Case Report ISSN (O):2395-2822; ISSN (P):2395-2814

Rapunzel Syndrome: A Case Report and Review of Literature

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Received: March 2020
Accepted: March 2020

ABSTRACT

Trichobezoar is a collection of dense mass of hair in stomach and when this extends into small intestine it is called Rapunzel syndrome (RS). We report here, a case of RS in an adolescent girl who had presented with epigastric pain and swelling. Gastroscopy confirmed the presence of trichobezoar. She underwent upper midline laparotomy with gastrotomy and a large dense mass of hair. Extending up to first part of duodenum was removed. Her parents revealed their daughter's impulsive nature of scalp-hair Pulling. Following surgery, psychiatric consultation was sought to prevent recurrence. Trichobezoar is a very rare cause of upper abdominal mass and should be considered if there is a very strong history of impulsive hair pulling. Surgical removal of a large trichobezoar is the only treatment of cure followed by psychiatric treatment and counseling to prevent recurrence.

Keywords: Rapunzel Syndrome, Trichobezoar.

INTRODUCTION

''Rapunzel'' was the name of the maiden in the ''Grimm brothers'' fairy tale in 1812, whose long hair flowed out of her prison tower allowing her prince to rescue her.[1] Because of the resemblance of the tail of trichobezoar extending into the small intestine to the hair of Rapunzel, this condition is given the name-Rapunzel syndrome (RS). RS was first described in the literature by Vaughan et al. in 1968.[2] In which a dense compact mass of hair (trichobezoar) was found in the stomach with extension into intestine through duodenum in patients with a history of psychiatric disorder. Trichotillomania is the habit of hair pulling and trichophagia is the morbid habit of chewing the hair. Bezoars are compact masses formed of indigestible materials found in stomach. The term ''bezoar'' is believed to originate from the Arabic word ''badzehr'', Persian word ''padzahr'' or the Turkish word "panzehir", all of which means substance that acts as antidote or counter poison.[3] Bezoars are classified according to the content of the bezoar, such as trichobezoar (hair), phytobezoar (vegetable fibers), lactobezoar (milk products), pharmacobezoar (drugs), diospyrobezoars (persimmon fibres), cotton bezoars (cotton fibers).[4] Trichobezoars were first described by baudomont in 1779.[5] The presentation of RS forms a wide spectrum, varying from asymptomatic state to gastrointestinal ulceration, obstruction or perforation in young psychiatric patient.

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CASE REPORT

A 14-year-old adolescent girl presented with loss of scalp hair of 2 months duration; upper abdominal pain and vomiting since last 1 month duration. She was apparently in good health 2 months before when her parent noticed lack of scalp hair growth and gradual loss of scalp hair mostly from the sides. There was no itching and discharge from the scalp. She did not use any unusual ''hair tonic'' or ''conditioner''. One month prior to admission, she developed mild dull aching constant upper abdominal pain without any food-pain relation, flatulence or retrosternal burning sensation. Soon she noticed mild slowly increasing upper abdominal swelling, gradual loss of appetite and occasional vomiting. There was no jaundice, hematemesis, fever, breathlessness, palpitation, cough, oliguria, and hematuria. Her diet was mixed and her bowel movements were normal. Her past and family history was unremarkable. She belonged to poor socioeconomic status. She was adequately immunized, unmarried with normal menstrual history. Clinically anxious looking with normal vital parameters, BMI - 17kg/m2, mild anemia, mild bilateral pitting pedal edema extending up to the knees. There was no jaundice, lymphadenopathy, or raised jvp. Abdominal examination revealed small epigastric swelling moving with respiration, normal skin texture, and absent distended veins. A 6×4cm2 firm, non-tender, well-delineated lump occupying the epigastrium, with smooth surface, non-pulsatile, moving well with respiration, no rigidity or guarding. There was no other abnormal intra-abdominal mass, no succussion splash, no free fluid, no bruit, normal bowel sounds were present, all hernial sites were normal. Per rectal examination was normal. Other Systemic examination- Scalp-

non-scarring, non-scaly frontal, temporal baldness with irregular outline, broken hairs with variable hair length- all features suggestive of traumatic alopecia (patients profile photograph [Figure 1]). Mother gave very reliable history of her daughter's intermittent aggressive behavior with impulsive pulling of scalp hair followed by chewing, for the past few months. A provisional diagnosis of trichotillomania, trichophagia with a strong possibility of trichobezor with nutritional defeciancy was entertained.

Figure 1: Photograph of patient with hair loss

Laboratory investigations

Hb- 9gm%, TLC- 8500/cumm., DLC (P-77%, L-20%, E-02%, M-01%), RBC- hypochromic, microcytic, Platelets—2.5 Lakh/cumm. Urine examination was normal, LFT- normal, blood urea-25mg%, s. Creatinine - 0.8mg%, s. electrolytes-normal. Plasma glucose 109mg%, s. Iron 23 microgm%, s. Ferretin-11mg%.

