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Trichobezoar with long hairy tail, not fairy tale (Rapunzel syndrom)

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Abstract: *Bezoar is an uncommon condition which occur by ingestion of indigestible material like hair(trichobazoar) furr fruit and some vegetable(phytobazoar). most commonly found is trichobazoar. from the accumulation of*
Such material in intestinal track lead to formation of a large mass of bazoar. The "bezoar" is a Persian word means "protection from poison. The first bezoar in human was in 1779 during an autopsy of a patient who died from sepsis due to peritonitis after complication of bezoar due to gastric perforation.[1] Trichotillomania (hair pulling) was first described in literature in 1889.in Trichotillomaniacs only 30% eat their hair and in these trichophagia patient only 1% eat excessively to that point to form trichobazoar in their .[2] trichobazoar occur mainly in young females with psychiatric disorder, one who pull out one's own hair (Trichotillomania) and eat it (trichophagia). Other psychiatric disorders, such as mental disorders, obsessive compulsive disorder, abuse, depression and anorexia nervosa may also be associated with trichobezoar.[3]

Key Words: *Rapunzel syndrome,trichobazoar,developmental delay.epigastric lump*

I. Introduction

Rapunzel Syndrome is a rare form of trichobazoar. It's name given after a tale written in 1812 by the Brothers Grimm she was a young maiden, Rapunzel, who had very long hair she lowered her hair to the ground from the top of castle, which was actually a prison with no stairs only witch can fly and reach on the top, permit her young prince to climb up to her window and rescue her.

Trichobezoars, commonly occur in females with psychiatric disorder who chew and swallow their own hair. But Only 50% patient give history of trichophagia.Trichobezoars most common type of bazoar mention in literature and they form 55% of all bezoars. In very rare cases of trichobazoar the tail of mass formed by hair extends through the pylorus beyond the duodeno-jejunum junction into the small bowel called Rapunzel Syndrome.in this condition mass causes symptom and sign of partial or complete gastric outlet obstruction.[4,5]

II. Case Report

A 13 year old female patient presented in pediatric opd with chief complain of pain upper abdomen since 2 month decrease oral intake 1 month regurgitation 15 days no h/o vomiting and hematmesis

On examination there was a lump firm to hard in consistency 15 to 20 cm size involving the left hypochondrium epigatrium and extending into right hypochondrium it broad from left and upper side and gradually narrow lower and right side. so basically it took the shape of stomach. so the suspicion of trichobazoar came in seen asking about any psychiatric illness there was a negative history
But patient had delay of all developmental mile stone.
Sitting-on 2 year, normal child start sitting without support on 9 month
Standing 2 and a half year, normal child start standing 0n 18 moth

Walking and talking on 3 year, normal child able to speak two to three word at the age of 2 year and walks on age of 18 month

So there is delay of joining in school now at the age of 13 year patient reading in 2nd standard so patient have low i.q. level usg also give a suspicion of intaluminal pathology so we went for upper g.i. endoscopy and it came to be trichobazoar in the stomach going toward duodenum so decision of laparotomy is taken .

Abdomen is opened from a upper abdominal transverse incision then stomach was mobilized and a transverse incision given on its anterior surface slowly n carefully whole of the lump (trichobazoar)and its continuation into the duodenum and jejunum is takenout as a whole there was a long tale about 60 cm long and the size of mass in stomach was 16 *10 cm size. after removing the hair ball with its tail stomach is closed in two layers and abdomen is closed in layers.post –op course was un eventful. Patient is discharged with advise to follow up with pediatric surgery and psychiatric opd

