



## IRON DEFICIENCY ANEMIA AND HYPOALBUMINEMIA SECONDARY TO A RARE CASE OF RAPUNZEL SYNDROME.

### Biochemistry

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### ABSTRACT

**Background:** The term "RAPUNZEL SYNDROME" was coined by Vaughan and associates in 1968 for unusual manifestation of a trichobezoar in which the mass extends from the stomach and duodenum through a large portion of small intestine. Bezoars are collection of non-digestible foreign bodies such as hair that usually accumulate in gastrointestinal tract. Trichobezoar is underdiagnosed entity that has to be considered in childrens and adolescents especially females, suffering from trichotillomania (compulsion to pull hair) and trichophagia (compulsion to swallow hair). When undiagnosed, gastric bezoars may also cause severe anemia either due to malabsorption or gastrointestinal bleeding. **Objectives:** 1) To emphasize the presentation and focus on prompt diagnostic findings in a case of Rapunzel syndrome. 2) To explain the need of routine biochemical investigations like S. Iron, S. Ferritin, S. Albumin, S. Globulin, Total proteins in case of Rapunzel syndrome so as prevent further complications. **Methodology:** A 17 year old female presented to casualty with complaints of pain in epigastric region since 4 months which was spasmodic on and off in nature and non-radiating. History of nausea and vomitings and history of strange behaviour noticed by guardians-plucking hair and swallowing. USG suggested hyperechoic curvilinear dense strip with acoustic shadowing. CT scan shows an intragastric well circumscribed inhomogenous mass consisting of mottled gas pattern. **Result:** patient was diagnosed with Rapunzel syndrome associated with iron deficiency anemia and hypoalbuminemia. **Conclusion:** Rapunzel syndrome is a rare entity that can be complicated with life threatening events. A prompt diagnosis of trichobezoar so as to prevent deficiencies which may lead to complications.

### KEYWORDS

#### INTRODUCTION:

Rapunzel syndrome is a rare and unusual form of trichobezoar. Since 1968, there have been only 64 documented cases in the literature. (1) The term "RAPUNZEL SYNDROME" was coined by Vaughan and associates in 1968, for an unusual manifestation of trichobezoar in which the mass extends from the stomach and duodenum through a large portion of small intestine. Bezoars are the collection of non-digestible foreign bodies such as hair that usually accumulate in gastrointestinal tract (2). Trichobezoar is an under diagnosed entity that has to be considered in childrens and adolescents especially females, suffering from trichotillomania (compulsion to pull hair) and trichophagia (compulsion to swallow hair). This condition is commonly observed in adolescents, particularly in young females with trichotillomania and trichophagia (5,6). Bezoars mostly originate in the stomach, probably due to its capacity and its related to abnormal diet pattern which may result in malnutrition and iron deficiencies. About one-third of trichobezoar patients, in turn, develops Rapunzel syndrome, a condition in which the body of the hairball lies in the stomach, and its tail extends to the duodenum, ileum or all the way to the colon (9). When undiagnosed, gastric bezoars may also cause severe anemia either due to malabsorption or gastrointestinal bleeding (3).

As trichobezoar enlarges, more serious complications can develop such as malabsorption, protein losing enteropathy, nutritional deficiencies including iron deficiency anemia (4). Rarely, gastric bleeding and gastrointestinal obstruction may be observed as complications of large trichobezoar (5,6,7,8).

#### CASE PRESENTATION:

17yr old female came to casualty with complaints of pain abdomen in epigastric region since 4 months which was spasmodic on and off in nature, non radiating. History of vomiting and nausea which was non bilious, non-projectile not associate with blood. History of strange behavior noticed by guardians-plucking hair and swallowing since 6 months. Menstrual history – menarche -14yrs, irregular cycles (once in 3 months), 2-3 days flow.

On examination :GSC-13/15

Pallor present, moderately built.

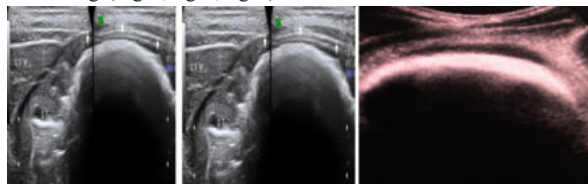
Local examination- Afebrile on touch, presence of hard mass noticed at epigastric region felt was not moving on respiration and borders were ill defined. Diagnosis is usually made by findings based on the clinical grounds and by laboratory and radiological findings like barium swallow and CT scan. Similarly ultrasound can be helpful

which shows echogenicity in the stomach (10). Management classically involves surgical removal of the hairball, adequate diet and psychological management of the underlying cause (5).