Abdominal ultrasound– Hyper-echoic curvilinear strips with dense acoustic shadow in the epigastric region.

Barium studies- distended stomach with flocculations of barium giving it a mottled appearance with dense acoustic shadow in stomach [Figure 2]. Upper GI Endoscopy- a mass made up of black hair with entangled food and fibrous materials occupying the entire stomach and extending into first part of duodenum.

CECT abdomen- a large area of hypointense signal intensity seen in stomach on all pulse sequences, shaped according to gastric lumen and extending into proximal part of duodenum.

She underwent upper midline laparotomy with gastrotomy. A large trichobezoar, occupying fundus, body, greater curvature, lesser curvature extending through the pylorus into the first part of duodenum was removed [Figure 4a and 4b]. Gastric biopsy specimen histopathology revealed chronic non-specific gastritis. She made uneventful recovery. Psychiatric consultation was taken and she was diagnosed as trichotillomania and managed successfully using the combination of pharmacotherapy and a package of behavior therapy. She has been on regular psychiatric OPD follow up and there is no recurrence.

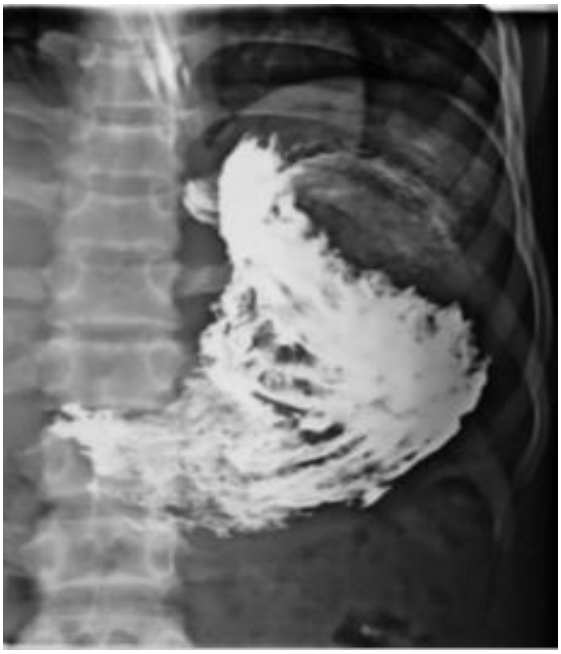

Figure 2: barium study

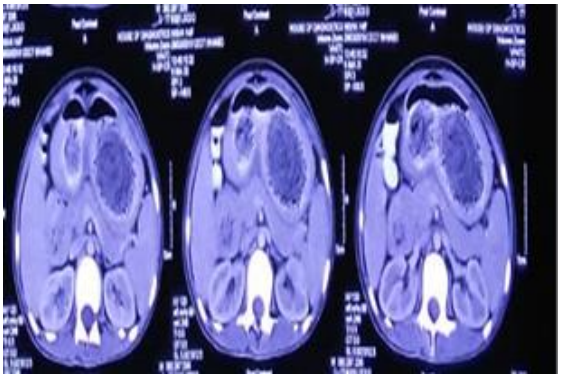

Figure 3: CECT whole abdomen

DISCUSSION

We present here an adolescent girl with RS, an uncommon form of trichobezoar. RS was first described in literature by Vaughan et al. in 1968.[2] Most cases of RS are reported by surgeons, pediatricians, and gastroenterologists.[6-13] Very few cases are reported in psychiatric literature. An explanation for such disparity is due to the fact that most cases of Trichotillomania are referred to psychiatrist early before the development of rs.[14,15] The formation of trichobezoar is complex. It occurs