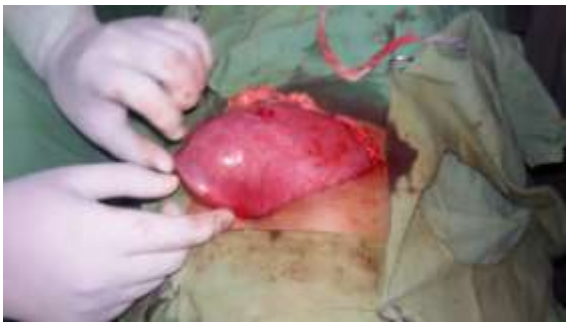

Figure 1 –mobilise the stomach from laparotomy incision

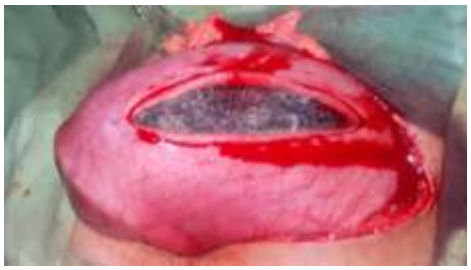

Figure 2&3- shows trichobazoar in luman

Figure 4-large trichobazoar removed from lumen

Figure 5-whole trichobazoar specimen

Figure 6-60 cm long tail

III. Discussion

More than 90% of patient of trichobazoar is female and of young age group mainly less than 30 year and suffer from a psychiatric illness or has a stressor event in life or pica syndrome or developmental delay[6]

Pathogenesis of trichobazoar not very studied but it is said that smooth surface of hair and rougal fold prevent it from propelling action of peristalsis .more and more hair accumulates and get meshed up to form ball in stomach narrowing ar pylorus and bend at duodenum make trichobazoar to form a ball in stomach and this take a shape of trichobazoar[7].

Decomposition and fermentation of trapped Food in hair ball gives the bezoar, and the patient's breath, a foul and putrid smell. Rapunzel syndrome is a rare condition in which hairy tail extend beyond duodeno-jejunal junction [8] Trichobazoar present with the chief complain of a palpable abdominal mass in (87.7%) present , abdominal pain (70.2%), nausea and vomiting (64.9%), weight loss (38.1%), constipation or diarrhea (32%) and hematmesis (6.1%). The laboratory investigations show low hemoglobin in about 62%[9].

Patient of trichobazoar are usually pallor (Iron deficiency anaemia present in

many cases). it may be due to impaired gastrointestinal absorption ,due to

pica syndrome, erosions and ulceration and malena occurred due to Pressure necrosis. Due to obstruction at the ampulla of vater at 2nd part of duodenum Jaundice and acute pancreatitis may accured[10,11] clinical suspicion may occurred after careful history taking and clinical examination but to diagnose we should go for imaging modality. hyperechoic curvilinear dense strip at the anterior margin of the lesion associated with marked acoustic shadowing [12] .on barium it gives mottled filling defect it gives positive enhancement of mass like lacelike pattern because of residual contrast medium clearly seen on delayed films

on fluoroscopic examination in erect position swallowed barium held up by the mass and in few sec this trickle down defuse slowly downward make the contour of mass prominent

On CT, with the characteristic appearance as described above, CT best reveals the size and shape of themass and most precisely identifies its location. Further, on CT appearance we can differentiate it from other pathologies (such as neoplasm)[13] .

We can diagnose the trichobazoar on imaging but for confirmation we have to go for upper g.i. endoscopy to see the hair ball in stomach it can be therapeutic for small volume trichobazoar, The first report of successful endoscopic removal of trichobazoar In relatively small one, only 55 g [14].endoscopic lithotripter and laser fragmentation are emerging now adays for minimal invasive surgery for trichobazoars.they give two advantages: first, they can remove small trichobezoars and remove small but in continuation trichobazoars which extent into duodenum and jejunum but for large trichobazoars erosion and perforation are frequent by endoscopic removal[15].

Laparoscopic removal of bezoar was first reported by Nirasawa 1998[16] for the removal of giant trichobezoar, the laparoscopic approach could be troblsome .other alternative is Minilaparotomy to laparoscopy in cases of moderately large trichobezoars [17]. In a study by Koulas et al. including over 23 cases, the morbidity was 28% (wound infection, incisional hernia), whereas the endoscopic morbidity was 11%. Mortality was 4% and for the surgical and endoscopic groups it was 0%. An open approach is safest for removal of giant one. while also avoiding contamination of the peritoneal cavity and wound with gut content and hair.

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