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RAPUNZEL SYNDROME

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ABSTRACT
Rapunzel syndrome is a rare form of intestinal condition .The syndrome is name after a fairy tale princess who has incredibly long hair. It is an usual form of Bezoar which is extending from stomach to small intestine or beyond. Bezoar are concretion of human or vegetable fiber that accumulate in GIT. Trichobezoar (gastric hair ball) which a type of bezoar and it is common in psychiatric disorder where patient chew and swallow their own hair. People who compulsively chew or swallow their own hair are said to be have trichopagia. Such a cases are found in mentally retard or emotionally disturbed patients. Most of the cases of trichpbezoar are reported in females than males. The diagnosis of the syndrome is done by Contract- enhanced computed tomography(CT) imaging, Ultrasonography, contrast radiography, upper gastrointestinal endoscopy. The main symptoms of Rapunzel syndromes are epigastric pain and vomiting. Here in this review we have discuss some cases related to Rapunzel syndrome.

KEYWORDS: Bezoar, Trichopagia, Trichobezoar, Rapunzel syndrome, mentally retard.

INTRODUCTION
The word bezoar comes from Persian word “Padzhar” or Arabic word “Bedzer” which means ‘protecting against the poison’, i.e. antidote.[4][5] Bezoar can be divided according to their primary constituents into six types Trichobezoar (hair), phytobezoar (plant material), pharmacobezoar, lactobezoar, Fungal agglomeration and foreign bodies.[10][11]

The name Rapunzel syndrome inspired by fairy tale princess who has incredibly long hair.[8] the condition occurred when patient has hair ball (trichobezoar) in stomach, this hair ball has a tail that extend into small intestine or beyond.[1][2]

Trichobezoar can cause digestive problem such as abdominal pain, nausea, vomiting. trichobezoar are much common in females than males. in most of the cases syndrome observed in female aged less than 30 years.[1][4]

Trichobezoar is common type of bezoar in human which is mostly made of hair.[1] Trichobezoar associated with adultration of gastrointestinal motility but it underlying psychiatric disorder in mentally disturbed and mentally retard patient.[9]

In most cases trichobezoar reported in females though one report that male ate hair of his sisters. the majority of cases presented between 13-20 years of age. Then first report of Rapunzel syndrome was reported in 1968 by Dr. Vaughan.[1][6] after that 40 cases have been reported in the literature till date, within this 30% of these have been in India.

CASE REPORT
Case 1
An 11 year old female, she was referred with an abdominal mass associated with epigastric pain, vomiting after meal since 15 days. She had history of pica (habitual ingestion of foreign bodies) and habitual thumb sucking also she had no family or social strain.

On examination the patient was uncooperative and slightly dehydrated. Her body temperature, blood pressure and pulse was normal also chest and heart examination were normal. Her abdominal examination showed a firm mass in her epigastrium.

From the laboratory investigation it was clear that her WBC count, Hb, platelet count were normal and also there were no electrolyte disturbance and serum amylase, lipase and liver functions were normal.

Computed axial tomography showed a mass with a hazy outline, the mass showed no contrast enhancement and there were no contrast enhancement and there were air

pockets and that's why patient prepare for surgery. During surgery it was found that hair ball occupying the whole stomach and that had tail which is extending up to jejunum. That was extracted gently where gastric ulcer was also found.[16]

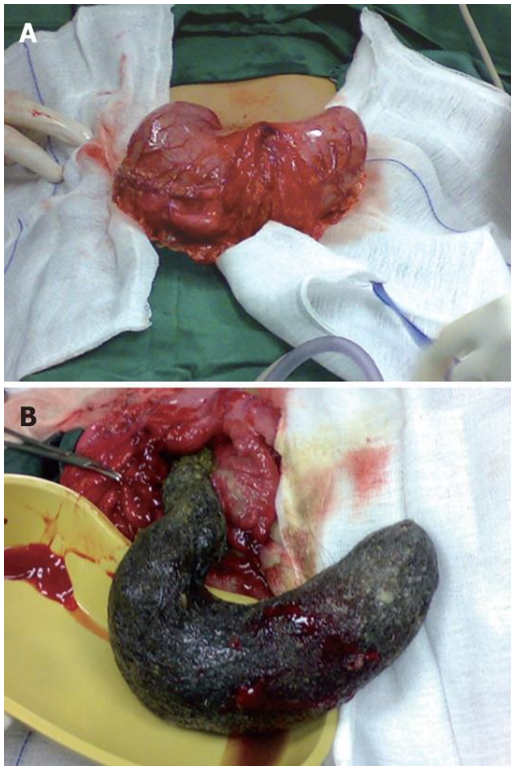

Fig 1: Hair ball occupying the whole stomach [A]and extending down to jejunum [B].

Fig 2: Full extend of the hair ball extending into jejunum.

Case 2

A 19 year old women who is pregnant for the first time, at the end of second trimester she suffered from upper abdominal pain, vomiting, constipation and abdominal enlargement for five days. There were psychiatric problem.

On examination her blood pressure, pulse and body temperature were normal also chest and heart examination were normal. Upper endoscopy disclosed the nature of mass as it was hair ball which occupying her whole stomach and then patient was posted for surgery. While doing operation it was found that hair ball was extending down to upper jejunum the gastronomy was closed in layers and then her abdominal closed with drainage.