in psychiatric patients with trichotillomania and trichophagia.[15] The formation of trichobezoar in these patients depends upon the quantity and duration of trichophagia, on an average only 1% patients with trichophagia develop trichobezoar.[8,16,17] The slippery surface of the hair tufts, resist the gastric peristaltic propulsion and tends to collect in the gastric mucosal folds. As more and more hair accumulates, a ball of hair is formed which becomes too large to escape and results in gastric atony and subsequently takes the shape of stomach.[9,18] Gastric mucous accumulates over the bezoar to make it glistening black. Decomposition and fermentation of fat over the bezoar imparts putrid smell to the patient's breath.[10] The acidic contents of the stomach denature hair protein and give the bezoar a jet black color.[11] The clinical presentation forms a very wide spectrum depending on the duration and quantity of trichophagia. The commonest presenting symptoms are abdominal pain, nausea, early satiety, and vomiting.[6-13] An upper abdominal mass remains the commonest presenting sign.[18] The signs and symptoms are due to mechanical effect of the mass and malabsorption of different nutrients. Other less common presenting features are gastrointestinal ulceration, obstruction, hemorrhage, perforation with peritonitis, acute pancreatitis, obstructive jaundice, and gastric emphysema.[12] Other malabsorption related complications include protein losing enteropathy, iron deficiency, and megaloblastic anemia. The gastrointestinal perforation and peritonitis are largely responsible for mortality which is about 30%.[8] Our patient presented with upper abdominal mass, and recurrent vomiting a high index of suspicion for trichobezoar arises from the classical clinical presentation in the background of psychiatric illness.[7,8] Various diagnostic modalities confirm the diagnosis. Abdominal ultrasounds reveal an intraluminal mass with hypo echoic arc-like surface and dense acoustic shadow. An abdominal CT scan shows a well-circumscribed lesion, composed of concentric whorls of different densities with pockets of air enmeshed within it, appears in the region of the stomach. Oral contrast fills the more peripheral interstices of the lesion and a thin band of contrast circumscribed it. The absence of significant post intravenous contrast enhancement precludes a neoplastic lesion. MRI abdomen shows a large mass of hypointense signal in stomach on all pulse sequences shaped, according to gastric lumen with extension into small intestine in case of RS .The gold standard for diagnosis is Gastro-duodenal endoscopy. The presence of a dark greenish brown or black intragastric mobile mass with a slimy surface and a strong odor due to decomposition of various organic residue interspersed with hair is confirmatory. The management of rs includes the removal of the trichobezoar and prevention of recurrence by psychiatric treatment of the underlying psychiatric illness. Removal of trichobezoar is done either by surgery or by Endoscopy. Endoscopic removal is more effective for small trichobezoars, also for phytobezoars and lactobezoars but not for large trichobezoars as in rs. Specialized bezotomes (device that pulverize bezoars) and bezotriptors (devices that fragment bezoars by acoustic waves) have been used to fragment large and solid trichobezoars. Surgical removal is recommended in a very large trichobezoar causing perforation or haemorrhage.[8] Surgical removal is done traditionally by upper midline laparotomy and gastrotomy.[7,8] Minimally invasive surgery is now available for small to moderate size trichobezoars6. Various other methods like lithotripsy, intragastric administration of pancreatic lipase, cellulose, acetylcystine are met with various success. Our patient underwent midline laparotomy and gastrotomy for the removal of trichobezoar extending into the duodenum. Recurrence of trichotellomania and trichophagia as a cause of rs is prevented by managing the underlyieng psychiatric ailment using combination of pharmacotherapy and behavior therapy.

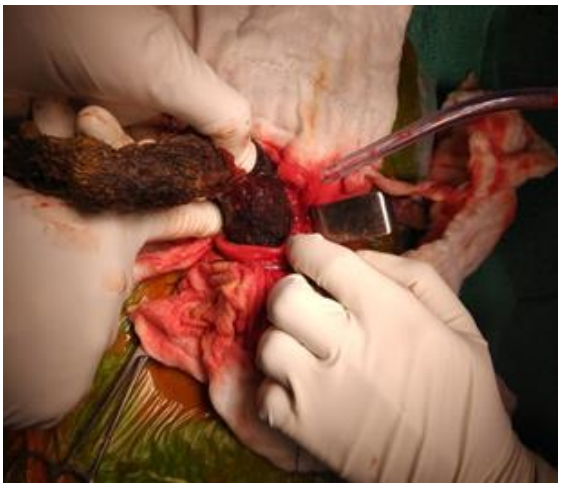

Figure 4a: Intra-operative retrieval of trichobezoar.

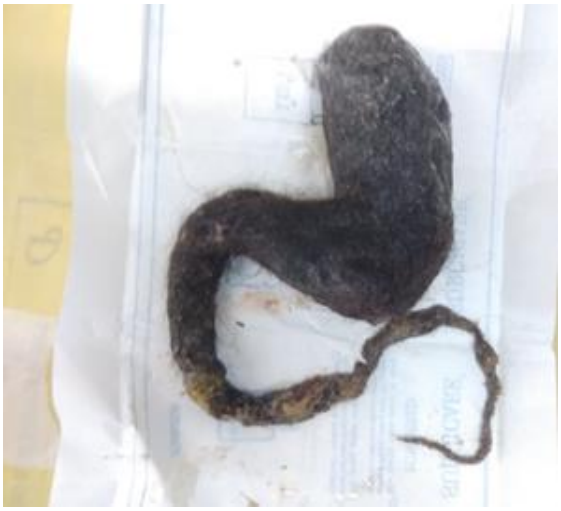

Figure 4 b: Trichobezoar.

CONCLUSION

RS, a gastric trichobezoar with intestinal extension is quite uncommon, and should be considered strongly in a young patient with abdominal pain, and non-tender abdominal mass with history of trichotillomania. Surgical removal is the gold standard treatment of choice. Psychiatric treatment prevents its recurrence.

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How to cite this article: Singh SK, Gupta AK, Parmar P, Chaudhary P, Lal R. Rapunzel Syndrome: A Case Report and Review of Literature. Ann. Int. Med. Den. Res. 2020; 6(3):SG01-SG04.

Source of Support: Nil, **Conflict of Interest:** None declared