Case Studies in Abdominal Pain

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Patient 1

21 y.o. M, 2nd opinion for abdominal pain and vomiting

2 years earlier: Episodes of burning mid-abdominal pain

1 year earlier: Acute burning mid-abdominal pain and vomiting; hypertension; no nausea

7-lb. weight loss

Abrupt cessation after 10 days
Patient 1

Workup

Normal laboratory studies
Normal head CT (except small pineal cyst)
Normal abdominal and pelvic CT (??Meckel’s inflammation)
Normal renal artery ultrasound
Normal MRCP
Normal colonoscopy
Normal laparoscopy
Patient 1

Fine after discharge

Similar episode 6 months later:

Hospitalized for dehydration
20-lb. weight loss
TPN
Patient 1
2nd Workup

Normal abdominal and pelvic CT
EGD: mild gastric erythema
Normal head CT
Normal UGI and SBFT
Normal colonoscopy
Normal video capsule endoscopy
Normal gallbladder ejection fraction
Negative tests for celiac disease, porphyria

Abrupt cessation after 10 days
Diagnosis?

Further tests?
Patient 1

Further History

FH: Migraine headaches
   Paternal grandmother
   Paternal aunt
   ? Father

Occasional marijuana use
Cyclic Vomiting Syndrome

Stereotypical episodes of vomiting with regard to onset (acute) and duration (<1 week)

Three or more discrete episodes in the prior year

Absence of nausea and vomiting between episodes

Supportive: history or family history of migraine headaches

Rome III, 2006
Cyclic Vomiting Syndrome

Treatment

Prevention: Discontinue marijuana and other triggers
Amitriptyline
Anti-seizure medications
Zonisamide
Levetiracetam

Treatment: Sumatriptan
Benzodiazepines
Antiemetics
Analgesics

Patient 1
Follow-up

Discontinued marijuana

Amitriptyline, increased to 50 mg hs

Sumatriptan prescription (not taken)

No attacks in over 4 years
Patient 2

79 y.o. M, 2nd opinion for abdominal pain and nausea

9 months earlier: mild intermittent nausea

7 months earlier: chronic nausea
diffuse abdominal discomfort
1 to 2 hours after eating

10-lb. weight loss

No vomiting, bloating, distention, change in bowel habits
Patient 2
Prior Evaluation

Routine labs normal

Amylase 176/110 U/L

Lipase 24 U/L

ESR normal

Celiac serology negative

*H. pylori* antibody negative
### Patient 2

#### Prior Evaluation (cont’d)

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Findings</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abdominal CT (x2):</td>
<td>diverticulosis, calcifications in aorta and</td>
</tr>
<tr>
<td></td>
<td>bilateral ileac vessels, kidney cysts, enlarged prostate</td>
</tr>
<tr>
<td>Abdominal ultrasound:</td>
<td>unremarkable</td>
</tr>
<tr>
<td>Prior colonoscopy:</td>
<td>diverticulosis, 6-mm adenoma</td>
</tr>
</tbody>
</table>
Patient 2
Upper Endoscopy

Normal

Histology: Gastric body and fundic mucosa with intestinal metaplasia, pseudopyloric metaplasia, and linear neuroendocrine cell hyperplasia

Antrum with reactive gastropathy and prominence of G cells

Interpretation: Autoimmune gastritis

Vitamin B12 level: 537
Patient 2
Prior Management

Prednisone 10 mg PO twice daily
Symptoms worsened
Discontinued by patient after 4-5 days
Trials of omeprazole twice daily and ranitidine prn: ?helped
Trial of rifaximin (for small intestinal bacterial overgrowth): no benefit
Placed on ondansetron 4 mg PO twice daily
Patient 2

Past history: HBP, laryngeal nodule, depression

Medications: Irbesartan, Lorazepam, Ondansetron

ETOH: 1-2 glasses of wine with dinner, discontinued 6 months earlier
Patient 2
Physical Examination

Chronically ill-appearing, sallow
P 78 reg, BP 124/80 mm Hg
Bilateral Dupuytren’s contractures
Chest, heart, abdomen unremarkable:
no abdominal mass, no succussion splash, no bruits
Patient 2
Additional Testing

Gastric emptying scan: normal
Serum gastrin (on omeprazole): 336 pg/mL
Parietal cell antibodies: positive (68.4 units)
Intrinsic factor antibodies: negative
Celiac serology: negative
Serum cortisol: 13.9 mcg/dL
Review of gastric histology: autoimmune gastritis
Diagnosis?

Further tests?
Patient 2

Workup

Doppler studies of mesenteric vessels:

70% stenosis of celiac trunk
Patient 2

Now what?
Patient 2

Placement of celiac artery stent

Resolution of discomfort, improvement in nausea (some GERD symptoms)

Worsening depression

Vitamin B12 supplementation
Chronic Mesenteric Ischemia

<5% of cases of intestinal ischemia

Almost always caused by mesenteric atherosclerosis

Despite collateral pathways, single-vessel disease (especially of celiac artery) does occur

Chronic Mesenteric Ischemia

Clinical Features

Abdominal cramping or discomfort usually within 30 minutes after eating, increasing and resolving over 1-3 hours

Progressive; pain can become continuous

Fear of eating ("sitophobia")

Weight loss

Possibly nausea, bloating, episodic diarrhea, malabsorption, constipation, GI bleeding
Chronic Mesenteric Ischemia

Diagnosis

Abdominal plain films and CT often normal; vascular calcifications may be seen.

