Rapunzel Syndrome

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RAD 4003 Pedi Elective
Dr. Susan Greenfield
Clinical History

7 yo female who presented with episodic bilious vomiting, abdominal pain for 1 week

- **Associated symptoms**: severe pain that **radiates to her back**, **weight loss**, decreased PO intake and urinary output

- **Hx of hair pulling** since 2yo and other nervous habits/ **hx of being bullied and constipation**

- **PMHx**: nocturnal enuresis

- **Family Hx**: Mother -OCD, anxiety

- **Physical Exam**: tender to palpation in the epigastric region. Palpable **rubbery mass** in the epigastric region/RUQ about 1 cm, hair thinning
Clinical History-Labs

- Glucose: 64
- **Lipase: 2255** uptrending to 7000s during inpt stay over 3 days
- Tbili: 1.4
- Normal electrolytes, creatine, AST/ALT,/Alk phos, GGT
- UA: ketones, leukocyte esterase, WBC

- Initial U/S: mild right hydronephrosis and gallbladder sludge, normal pancreas

- Pt was treated for acute pancreatitis and continued to vomit during the course of her stay
Normal Images

T2 MRI
- Duodenum/SI
- Stomach
- Kidney
- Liver

AP X-ray
- Normal air gas pattern

https://radiopaedia.org/cases/normal-upper-abdominal-mri?lang=us
6/29: Abdomen 1 view X-ray

Incidental finding of a **dense ovoid material** in the stomach with attempt of NG tube placement
6/29- MRI w/o contrast

T1 Axial
6/29- MRI w/o contrast

T2 Axial view

- Mass in the stomach
- Gas filled bowel
- Gallbladder
- Liver
- Kidney
6/29 - Axial T2 MRI

- Stomach
- Gallbladder
- Duodenum
6/29- MRI w/o contrast
Key Imaging Findings

Pt with n/v and epigastric pain and a palpable rubbery mass being managed for acute pancreatitis

• U/S showed some peripancreatic fluid but no apparent mass
• Abdominal XR showed a dense ovoid material in the stomach with the attempted placement of NG tube
• MRI showed a tubular structure that is T2 hypointense mass found occupying the stomach and extending into the first part of the duodenum
Differential Diagnosis

• **Bezoar**-trichobezoar, lactobezoar, phytobezoar
  • Presents with abd pain, bowel obstruction/constipation, hx of anxiety or psychiatric illnesses
• **Gastric carcinoma**
  • Rare
  • weight loss, n/v, abdominal pain, anemia
• **Intramural mass**
  • GI stromal tumor, metastasis, lymphoma
  • anemia, wt loss, n/v, abdominal pain, can be asymptomatic
• Post-prandial food
Final Diagnosis

• **Trichobezoar** that fully occupied the **stomach** and extended to the **duodenum** due to **trichotillomania** or nervous habit

• **Rapunzel syndrome**: trichobezoar that extends into the small intestine
Discussion-Trichobezoar

- Caused by ingesting hair with an underlying psychiatric disorder-trichotillomania/trichophagia
  - Hair pulling seen in 1-4% of population of that 5-18% ingest
  - Hair can not pass peristalsis → mass forms combining with food → obstructed at the pylorus over time
- Seen in female children 6-10 years old
- GI symptoms present when the bezor is more advanced in size
- Increased morbidity compared to phytobezoar
  - Phytobezoar-vegetable/fiber matter
    - most common
    - tx: medically managed
Discussion

- **Symptoms**: halitosis, abdominal pain, n/v, weight loss, early satiety, can also be asymptomatic/incidental findings on imaging
  - palpable mass in LUQ
- **Complication**: pancreatitis (rare), constipation, gastric ulcers, obstruction
- **Dx**: abdominal radiograph +/- barium, non contrast CT, confirmed with EGD to get samples (Gold standard)
- **Management**: endoscopic removal, surgery
  - if palpable/complicated bezoar: surgery
  - Common to reoccur
    - Need behavior modification/therapy
Discussion-Imaging

- Only 18% of bezoars are seen on X-ray
- **U/S**: determine gastric bezoar 25% of the time
- **CT scan** show floating mass at the air-fluid level
  - correlates 97% with surgical findings
- **MRI**-not as useful
  - harder to distinguish bezoar with surrounding tissue/structure
Treatment

Pt was taken to surgery

- Ex-lap, gastrostomy with removal of bezoar, EGD to remove remaining bezoar in the duodenum
- Bezoar occupied the entire stomach and extended into the first part of the duodenum
- Multiple ulcer in the stomach and the duodenum
- Recommended Habit reversal therapy
ACR appropriateness Criteria-Palpable mass

- There was not a ACR criteria for bezoar.
- Following ACR criteria based on pt’s presentation of abdominal pain, palpable mass, and acute pancreatitis
### ACR Criteria-Acute non-localized abdominal pain

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<thead>
<tr>
<th>Procedure</th>
<th>Appropriateness Category</th>
<th>Relative Radiation Level</th>
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<tbody>
<tr>
<td>CT abdomen and pelvis with IV contrast</td>
<td>Usually Appropriate</td>
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<td>MRI abdomen and pelvis without and with IV contrast</td>
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<td>Radiography abdomen</td>
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<td>FDG-PET/CT skull base to mid-thigh</td>
<td>Usually Not Appropriate</td>
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<td>WBC scan abdomen and pelvis</td>
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<td>Nuclear medicine scan gallbladder</td>
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<td>Fluoroscopy upper GI series with small bowel follow-through</td>
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<td>Fluoroscopy contrast enema</td>
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ACR Criteria-Acute Pancreatitis

- The imaging modality of US, Abdominal, Radiograph, and MRI without contrast were appropriate.
- CT without contrast might have been better to visualize the mass.

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Cost of Imaging-MHH Inpt

• Cost of bezoar imaging:
  • Abdomen 1 view X-ray (1): $670
  • Abd MRI w/o contrast (1): $4,610
  >TOTAL: $5,280

• Total cost of imaging during inpatient stay:
  • Abdomen 2 view X-ray (1): $771
  • Abdomen 1 view X-ray (5): $670x5 = $3,350
  • US Abdomen limited (3): $1,493x3 = $4,479
  • Abd MRI w/o contrast: (1): $4,610
  • Chest X-ray 1 view (1): $683
  >TOTAL: $13,893

https://www.memorialhermann.org/patients-caregivers/memorial-hermann-charge-master/
Take Home Points

- Pt is symptomatic when the bezoar is quite extensive
- Consider trichobezoar on the differential in female children with abdominal mass/pain, GI symptoms, underlying psychiatric illnesses
- Best diagnosed with non contrast CT and confirmed with EGD + sampling
- Acute pancreatitis is a rare complication of bezoar
- Treatment is surgery severe, advanced bezoar
References

• https://www.uptodate.com/contents/gastric-bezoars?search=trichobezoar&source=search_result&selectedTitle=1~5&usage_type=default&display_rank=1#H15
• https://acsearch.acr.org/list
Questions?