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Case Report

An unusual presentation of Rapunzel syndrome, a double impacted trichobezoar

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Abstract: Bezoars are collections of interwoven organic or inorganic matter found in the alimentary canal. Rapunzel syndrome represents a rare presentation of trichobezoar where the tail of main gastric bezoar extends into the lumen of small intestine. We present the case of a 09 year old female child who presented to us as a case of acute intestinal obstruction as a rare manifestation of Rapunzel syndrome, with migration of gastric bezoar into the small intestine. Successful surgical retrieval was performed in emergency scenario and the patient had an uneventful recovery.
Keywords: Trichobezoar, Rapunzel syndrome, Trichotillomania, Trichophagia, Intestinal obstruction, abdominal mass.

INTRODUCTION

The term bezoar is derived from Persian word ‘bedzehr’ or ‘padzehr’ meaning anti-poison. Bezoars medically are a collection of tightly interwoven indigestible matters present in the alimentary canal. Subsequent nomenclature is governed by the constituent matter of the bezoar as hairs- trichobezoar, vegetables- phytobezoar, and milk curd- lactobezoar. Rare forms of bezoars as stone and fur bezoars have also been reported in literature. Trichobezoars are the most common of the bezoars comprising more than 50% of the bezoars described in the literature. Rapunzel syndrome has been described in the literature as a rare presentation of trichobezoar where the tail of main gastric bezoar extends through the pylorus to small intestine causing partial or complete gastric obstruction [1]. It results from a psychiatric illness trichotillomania in which the patient achieves pleasure and fulfillment through pulling and ingesting hair (trichophagia). The problem is almost exclusively seen in young females who may also show accompanying features of depression, anxiety or poor self image and esteem [2, 3].

CASE PRESENTATION

A 09 year old female child who presented to us in the surgical emergency with pain abdomen, recurrent episodes of bilious vomiting, obstipation and distension of abdomen of 03 days duration. There was no history of fever, loose motions, gastrointestinal bleed, and similar episodes in past or the presence of any medical co-morbidity. On examination the child had tachycardia with a pulse rate of 110/minute. The child was afebrile, had a normal blood pressure and a normal hydration status. Per abdomen examination showed marked distension, no features of peritonitis, the bowel sounds were absent. Digital rectal examination showed an empty rectum. Supine and upright X ray abdomen (Fig-1) showed dilated small bowel loops. Ultrasound abdomen suggested grossly dilated small bowel loops with poor peristalsis. The child was resuscitated with intravenous fluids and taken up for emergency exploratory laparotomy as a case of acute intestinal obstruction.

Per operative findings included minimal ascites, large sized trichobezoar measuring 10 cm X 4.5 cm with a distal tailing of hair strands (Fig-2) stuck in jejunum 20 cm distal to the duodenojejunal flexure with collapsed proximal jejunal loop, smaller trichobezoar measuring 4.5cm X 3.5cm with proximal tailing of hair strands (Fig-3) stuck in terminal ileum 10 cm proximal to the ileocaecal junction with collapsed distal ileum and the large bowel. Proximal larger trichobezoar had assumed the shape of stomach confirming its gastric origin.

Both bezoars were retrieved through separate enterotomies considering the risks involved with in situ manipulation of unknown foreign bodies in trying to achieve a single enterotomy. Both enterotomies were repaired primarily. Post operative period was uneventful. In the post operative period the child confessed to ingestion of hair stuck in the comb of herself and her mother over a period of 02 years.

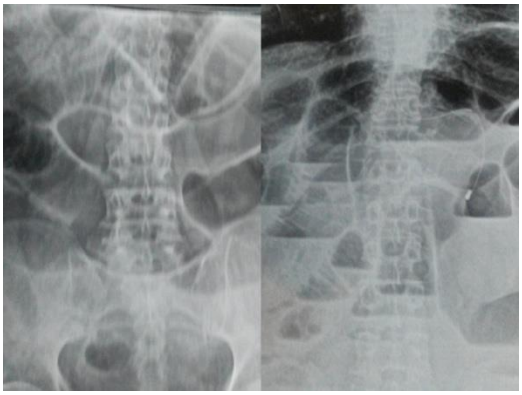

Fig-1: Supine and upright X ray abdomen

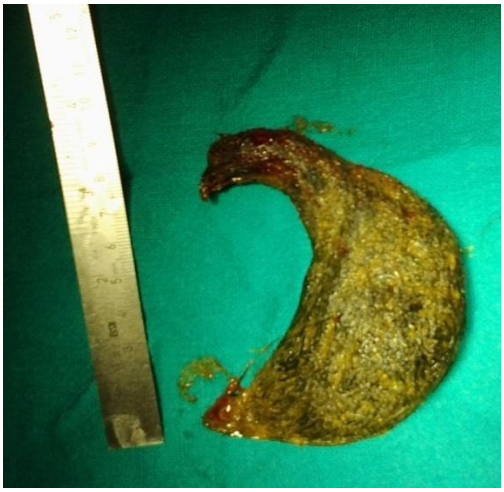

Fig-2: Trichobezoar measuring 10 cm X 4.5 cm with a distal tailing of hair strands

