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Surgery

CASE REPORT: RAPUNZEL SYNDROME WITH MULTIPLE GASTRIC ULCERS PRESENTING AS GASTRIC OUTLET OBSTRUCTION.

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ABSTRACT Rapunzel syndrome is an unusual type of trichobezoar in which swallowed hairs are present into stomach with its tail extending into small intestine. It may presents with abdominal lump, pain abdomen, nausea and vomiting, constipation or diarrohea.

This was a case of an 18 years female having Rapunzel syndrome with multiple gastric ulcers presenting as gastric outlet obstruction. Patient was managed by exploratory laparotomy with anterior gastrotomy and removal of trichobezoar.

KEYWORDS : Rapunzel syndrome, gastric outlet obstruction, gastric ulcers, gastrotomy

Introduction

Bezoars are collections of indigestible foreign materials usually in stomach. Term have its origin from Arabic word *badzehar* meaning "protection from poison" [1]. Virtually exclusive in young psychiatric female patients with history of ingestion of hairs [5]. Phytobezoar is most frequent type of bezoar in adults, while trichobezoar is more common in children and teenage girls.

Rapunzel syndrome is bezoar extending into the small intestine. Patients presentation may be with abdominal lump, pain abdomen, nausea and vomiting, weight loss, malnutrition, hematemesis, diarrhea, or constipation.

Case summary

An 18 years aged daily wager lady, presented with complaints of sudden, progressive, colicky pain for last 5 months and repeated episodes of vomiting for last 3 months. Vomitus was preceeded by nausea, contained food particles and usually occurred 1-2 hours after meals. Patient also noticed left upper quadrant abdominal lump for last one month. There was history of reduced appetite and significant weight loss. History of consumption of hairs was given by parents. There was no history of periodicity of pain, fever with night sweats, malena, haemetemesis or contact with pets. There was no history of drug abuse, physical abuse in childhood or psychiatric illness.

She was anemic and poorly nourished with BMI of 17. Bilateral pitting type pedal edema was present. A non tender hard intra-abdominal lump of size 15X12cm with well defined inferior and lateral margins moving well with respiration was present in left hypochondrium , epigastric and umbilical regions. Deep palpation over lump revealed crepitations. Lump was dull on percussion. A provisional diagnosis of gastric outlet obstruction was made.

Her hemoglobin was 8.2gm%. Ultrasound abdomen performed revealed normal study which was not corroborating with our findings, so, CECT abdomen was planned. CECT abdomen revealed grossly distended stomach with large intra luminal mass having mottled appearance due to air trapped between it. Outline of the mass was demarcated by fluid in stomach. There was abrupt cut off of bowel lumen at distal ileum (well defined narrowing at distal segment of ileum with bird beak deformity). The proximal small bowel loops were grossly dilated with maximum diameter of 4cm, distal bowel collapsed. The diagnosis of gastric trichobezoar extending to small intestine was made.

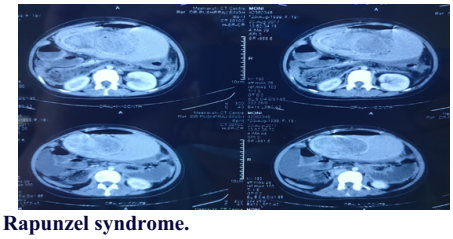

Fig. 1: Rapunzel syndrome.

Upper GI endoscopy confirmed the diagnosis of trichobezoar with complete obstruction of pylorous causing gastric outlet obstruction.

Midline laprotomy performed and it revealed distended stomach with normal appearing serosa. Gastrotomy was done and trichobezoar was removed along with its long tail extending into small intestine.

Fig 2. Specimen.

Once trichobezoar was removed multiple gastric ulcers of variable sizes were seen.

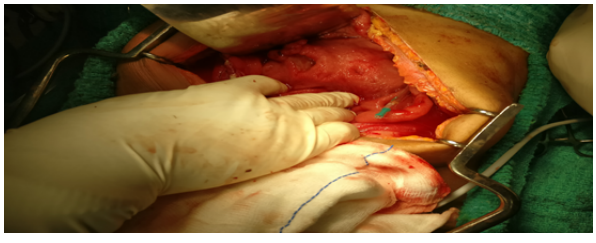

Fig 3. Multiple gastric ulcers (Ryles tube also visible).

