A Rare Case of Rapunzel Syndrome.

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Abstract:

Rapunzel syndrome is a rare presentation occuring predominantly in young women or girls with psychiatric or emotional disorders and consists of trichobezoar with a tail extending into small bowel with symptoms suggestive of obstruction. With less than 40 cases reported in literature till date we report a case in a young female diagnosed clinico-radiologically and treated surgically with removal of an intact trichobezoar extending from stomach till the duodenum.

Key-words: Rapunzel syndrome, trichobezoar.

Introduction:

Bezoars consist of nondigestible foreign material present in the stomach or intestine and are categorized by content into four categories. Phytobezoars originate from vegetable and plant matter, trichobezoars result from the ingestion of hair, lactobezoars, which consist of milk curd and pharmacobezoars or medication bezoars.

Rapunzel Syndrome is an uncommon presentation of trichobezoar, involving strands of swallowed hair extending as a tail through the small intestine, beyond the stomach. This was first described in 1968 by Vaughan et al[1], with less than 40 cases reported till date. It is predominantly found in emotionally disturbed or mentally retarded youngsters with history of trichotillomania and trichophagia[2], We report a case in a young girl with psychoneuronal disorder presenting with complaints of vomiting, abdominal pain and progressive weight loss.

Case Report:

A 13 year old girl presented to us with complaints of persistent vomiting, abdominal pain and progressive weight loss since 6 months. On examination the patient was pale and cachexic in appearance. Her hemoglobin was 8 gm/dl with microcytic hypochromic picture on peripheral blood smear. Other routine blood examinations were within normal limits.

Barium Meal examination revealed filling defect in stomach with persistent barium within the interstices of the filling defect seen extending into the duodenum.

Based on this and clinical features possibility of Rapunzel syndrome was kept and on further questioning the patient's mother admitted that she was on antipsychotic medications and had history of psychoneuronal disorder with a habit of pulling out

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her hair and secretly swallowing it.

CECT was the next investigation advised. The findings were:

- An intragastric non enhancing well-circumscribed inhomogenous mass consisting of 'mottled gas pattern' pattern due to the presence of entrapped air and food debris, the tail extended into duodenum
- Normal stomach wall was traced completely separate from the lesion
- Mucosal edema and wall thickening was seen in duodenum

Management:

Patient was taken for elective operative procedure in which a hard lump was felt in stomach which was moveable inside. Gastrotomy was done and the mass was extracted with its extension present in the duodenum. Patient was discharged on post operative day 8 without any postoperative complications.

