VOLUME-9, ISSUE-2, FEBRUARY-2020 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Medicine

RAPUNZEL SYNDROME IN A 15-YEAR-OLD INDIAN GIRL WITH AUTOIMMUNE HEPTITIS: A CASE REPORT AND LITERATURE REVIEW

Dr Priynka Ranga	Senior Resident Medicine MAMC Agroha, Hisar, Haryana.
Dr Pardaman Singh*	Assistant Professor , Radiology MAMC Agroha, Hisar, Haryana. *Corresponding Author
Dr R. K. Goel	Professor Medicine MAMC Agroha, Hisar, Haryana.
Dr Deepak Gehlan	Associate Professor Medicine MAMC Agroha, Hisar, Haryana.
Dr Kamal Mehta	Post Graduate Medicine MAMC Agroha, Hisar, Haryana.
Dr Parteek Tendon	Post Graduate Medicine MAMC Agroha, Hisar, Haryana.

ABSTRACT

INTRODUCTION: Bezoars are masses formed by the accumulation of intra-luminalnon-digestible substances that can lead to obstruction of the stomach and the small intestine. The anatomical changes in the gastrointestinal tract are known to cause bezoar formation. The so-called Rapunzel syndrome is the extension of the bezoars down to the duodenum and the jejunum, which is a rare condition. This may occur in subjects with mental retardation and/or psychiatric disorders.

CASE PRESENTATION: We present a 15-year-old female who was admitted to our department with history of pain in abdomen, loss of weight, recurrent vomiting,amenorrhea, easyfatigability for last 6 months. For which patient underwent investigations in which patient was havingclinical picture of malnutrition with normal level of LH,FSH,prolactin, thyroid profile and patient USG abdomen revealed coarse hepatic echotexture. Patient was Further found to be positive for ANTI LKM -1 (3 TIMES OF NORMAL VALVE) and diagnosis of autoimmune hepatitis was made and was put on immunosuppressant. CECT abdomen was ordered which reaveled a giant trichobezoarcompletely filling the stomach and duodenum causing partial obstruction ,needing surgical interference for removal and psychiatric therapy. We believe that this is a rare case presentation of Rapunzel syndrome in a 15 years old young female with Autoimmune liver disease and Trichotillomania . The link between Autoimmune liver diseases and neuropsychiatry disorders is still poorly understood and this case report may help us to understand the link between the two disorders.

CONCLUSIONS: Trichobezoars should be considered as a differential diagnosis in children complaining of recurrent abdominal pain,vomiting with epigastric mass and progressive weight loss. The link between autoimmune liver diseases and neuropsychiatry disorders is still poorly understood and this case may help us to understand the link between the two disorders. It is an original case report of interest to physician and psychiatry specialists.

KEYWORDS :

INTRODUCTION

A bezoar is an accumulation of food or foreign material in the intestinal tract. The term bezoar derives from the Arabic word Badzehr, which means antidote [1]. Bezoars were used as antidotes against many diseases like leprosy, snake bite, plague and epilepsy by physicians from 12th to 18th centuries [2]. It can be classified according to the primary constituent as trichobezoar (hairball), phytobezoar (food particles), trichphytobezoar (mixed), mycobezoar (fungal agglome rations), lactobezoar (milk component), or pseudobezoar[3]. Trichobezoars are the commonest type of bezoars and it is often associated with trichotillomania (hair pulling) and/or trichophagia (hair swallowing). Occasionally, trichobezoars have a tail that extends to the cardia, pylorus, and duodenum, or even further to the jejunum and ileum forming what is called Rapunzel syndrome[4].

CASE PRESENTATION

A 15-year-old female with a low socio-economic level admitted to our department with complaint of recurrent pain abdomen associated with vomiting intermittently, fatigue, facial puffiness,loss of weight, amenorrhea,sleep disturbance since last 6 month. There was recurrent dull aching pain mainly in abdomen not related to meals or physical activity. Her personal history revealed nothing significant except for the previous history of jaundice 3 years back. There was no past history of previous hospital admission, alcohol drinking by the family, drug intake. There was no history of blood transfusion and no prior history of early deaths, liver disease or autoim munity existed in the family. General examination revealed under built ,fully conscious, oriented, cooperative female with depressed mood. Pallor was present with puffiness rounds eyes but no jaundice,cyanosis, lymphadenopathy. Normal vital signs for age and sex were present. Abdominal palpation revealed a mobile well-defined mass occupying the upper half of the abdomen. The mass was not tender and was firm in consistency. The spleen was enlarged tip palpable but no hepatomegaly or ascites was present. Abdominal ultrasonography revealed coarsely echotextured of liver with spleen size around 13.5cm with normal portal pressure.

