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Rapunzel Syndrome: A Rare Presentation with Multiple Small Intestinal Bands and Perforation

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Abstract: ***Introduction*: Rapunzel syndrome was 1st reported in literature by Vaughan et al in 1968.Rapunzel Syndrome is an extremely rare intestinal condition resulting from ingestion of hair (trichophagia) extending from stomach to small intestine. *Case Report*: We present a case report of 16 year old female with complain of pain abdomen,vomiting and epigastric mass. UGI Endoscopy suggestive of Gastric Trichobezoars.Patient underwent laparotomy gaint trichobejoar with jejunal perforation for which jejunojejunal anastomosis done. Patient discharged with full recovery. *Discussion*: Rapunzel syndrome occurs predominantly in young women with psychiatry disorder and consist of presence of rare type oftrichobejoar.Trichotillomania and trichophagia were only reported after noticing bald patches on scalp resulting from pulling out and swallowing of hairs.CECT abdomen reveals extend of trichobezoar and gold standard is upper GI endoscopy.Treatment of trichobezoar includeremoval by endoscopy,gastrotomy or enterotomy and laparoscopic etc. *Conclusion*: CECT and endoscopy is investigation of choice Laparotomy is considered excellent treatment option. Psychiatric assessment and long term follow up advised as a regular part of treatment to prevent recurrence.***

Keywords: Rapunzel syndrome, Trichobezoars, Laparotomy, Upper GI endoscopy, CECT abdomen

1. Introduction

Rapunzel syndrome was 1st reported in literature by Vaughan et al in 1968. Bezoars have been known to occur in the form of undigested masses found in GI tract. The masses classified as follows:- phytobezoar-plants, lactobezoars-milk, trichobezoar- hairs and pharmacobezoars composed of medicines. Rapunzel syndrome represents trichobezoar with a tail extending in small intestine. Patient presenting with pain abdomen, vomiting, non-tender abdominal masses should undergo CECT, endoscopy. In terms of treatment multiple options were reported, among which medical treatment and enzymatic degradation, endoscopic, laparoscopic or laparotomic removal.

2. Case Report

A 16 year old girl presented with complain of pain upper abdomen, vomiting andabdominal mass in epigastric region frompast one month. Pain abdomen was acute in onset, non-progressive, non-radiating and not relieved by medication. Vomiting was multiple, non- projectile non-bilious in nature.There was no past illness or any psychiatric illness and was not on any medications.On examination patient was pallor and hypotensive (90/60mmhg).On abdominal palpation revealed a hard, non-tender,mobile lump of size 10x5 cm in left upper hypochondrium. There was shift of frontal hair line.Lab investigation Hb-9.3g/dl, TLC-4.13*10^3/ul, PLT-162*10^3/UL, LFT-WNL, RFT-WNL, SE-133/3.11, RBS-98.

Radiographic investigations

X ray FPA	Distended Reverse J shaped stomach at the level of fundus in right side (figure 1)
USG whole abdomen	Echogenic mass with intense acoustic shadow seen in stomach
CECT abdomen pelvis	Stomach distended,with defined mixed density predominant hyperdense rounded material with internal multiple air foci size 5.3x8.2x10cm s/o ?Bejoar (FIGURE2)
UGI ENDOSCOPY	Gastric trichobezoar

