, iron deficiency anaemia, and intussusceptions. [2,4] The reported complications of Rapunzel include gastric syndrome ulceration, incomplete pyloric obstruction, obstructive acute pancreatitis, iaundice, complete intestinal obstruction, gastric perforation, peritonitis, and mortality. [5-8] Elucidating a patient's history of trichotillomania and/or trichophagia is essential for the diagnosis of Rapunzel syndrome or trichobezoars. In some patients, putrid halitosis and patchy alopecia can be clues on physical Imaging studies including examination.

plain abdominal radiograph, contrast upper gastrointestinal series, and abdominal CT scan may show the trichobezoar as a mass or filling defect in the stomach and small bowel. Among these modalities, CT scan with contrast can delineate the extension of trichobezoars. ^[9] Upper gastrointestinal endoscopy is considered to be the gold standard. The abdominal CT scan is the most accurate imaging test concerning the presence of trichobezoars. ^[10]

Surgery is indicated when a very large or solid bezoar causes perforation or haemorrhage, or in the case of Rapunzel syndrome, when there is significant extension of the bezoar. Surgical removal is accomplished by gastrotomy or enterotomy. Traditionally, a gastric trichobezoar was removed by gastrotomy through an upper midline laparotomy. [11]

CONCLUSION

A long history of gastrointestinal problem, in a paediatric age group with history of trichophagia, early endoscopy is recommended. All patients with Trichobezoar should be referred for psychiatric evaluation after surgery to avoid recurrence.

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