Image 1: Barium meal examination

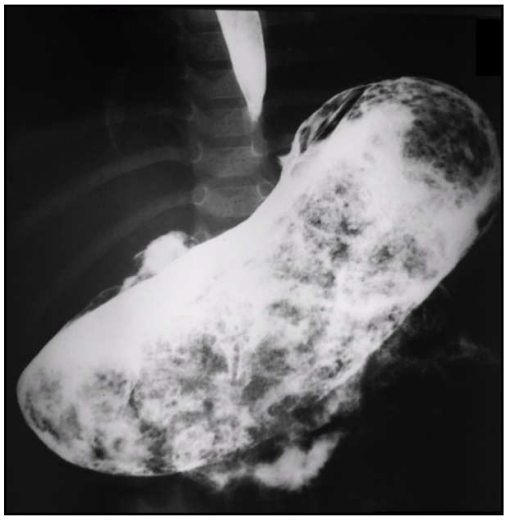


Image 2: Intraoperative specimen of gastric trichobezoar with duodenal tail

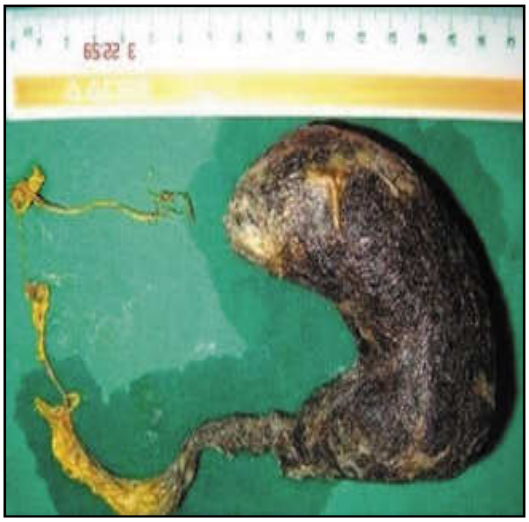
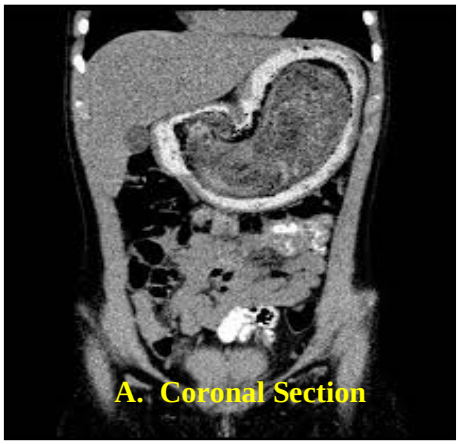
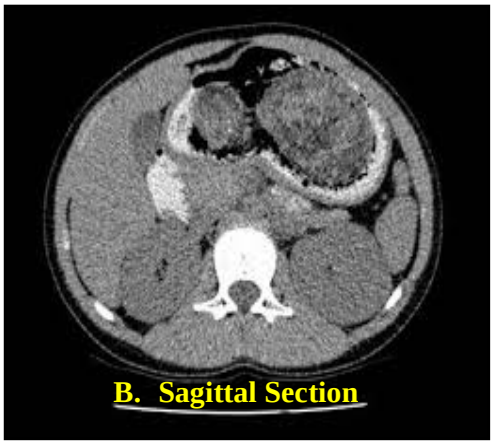


Image 3 CECT of Abdomen & Pelvis





Prophylactic head shaving was done and she was referred to psychiatric department where she was advised for regular follow up without any antipsychotic treatment for trichotillomania and trichophagia.

Discussion:

Rapunzel syndrome is named after a tale written in 1812 by the Brothers Grimm about a young maiden, Rapunzel, with long tresses who lowered her hair to the ground from high in her prison tower to permit her young prince to climb up to her window and rescue her.

Rapunzel syndrome is the term for a *trichobezoar* (gastric 'hair ball') which has a tail-like extension into the small bowel through the pylorus causing gastric outlet obstruction.

Human hair (especially long hair) is resistant to digestion as well as peristalsis. So it tends to stay in the stomach and over a period of time may form a large 'hair ball'. A trichobezoar may extend up to the pylorus, duodenum, or even jejunum. A part may break off into small bowel and cause small bowel obstruction.

As per literature Rapunzel syndrome occurs predominantly in young women or girls with psychiatric or emotional disorders and consists of 1) a trichobezoar with a tail 2) extension of tail into small bowel and 3) symptoms suggestive of obstruction.

The symptoms of Rapunzel syndrome is caused either by gastric outlet obstruction or its complications and includes:

- Anorexia, bloating, early satiety
- Weight loss
- Vomiting immediately following meals
- Acute epigastric pain
- Patchy hair loss seen in scalp hair

Complications:

- Obstructive jaundice
- Mechanical small bowel obstruction
- Small bowel perforation
- Peritonitis
- Acute pancreatitis

Radiographic features:

1. *Abdominal radiograph:*
    - Distended stomach shadow with an intragastric mottled gas pattern, outlined by fundal gas, which may resemble a food-filled stomach
    - Free gas shadow under diaphragm may be seen on erect radiograph if bowel perforation is present

2. *Fluoroscopy:*
    - Barium studies may show an intraluminal filling defect with mottled gas pattern without attachment to bowel wall. Over the time the interstices of trichobezoar are filled with barium. This barium may remain for a considerable period of time and can be seen in delayed radiographs when the barium has exited the stomach and duodenum
3. *Ultrasound:*
    - Echogenic mass with intense acoustic shadow seen within stomach and pylorus region.
    - Complex intraperitoneal free fluid if complicated by bowel perforation
4. *CT Scan:*
    - CT is the best imaging modality for showing the size and configuration of the trichobezoar and most accurately identifying its location
    - 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
    - Body of the mass in stomach while tail may extend to the duodenum or jejunum
    - Normal stomach wall can be traced completely separate from the lesion
    - No contrast enhancement
    - Mucosal edema and wall thickening may be seen in duodenum and jejunum
    - Intraperitoneal fluid with free gas can be seen if perforation is present

Treatment and prognosis:

Medical management is restricted to correction of anaemia and weakness. The treatment is essentially surgical. Laprotomy with extraction of bezoar is done with exploration of rest of the small bowel to look for detached bezoars. Small bowel segments showing extensive ulcerations and gangrene are resected. Psychiatric evaluation is suggested for underlying illness.

Differential diagnosis:

Although the imaging features allow confident diagnosis of trichobezoars, depending on the investigative modalities, the differentials to be considered are:

- gastrointestinal tumour such as a GIST extending into the stomach lumen
- other type of bezoar (e.g. phytobezoar)

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