The recovery period was very complicated by fetal loss and wound infection.[16]

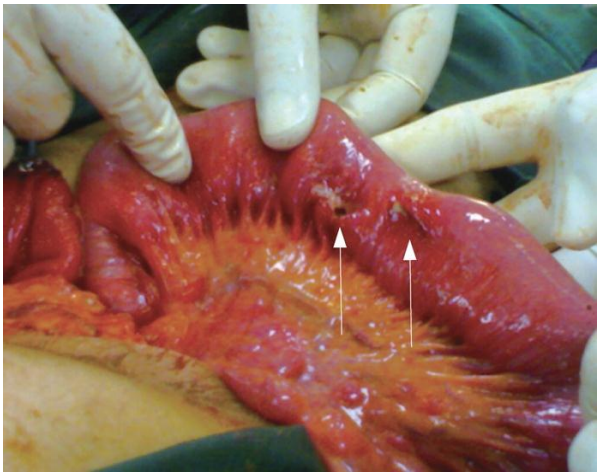

Fig 3: Two ruptured bowel at the mesenteric border of the proximal jejunum.

Case 3

A 10 year girl referred with epigastric pain and vomiting for 6 months in May 2001, on examination it was found that she was anaemic other than this WBC count, blood urea, blood sugar, serum creatinine, serum amylase, serum electrolytes and liver function were normal. chest radiography, urine analysis and ECG were normal. Barium meal study revealed that there was a large filling defect in stomach which was suggesting bezoar. Gastrotomy was performed and trichobezoar was removed and then patient was discharged.

After five year the same patient was again admitted with large abdominal mass with nausea, vomiting and loss of appetite, on physical examination Barium meal study and ultrasonography revealed that large mass in stomach which was extending up to jejunum then again gastronomy was performed and 62 cm mass of hair removed.[17]

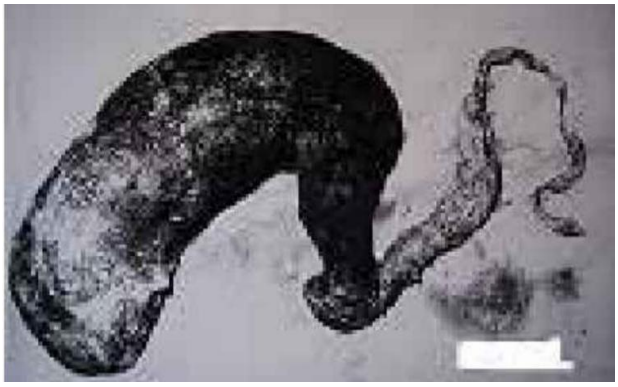

Fig 4: Photograph shows the trichobezoar extracted at the first laparotomy.

Fig 5: Photograph shows the trichobezoar extracted at the second laparotomy.

Case 4

A 7 year old female with a history of poorly localized abdominal pain. Her behavior was normal. Her mother commented on her self hair eating habit since 3 month but there were no baldness. There were also no changes in her bowel habit.

From the result of ultrasonography, Gastronomy was performed and the mass removed in only one piece. The patient was discharged and then she showed improvement in behavior.[18]

Fig 6: Trichobezoar found in stomach of 13cm× 7cm × 5cm.

DISCUSSION

Baudamant was the first who reported trichobezoar in 1779.[5][7] Trichobezoar is combination of two words 'trich' and 'bezoar'. In Greek trich means 'hair' and bezoar meaning 'antidote' in Arabic or Persian.[4][5] The bezoar formation usually because of ingestion of indigestible material. The symptoms of rapunzel syndrome are more common in female under the age of 30[3], and youngest case reported is of 6 month old infant.[5] There are six type of bezoar Trichobezoar (hair), phytobezoar (plant material), pharmacobezoar, lactobezoar, Fungal agglomeration and foreign bodies.[10][11]

The normally occurring symptoms are nausea, vomiting, abdominal pain, abdominal mass, loss of appetite, weight loss and aneamia.[12] If it left untreated for long period of time then it may cause ulceration, haemorrhage. other malabsorption related complications includes iron deficiency, megaloblastic anaemia.

In young girls with psychiatric disordered or mentally retard, trichobezoar is identified as secondary syndrome.[14] The actual reason why hair collect in stomach is not understood[6], Debakey and Oschner suggest that entrapment of hair in stomach is very irritating. Still psychiatric problem is usually present in such cases. Pregnancy complicate the situation and many cases that result into fetal loss and it is always the possibility.

According to classic series of Debakey et al upper abdominal mass remain common sign. The patient who get affected by this syndrome remain asymptomatic for many years, as the size of bezoar increases symptoms start develop that's the reason why many cases observed in that countries where women usually have long hair.[10][14]

The treatment aim to complete removal of trichobezoar from the stomach. Small size bezoar are removed by endoscopy. Almost in 97% cases Contract- enhanced computed tomography (CT) imaging is use to diagnose bezoar.[1][13] It use as detection tool. trichobezoar cover whole stomach, duodenum or jejunum that's why diagnostic tool such as, US, contrast radiography, upper gastrointestinal endoscopy are used to diagnose rapunzel syndrome.[1][4][13] Gastronomy is not indicated in all cases if patient presents intestinal obstruction it happen in most cases.[4] Large bezoar are difficult to remove in one session of endoscopy hence fragmentation is done and the it removed. There is no effective medical management for this, endoscopic removal is succeed in many cases due it's extension in small intestine. Surgical removal is the choice for removal of hair ball.[14][15] Now a days surgeons use laproscopic techniques for small bezoar.[5] Other methods that found successful are lithotripsy, intragastric administration of enzymes. Recurrence of Rapunzel syndrome is extremely rare, still to avoid recurrence parent counseling is very important.

CONCLUSION

Surgeons, radiologist and physician should consider Rapunzel syndrome in different diagnosis manner especially in young female. In suspicion of bezoar diagnosis should recognize as early as possible in order to get treatment. timely diagnosis and treatment is very important so that can prevent complications, morbidity and fatal outcomes. To avoid recurrence patient should have a proper psychiatric counseling and medications.

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