Extensive workshops of investigations were done to reach to the diagnosis of coarse echotexture of liver: A complete blood count revealed hypochromic microcytic anemia (Hb 8 gm/dL) with normal TLC (8.64/mm^3) and platelets count (335/mm^3). ESR (first hour 40) and CRP (30 mg/dL) levels were mildly elevated. The renal function tests were normal (BUN 16mg/dL&creatinine 0.7mg/). The liver function tests were normal bilirubin (total 0.81 mg/dL, direct 0.19 mg/dL), ALT (39 IU/L), AST (44 IU/L) levels. The laboratory tests excluded viral infections like (hepatitis virus B, C, HIV). Immunological testing for ANA, Anti-mitochondrial antibodies were negative. Anti-LKM-1 antibodies via ELISA method was positive (68.50 EU/ml, n<20).Total serumprotein(6.9) normal, low albumin(1.8), high globulin(5.1).IgG levels were slightly elevated(220 mg/dl,n=200mg/dl). Serum calcium was low(7.9mg/dl).LH, FSH, prolactin and thyroid profile were in normal range.

Patient was put on steroid and azathiopurine treatment for one

month with tapering dose.All parameters wereimproving but patient was still having vomiting and pain in abdomen.Patient then underwent for CECT abdomen that suggestive oflarge heterogeneous mass with mottled gas pattern in stomach extending to pyloric region, duodenum, jejunum, proximal ileum with mild mucosal thickening suggestive of RUPANZAL SYNDROME.

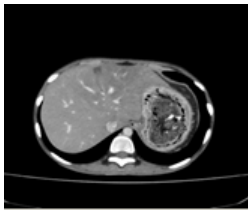

Figure 1

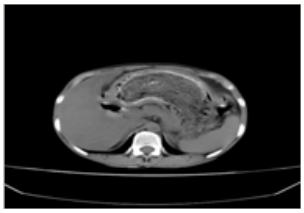

Figure 2

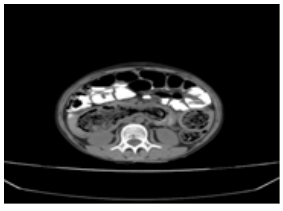

Figure 3

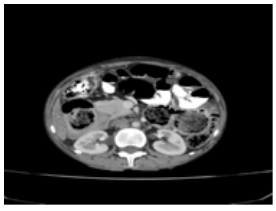

Figure 4

Fig 1 to 4:-**Serial axial post contrast CT images of upper abdomen shows Huge, well defined, multi-layered, heterogeneous, solid appearing, non-enhancing mass in the gastric lumen extending from the gastric fundus to the pyloric canal down to jejunum. The mass lesion is separated from the gastric wall by contrast in stomach .**

After the initial medical management patient was stabilized. Endoscopic removal of the mass was not possible hence surgical intervention was planned.