Patient was send for further Biochemical evaluation, ultrasound and CT was done.

#### USG findings:

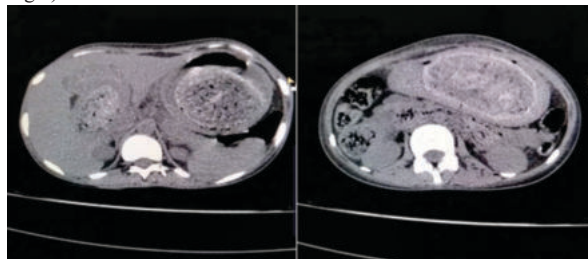
A large complex mass was noted in upper abdomen, seen as a huge echodensity in the region of stomach. The mass was large and was casting a huge semilunar or crescentic echodensity with distal shadowing. (Fig. 1, Fig. 2, Fig. 3)



**Fig.1** **Fig.2** **Fig.3**  
(Fig. 1, Fig. 2, Fig. 3) USG showing large complex mass and was casting a huge semilunar or crescentic echodensity with distal shadowing.

#### CTSCAN:

Shows an intragastric well-circumscribed inhomogenous mass consisting of 'mottled gas pattern' or 'compressed concentric rings' pattern due to the presence of entrapped air and food debris. (Fig. 4 ; Fig. 5)



**Fig.4** **Fig.5**  
(Fig. 4, Fig. 5) Shows an intragastric well-circumscribed in homogenous mass consisting of 'mottled gas pattern' or 'compressed concentric rings' pattern.

The Biochemical findings are given in Table 1

| Parameters              | Results                 |
|-------------------------|-------------------------|
| Haemoglobin             | 7 g/dl (12.0-15.0 g/dl) |
| Serum.Iron              | 33 mu. g/dl (50-170)    |
| Serum.Ferritin          | 23 ng/ml(30-400)        |
| TIBC                    | 654 mu. g/dl(215-535)   |
| %Transferrin saturation | 5.04%(13-45)            |
| Albumin                 | 1.5 g/dl (3.4- 5.4)     |
| Globulin                | 2.81 g/dl (2.5-3.4)     |
| S. AL/GL Ratio          | 0.54 (0.9-2)            |
| Total proteins          | 6.59 gm/dl (5.7-8.2)    |
| Vit.B12                 | 380 pg/dl (160- 950)    |

### Case Discussion

Patient was diagnosed with Rapunzel syndrome associated with Iron deficiency anemia and Hypo albuminemia. The treatment of large bezoars and concretions is essentially surgical via gastrotomy(Fig. 6). It is mandatory to do a thorough exploration of the rest of the small intestine and the stomach to look for retained bezoars. After appropriate transfusion of erythrocytes and plasma about 3 units and one pack of albumin, patient underwent surgical intervention by gastrotomy.



**Fig. 6**

**Fig. 7**

(Fig .6, Fig.7) Showing trichobezoar after surgical intervention by gastrotomy.

Gradual supplementation of iron and micro nutrients allow adequate catch up with normalization of biochemical nutritional parameters. After surgery and gradual re-feeding, normalization of biochemical and nutritional parameters is observed in most of patients (5,6,8).

Follow up visit - As advised the patient had a followup visit with repeat of biochemical parameters. (Table. 2)

Table. 2

| Parameters              | Results                  |
|-------------------------|--------------------------|
| Haemoglobin             | 11 g/dl (12.0-15.0 g/dl) |
| Serum.Iron              | 70 mu. g/dl (50-170)     |
| Serum.Ferritin          | 40 ng/ml(30-400)         |
| TIBC                    | 320 mu. g/dl(215-535)    |
| %Transferrin saturation | 21.8%(13-45)             |
| Albumin                 | 3.2 g/dl (3.4- 5.4)      |
| Globulin                | 3.1 g/dl (2.5-3.4)       |
| S. AL/GL Ratio          | 1.03 (0.9-2)             |

### CONCLUSION :

Iron deficiency anemia and hypoalbuminemia is seen as a complication of gastric trichobezoar formation. The Rapunzel syndrome is a rare entity that may be complicated by life – threatening events. Patient was recommended further supplementation of iron, Vitamin B complex, protein supplementation and other micronutrients. Prophylaxis against recurrence should be by proper psychiatric counselling and monitoring A Prompt diagnosis of trichobezoar so as to prevent deficiencies which may lead to complications.

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