Thorough irrigation of stomach and peritoneal cavity was done and anterior gastric wall was closed in two layers. Patient made an uneventful recovery and discharged on post operative day7.

Discussion:

Bezoars are classified according to their composition as: phytobezoars, consisting of vegetable matter, pharmacobezoars or medication bezoars, lactobezoars, occur secondary to infant formula and consist of milk curd, and trichobezoars, which are conglomerations of hair or hair like fibres. Phytobezoars are more commonly found in patients who have undergone surgery for stomach and have impaired gastric emptying [2]. Diabetics and autonomic neuropathy are also risk factors.

Trichobezoars result from compulsive pulling out of hair (trichotillomania) and then swallowing the hair (trichophagia). The common presentation of trichobezoar is in young females usually with underlying psychiatric disorders; in their second decade of life [3]. Trichobezoar is seen in around 1% patients with history of trichophagia [4]. When hairs are ingested, due to their indigestibility, they become entrapped within the mucosal folds, attract more hairs

and become large sized as days pass. As more hair accumulates, peristalsis causes it to be enmeshed into a ball. The ball of hairs slowly enlarges to acquire the shape of stomach, usually as a single solid mass. Bezoar become black colored due to denaturation caused by acid produced by stomach.

Rapunzel Syndrome is an uncommon presentation of trichobezoar, in which strands of swallowed hairs extend as tail through small intestine, beyond the stomach. Rapunzel syndrome is named after 'Rapunzel' – the maiden in the Grimm brothers' fairy tale whose long hair flowed out of her prison tower allowing her prince to rescue her. Vaughan et al first described it in 1968 [5]. Rapunzel syndrome is generally defined by: (a) the presence of a trichobezoar with a tail, (b) the extension of the tail to at least the jejunum and (c) the symptoms suggestive of obstruction [6].

Majority of cases of trichobezoar present late, due to the low index of suspicion by the physician. The common presentation is in young females usually with underlying psychiatric symptoms as palpable abdominal mass, abdominal pain, nausea and vomiting, weakness and weight loss, constipation or diarrhea, early satiety and haematemesis. They may extend in duodenum and jejunum as in our case and may present as a case of gastric outlet obstruction [7] or partial intestinal obstruct presenting with abdominal pain.

With high index of suspicion, trichobezoars can be diagnosed with imaging and endoscopic techniques. Abdominal Ultrasound shows a dense, echogenic rim with sharp, clear posterior acoustic shadowing in the epigastric region, but it is not pathognomic [8]. Contrast-enhanced CT or barium studies are investigations of choice to confirm the diagnosis. In barium study, the hair ball gets evenly coated with a small amount of contrast and becomes visible. It also helps to detect extension, dislodgment (satellite masses) or synchronism. On CT scan, trichobezoars in the stomach appear as a concentric inhomogeneous mass with entrapped air surrounded by contrast material. The presence of tail is reflected by small round hypodensities [9]. Upper GI endoscopy is diagnostic as well as therapeutic for smaller trichobezoars [10].

Treatment of bezoar consists of removal of bezoar and prevention of recurrence by addressing the underlying physical or emotional cause. Trichobezoars are resistant to enzymatic dissolution. Depending on its consistency, size and location, bezoar removal can be performed by endoscopy or surgery. Small to moderate sized trichobezoars can be removed by laparoscopy [11, 12] or minilaprotomy [13]. Surgery in form of laprotomy with gastrotomy or enterotomy is indicated in case very large bezoars, solid bezoar causing perforation or hemorrhage, or in case of Rapunzel syndrome, when there is significant extension of the bezoar. Patient should be started on tricyclic antidepressants as trichotillomania is classified as obsessive-compulsive disorder. The patient's long-term prognosis is excellent if behavioral therapy is used to control trichophagia, and psychological/psychiatric follow-up is maintained.

Conclusion

Rapunzel syndrome though uncommon, should be included in differential diagnosis of young female patients with a history trichophagia along with symptoms of gastric outlet syndrome, presenting as, abdominal lump and pain, nausea and vomiting. It can be diagnosed with upper GI endoscopy, CECT abdomen and barium studies. Management options for the treatment of the Rapunzel Syndrome include surgical removal by laparotomy or laparoscopically, while laparotomy is widely considered as the treatment of choice for complicated trichobezoars. Patient must be treated and Counselled by a psychiatrist so as to undergo behavioral changes, so that recurrence can be prevented.

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