DISCUSSION

Rapunzel Syndrome is a form of trichobezoar. It is named after a tale written in 1812 by the Brothers Grimm about a young maiden, Rapunzel, with long hair who lowered her hair to the ground from a castle, which was a prison tower to permit her young prince to climb up to her window and rescue her. It was first described by Vaughan et al in 1968[5]. Various criteria have been used by different clinicians to report their cases as Rapunzel syndrome. Some have defined it as a gastric trichobezoar with a tail extending up to the ileocecal junction[6], others have said it is simply a trichobezoar with a long tail, which may extend to the jejunum, ileum or the ileocecal junction, and still others have defined it as a trichobezoar of any size which presents in the form of an intestinal obstruction[7] . However the most accepted definition is that of a gastric trichobezoar with a tail extending to the small bowel. Various types of bezoar are described depending on their content. Trichobezoar - made up of hairs, Lactobezoar- made up of milk curd, Phytobezoar - made up of vegetable or fruit fibre, Trichophytobezoar- mixed, pharmacobezoars (drugs) or diospyrobezoars (persimmon fibers). Cotton bezoars are also rare, resulting from the ingestion of strings unravelled from clothes, especially by mentally retarded patients[8,9] . Trichobezoar are formed as the ingested hairs get stranded and are retained in the folds of the gastric mucosa. Their slippery surface prevents their propulsion by peristalsis. The initial hold up by the pylorus helps entangle new hairs. As more and more hairs are added, peristalsis causes them to be enmeshed like a ball. The ball grows in size, assumes the shape of the stomach and can lead to gastric atony if size is too long. It has a glistening shiny surface due to the mucous covering it. The putrid smell is due to decomposition and fermentation of fats in the interstices. The mean age of presentation ranges from 1 yr 2 months to 22 years with 68.9% cases in the age group of 5- 15 yr. Rapunzel syndrome is mainly reported in females (>95% of cases), however 2 males have also been reported[9] , which may be attributed to the traditional long hairs in females[10] . Symptoms of this disease are very vague and varied; the most common presenting symptoms are abdominal pain (46.66%), nausea and vomiting (44.44%), obstruction (20%), abdominal distension (8.88%), weight loss (8.88%), peritonitis (6.66%), abdominal mass (6.66%), sepsis (2.2%), cardiac arrest (2.2%) and unusual pallor (2.2%). Due to non specific symptoms and low index of suspicion by the physician majority of cases present late. Mortality has been reported in 4 out of 45 cases (8.9%). Mortality is more in younger age and it is because of complications. There are co-existing factors usually like mental retardation, psychological problems, family problem/parental discontent and bereavement[11]. Trichobezoars are nearly impossible to diagnose by plain X ray alone. Ultrasound has been shown to be effective in diagnosing bezoars in up to 88% of cases if a "clean" acoustic shadow, which represents a solid mass, can be visualized. However, a calcified mass, neuroblastoma, aneurysm, abscess, or fecal material can appear similar on an ultrasound[12]. CT abdomen with contrast has been shown to diagnose 97% of bezoars which appear as free-floating filling defects in the stomach[8]. On CT scan, a well-circumscribed lesion, composed of concentric whorls of different densities with pockets of air enmeshed within it, appears in the region of stomach. Absence of significant post intravenous contrast enhancement precludes a neoplastic lesion. Endoscopy is diagnostic in almost all cases. Clinically epigastric mass must be differentiated from a pancreatic pseudocyst, a neuroblastoma, a horseshoe- shaped kidney, an enlarged spleen, a gastric neoplasm, an enlarged liver and a Wilms' tumour. No effective medical management is available. Endoscopic removal is reserved for small Trichobezoars only. Endoscopic removal is unlikely to succeed in Rapunzel syndrome due to large size and extension in small intestine. Surgical removal is the treatment of choice in Rapunzel syndrome and is accomplished by gastrotomy&/or enterotomy. Since the advent of minimally invasive surgery, surgeons now use laparoscopic techniques for small to moderate-size bezoars only. Various other methods like extracorporeal shock wave lithotripsy, intragastric admini stration of enzymes (pancreatic lipase, cellulose) demonstrate varying success. The complications of Rapunzel syndrome include attacks of incomplete pyloric obstruction, complete obstruction of the bowel, perforation, peritonitis and death. Because of the inequality in the length of bowel and mesentery, there is unequal pressure along the mesenteric border of the bowel. As the bowel becomes more compressed, pressure necrosis may develop along the mesenteric aspect of the bowel wall at points of maximum tension. The other complications include internal fistulae, obstructive jaundice, megaloblasticanemia, protein-losing enteropathy and short bowel syndrome.

CONCLUSION :

Surgeons, physicians, and radiologists should consider Trichobezoarin the differential diagnosis of gastrointestinal obstruction in young females, especially in the presence of an upper abdominal mass. Timely diagnosis and treatment can prevent the morbidity, complications and fatal outcome. As recurrences are known, each patient should have a proper psychiatric evaluation and follow-up

REFERENCES

1. Samad A, Ahmad M, Latif Z (1997) Bezoars: a review and report of two cases. Journal of the College of Physicians and Surgeons Pakistan 7: 263-265.
2. Khattak S and Asghar K (2004) Trichobezoars. Gomal Journal of Medical Sciences2: 25-26.
3. MewaKinoo S, Singh B (2012) Gastric trichobezoar: an enduring intrigue. Case Rep Gastrointest Med 2012: 136963.
4. Rabie ME, Arishi AR, Khan A, Ageely H, Seif El-Nasr GA, et al. (2008) Rapunzel syndrome: the unsuspected culprit. World J Gastroenterol 14: 1141-1143.

5. Vaughan ED, Sawyers JL, Scott HW. The Rapunzel syndrome.An unusual complication of intestinal bezoar. Surgery 1968; 63(2): 339-343.
6. Kaspar A, Deeg KH, Schmidt K, Meister R. Rapunzel Syndrome, a rare form of intestinal Trichobezoar. KlinPeadiatr 1999; 211(5):420-422.
7. Faria AP, Silva IZ, Santos A, Avilla SGA, Silveria AE. The Rapunzel syndrome - a case report :trichobezoar as a cause of intestinal perforation (in Portuguese). J Pediatria (Rio J) 2000; 76(1):83-86.
8. Sarin YK. Rapunzel Syndrome. Indian Paediatrics 1998; 35: 682-683.
9. Emre AU, Tascilar O, Karadeniz G, Irkorucu O, Karakaya K, and Comert M. Rapunzel Syndrome of a Cotton Bezoar in a Multimorbid Patient. Clinics. 2008; 63(2): 285-288.
10. Gonuguntla V, Joshi DD. Rapunzel Syndrome: A Comprehensive Review of an Unusual Case of Trichobezoar. Clin Med Res. 2009; 7(3): 99-102
11. Duncan ND, Aitken R, Venugopal S, West WM, Carpenter RA. The Rapunzel syndrome.Report of a case and review of the literature.West Indian Med J 1994; 43:63 65.
12. Frey AS, McKee M, King RA, Martin A. Hair Apparent: Rapunzel Syndrome. Am J Psychiatry 2005; 162(2): 242-248.