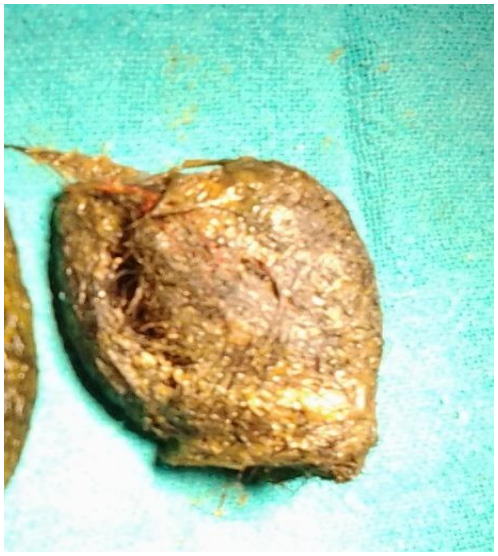

Fig-3: Trichobezoar measuring 4.5cm X 3.5cm with proximal tailing of hair strands

DISCUSSION

The common presentation of tichobezoar is in a young female with an underlying psychiatric disorder. Trichobezoar in itself is a rare entity as its prevalence in patients with trichophagia has been described to be less than 1% [4]. Stomach is the most common site for trichobezoar formation as the ingested hair strands escape peristaltic propulsion and are retained in prepyloric gastric folds and persistent peristalsis enmeshes them into an interwoven ball. Studies have linked trichobezoar to childhood neglect, abuse, mental retardation or bereavement [5, 6]. Majority of the cases are detected late due to an ignorant patient and rarity of the condition resulting in a low index of suspicion amongst the physicians.

The patient usually presents with a palpable abdominal mass (87.7%), abdominal pain (70.2%), nausea and vomiting (64.9%), weakness and weight loss (38.1%), obstruction or diarrhea (32%) and low hemoglobin (62%) [7].

Treatment modalities available include endoscopic therapy and surgical intervention. Endoscopic therapy is more effective for soft bezoars like phytobezoars (vegetable) and lactobezoars (milk and curd). Use of specialized bezotomes and bezotriptors that pulverize and fragment the bezoars has been described in literature [8]. However technical intricacies involved and limited availability of these tools offset their advantages over a formal laparotomy.

CONCLUSION

Trichobezoar is a rare but still under diagnosed clinical condition. This should be considered as differential diagnosis in a young female patient presenting with abdominal mass, intestinal obstruction or pain abdomen and a specific history of pica should be obtained. Patient has associated psycho-social issues that need to be addressed to avoid a recurrence. Also as cases of trichobezoar and further Rapunzel syndrome themselves being rare, our case presentation of a gastric trichobezoar migrating and causing small intestinal obstruction due to double impaction with its satellite tuft of hair becomes still rarer.

REFERENCES

1. Phillips MR, Zaheer S, Drugas GT; Gastric trichobezoar: case report and literature review. Mayo Clin Proc 1998, 73**:** 653-656.
2. Diefenbach GJ, Reitman D, Williamson DA; Trichotillomania: a challenge to research and practice. Clin Psychol Rev 2000, 20:289–309.
3. Carr JR, Sholevar EH, Baron DA; Trichotillomania and trichobezoar: a clinical practice insight with report of illustrative case. J Am Osteopath Assoc, 2006, 106: 647–652.
4. Irving PM, Kadirkamanathan SS, Priston AV, Blanshard C. Education and imaging. Gastrointestinal: Rapunzel syndrome. J Gastroenterol Hepatol, 2007; 22:2361.
5. Gockel I, Gaedertz C, Hain HJ, Winckelmann U, Albani MI, Lorenz D; The Rapunzel syndrome: rare manifestation of a trichobezoar of the upper gastrointestinal tract.] Chinurg, 2003; 74:753-756.
6. Sharma NL, Sharma RC, Mahajan VK, Sharma RC, Chauhan D, Sharma AK; Trichotillomania and

trichophagia leading to trichobezoar. J Dermatol, 2000; 27:24-26.
7. Gupta Naik S, Naik S, Chaudhary AK, Jain P, Sharma A; Rapunzel Syndrome Reviewed and Redefined. Dig Surg, 2007, 24:157-161.
8. Wang YG, Seitz U, Li ZL, Soehendra N, Qiao XA; Endoscopic management of huge bezoars. Endoscopy, 1998; 30:371–374.