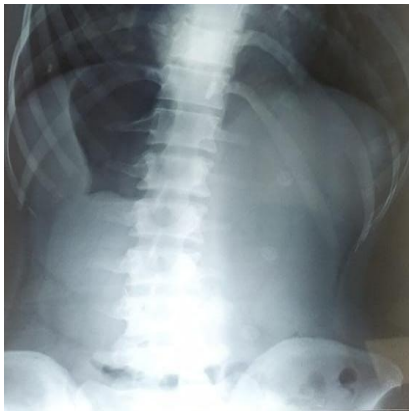

Figure 1

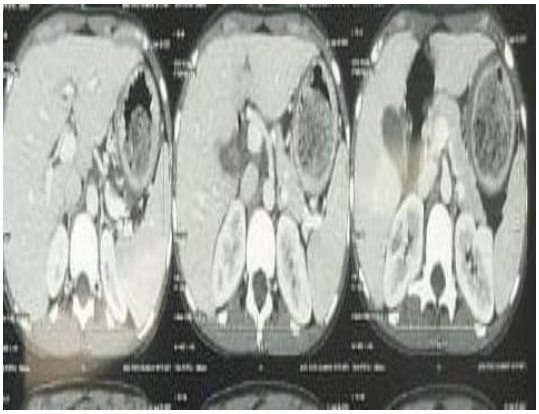

Figure 2

Psychiatric refernce done patient and relatines gave negative history for hair pulling and ingestion.After taking written informed conset and PAC fitness .Patient was planned for laparotomy. Patient underwent surgery through classical midline vertical incision given gastrotomy done gaint trichobejoar (figure 3) of of 6 cm diameter and 20 cm in length identified and removed which was extending upto Jejunum and was forming band insideuneven ly, over which whole jejunum was overlapping .There was a fibrous band which was 50 cm from DJ over the anti-mesenteric border of jejunum and was impending to perforate with compromised blood supply and there w as a another band over jejunum with 2*1 cm(figure 5) of perforation which was 70 cm from DJ for which jejunojejunal anastomosis was done.Patient started orally on day 5.Post operative period uneventful drain was removed on day 5. Patient discharged on POD-9 with taking orally and passing flatus motion on dischange patient advised to attend psychiatry opd to prevent recurrence

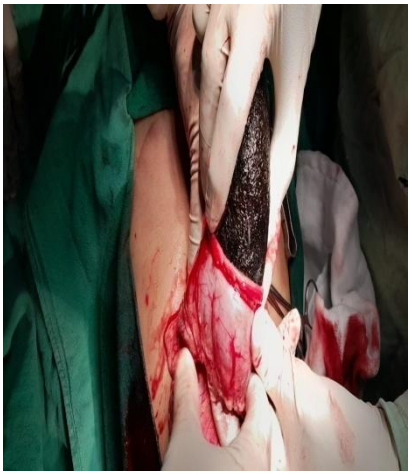

Figure 3

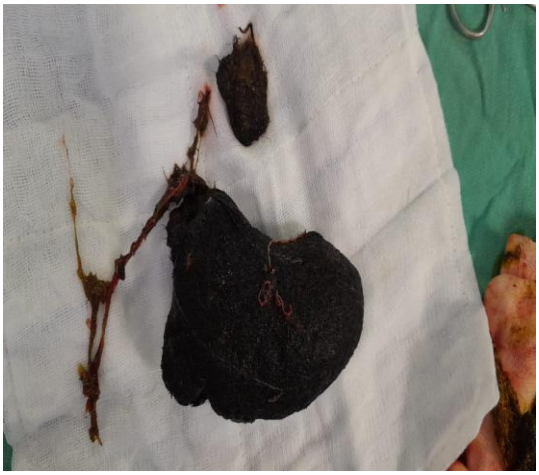

Figure 4

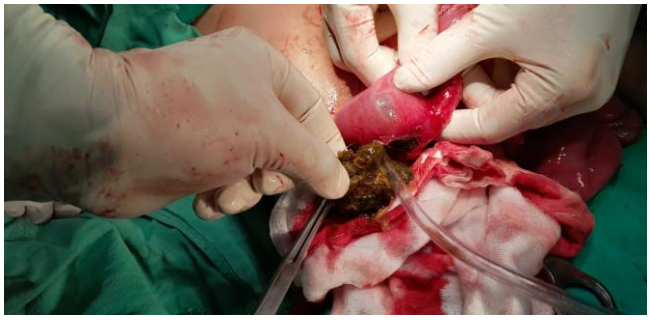

Figure 5

3. Discussion

It has already been confirmed in literature that rapunzel syndrome occurs predominantly in young women with psychiatry disorder and consist of presence of rare type oftrichobejoar.Trichotillomania and trichophagia were only reported after noticing bald patches on scalp resulting from pulling out and swallowing of hairs.Trichobezoar form when hair strands escaping peristaltic propulsionand retained in folds of gastric mucosa.As more hair accumulates,peristalsis causes it to be enmeshed into ball.Ball gets larger and causes gastric atony.Hair ball become more matted together and assume stomach shape or a single solid mass.Patint usually present with abodmen pain,vomiting and abdominal lump. Obstruction is most common clinical manifestation. CECT abdomen reveals extend of trichobezoar in stomach and

small intenstine and gold standard is upper GI endoscopy.Treatment of trichobezoar include use of chemical substances to dissolve hair ball and mechanical fragmentation or removal by endoscopy.However gastrotomy or enterotomy is advised in larger trichobezoar.When bowel is damaged showing necrosis, perforation which has been described in this case bowel resection and anastomosis have to be performed. Now a days use of laparoscopic techniques, extracorporeal shock wave lithotripsy,intragastric enzymes,medical administration seen for small to moderate size bezoars but due to small sample size these techniques in treatment of rapunzel syndrome remains to be defined. To decrease recurrence long term psychiatric follow up is advised.

4. Conclusion

Rapunzel syndrome is an uncommon trichobezoar commonly found in young female. Common presentation include pain abdomen, vomiting ,abdominal mass. CECT and endoscopy is investigation of choice Laparotomy is considered excellent treatment option. Psychiatric assessment and long term follow up advised as a regular part of treatment to prevent recurrence.

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