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Rapunzel Syndrome' Trichobezoar in a 27-Year-Old Lady : A Case Report

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ABSTRACT *Trichobezoars (hair balls) are usually located in the stomach, but may extend through the pylorus into the duodenum and small bowel (Rapunzel syndrome). They are almost always associated with trichotillomania and trichophagia or other psychiatric disorders. Though several treatment options are proposed, conventional laparotomy was the treatment of choice. A case of trichobezoar extraction by gastrotomy is reported here.*

KEYWORDS : Trichobezoar, Rapunzel syndrome, Laparotomy, Anterior gastrotomy

Introduction

Trichobezoar, a hair ball in the proximal gastrointestinal tract, is a rare condition almost exclusively seen in young females . Human hair is resistant to digestion, it accumulates between the mucosal folds of the stomach. Over a period of time, continuous ingestion of hair leads to the impaction of hair together with mucus and food, causing the formation of a trichobezoar. In most cases the trichobezoar is confined within the stomach. In some cases, however, the trichobezoar extends through the pylorus into jejunum, ileum or even colon. This condition, called Rapunzel syndrome, was first described by Vaughan et al. in 1968 [2]. In addition, incidentally, parts of the tail can break off and migrate to the small intestine, causing intestinal obstruction [3–5]. One should be aware of a trichobezoar in young females with psychiatric comorbidity.

Case report

A 27 year old female patient was presented with vomiting, abdominal discomfort on and off for the past two years. Careful history was recorded and clinical examination was done. On clinical examination, a mass in the epigastrium was observed extending to umbilical region, firm to hard in consistency and mobile. Endoscopy was done for diagnosis and it showed Bezoar in stomach and duodenum causing partial obstruction. CT scan abdomen was done which showed Trichobezoar in the stomach extending into the small bowel up to the ileum with features of intussusception (?. jejuno ileal). She was resuscitated; anaemia, hypoproteinemia were corrected. On laparotomy, stomach, duodenum, jejunum, and proximal 3rd of ileum were dilated. Anterior gastrotomy was done (Fig.1). The hair ball was removed enmass and its length was 115 cm (Fig.2). The stomach wall was closed in two layers - inner 2-0 vicryl, outer 2-0 silk, as continuous sutures. Postoperative period was uneventful. Symptomatic treatment was given with antibiotics, analgesics and supportive treatment. She was discharged in a good condition. She is doing very well now.

Discussion

In our case, the presentation is with hairball extending down to the small bowel, causing symptoms of gastric outlet obstruction. Rapunzel Syndrome is a rare form of trichobezoar. The commonly accepted definition is that of a gastric trichobezoar with a tail extending to the jejunum, ileum or the ileocecal junction. It is common in young females usually with mental retardation and an underlying psychiatric disorder; almost half of the patients present with trichophagia [2]. Sometimes, the aggregate of hair fragments and small pieces do pass through into the intestine, thus leading to sequelae such as ulceration, partial or total obstruction, intestinal perforation and peritonitis. The complications of Rapunzel syndrome ranges from attacks of incomplete pyloric obstruction to complete obstruction of the bowel to perforation to peritonitis and mortality [1,2].

Majority of cases of trichobezoar present late, due to the low index of suspicion by the physician, like our case. Patients are often asymptomatic, but may be presented with nausea, vomiting, anorexia, weight loss, vague abdominal pain or constipation which were the symptoms noticed in our case. The diagnosis is based on a combination of good history taking, and careful clinical examination. An abdominal radiograph may show a prominent gastric outline with an intragastric mass, outlined by gas in the distended stomach. Abdominal ultrasound shows a dense, echogenic rim with sharp, clear posterior shadowing in the epigastrium, Abdominal CT shows a mobile intragastric mass with a mixed density pattern due to the presence of entrapped air and food debris, and it will also delineate the extension of the trichobezoar. Endoscopy is diagnostic, in almost all cases. CT scan with contrast will delineate the extension of trichobezoar. Treatment may include endoscopic removal but with risk of bowel perforation, so this should be restricted to small trichobezoars only. In the early stages endoscopic removal is not without risk of bowel perforation and should be resolved for small Trichobezoars only [6]. Other methods including chemical dissolution, mechanical fragmentation, laser ignited mini-explosive technique were used successfully [7]. Laparoscopy has been also used with limited success but can be tried for cases depending upon the expertise [3]. Open surgery still remains the corner stone of large Trichobezoar removal especially if it has an extension into the bowel, which might be missed with other methods of treatment.

There have been few cases of recurrence following successful surgery [8]. Recurrence of Rapunzel Syndrome has been noted and it is seen to occur because the underlying emotional stress trigger was not corrected. However, treatment and psychological support of the mental as well as physical disorder is important for prevention of its recurrence.

Conclusion

Trichobezoar is an uncommon cause of obstruction to be thought of particularly in young females. If there is a long history of a gastrointestinal problem in young age with history of trichophagia, early endoscopy is recommended. As far as treatment is concerned, however, we consider conventional laparotomy to be the best choice in patients with trichobezoar and to be the only valid treatment in young females with Rapunzel syndrome. A multidisciplinary approach including psychiatric evaluation and proper management after surgery should be adopted to prevent recurrence of the disease.

Fig. 1 Removal of trichobezoar by anterior gastrotomy

Fig. 2 Showing removed hairball of length 115 cm

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