Duplex US, MRA, angiography: reveal anatomic limitations to splanchnic blood flow but do not establish presence or absence of intestinal ischemia.

Often at least 2 of 3 major splanchnic vessels are severely stenotic or occluded.

Chronic Mesenteric Ischemia

Treatment

Surgical revascularization is traditional approach

Percutaneous transluminal mesenteric angioplasty alone or with stent insertion is now preferred

Symptom relief in up to 99%, but relapse rate as high as 28%

Patient 3

53 y.o. M with right-sided chest and right subcostal pain for several months

Age 19: Fundoplication for refractory esophagitis
         Eventual resumption of medical therapy (PPI) to control symptoms

Age 38: History of binge drinking
         Epigastric pain and diarrhea
         Steatorrhea
         EUS: chronic pancreatitis
         Pancreatic enzymes
         Symptoms resolved
Patient 3
Past Medical History

Atrial fibrillation: Maze procedures twice
Homozygous factor V Leiden mutation
Protein C deficiency
Venous thromboembolism and pulmonary emboli
IVC filter
Anticoagulation
L4-5 and L5-S1 diskectomy
Depression
Patient 3

Presents with right-sided chest and right subcostal pain for several months

- Stabbing
- Right lower chest/right upper quadrant
- Worse with movement, not inspiration
- No dyspnea
- Limits ability to climb stairs
- Recent URI treated with levofloxacin
- Weight loss of 15 lbs. attributed to pain
Patient 3
Family History

Heart disease (father MI, age 46)
Pancreatic cancer (maternal grandmother)
Colon cancer (maternal grandfather)
Abdominal aortic aneurysm (maternal uncle)
Patient 3
Medications

Lanzoprazole
Creon
Warfarin
Atorvastatin
Celexa
Multivitamin
Patient 3
Physical Examination

RUQ discomfort on “twisting” body
Afebrile, normal pulse and BP
Normal heart and lungs
Abdomen obese, 2-cm area of distinct tenderness in RUQ, no guarding or rebound, no mass or organomegaly, normal bowel sounds
No rash
Normal neurologic examination
Patient 3
Studies

CBC: normal
CMP: normal
Amylase: 96 U/L
Lipase: 140 U/L
INR: 2
CXR: sternal wires, otherwise normal
CT chest and abdomen: 2 small fat hernias, no pulmonary emboli, no aortic dissection, normal pancreas, no gallstones, normal liver
EKG: RBBB and left anterior hemiblock (unchanged)
Diagnosis?

Further tests?
Patient 3

Neurologic consultation:

Anterior cutaneous nerve entrapment syndrome (ACNES)

R/o thoracic radiculopathy

Unlikely: biliary disease

pancreatitis

ischemic bowel disease
Anterior Cutaneous Nerve Entrapment Syndrome (ACNES)

Entrapment of a cutaneous branch of a sensory nerve derived from a neurovascular bundle emanating from spinal levels T7 to T12

Related to intra- or extra-abdominal lesion, edema, or fibrosis (scar)

Pain is discrete, localized

“Hover sign”: guarding from the examiner’s hand

“Carnett’s sign”: increased tenderness with tensing of abdominal muscles

Anterior Cutaneous Nerve Entrapment Syndrome

Treatment

Avoidance of certain movements
Non-narcotic analgesics
Physical therapy
Injection therapy
? Laparoscopy, lysis of adhesions, subcutaneous nerve resection

Patient 3
Follow-up

Gabapentin

Thoracic MRI: no radiculopathy

Nerve block (trigger point injection)

Resolution
A 59-year-old man with fatigue, abdominal pain, anemia, and abnormal liver function
Presentation

Epigastric distress, ankle edema for 3 days
Personal distress
Difficulty sleeping
Dysgeusia
Nausea

T 37.3°  BP 110/68 mm Hg  ankle edema
Hct 30.6
Omeprazole and sucralfate
Next Days

Pain in both legs, dysgeusia

Loose stools

Increased abdominal pain, pain in right knee and both shoulders, fatigue

T 37.2° C  P 89/min  BP 131/89 mm Hg

Tender right knee
ED Visit

Diffuse abdominal pain, worse with eating, radiating to left side of chest, neck, shoulder, back

Constipation for several days, nausea, shortness of breath

BP 187/90 mm Hg / 129/80 mm Hg
Key Test Results

<table>
<thead>
<tr>
<th>Test</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hct</td>
<td>31.6%, basophilic stippling</td>
</tr>
<tr>
<td>ESR</td>
<td>3 mm/hr</td>
</tr>
<tr>
<td>Na</td>
<td>138 → 126 mmol/L</td>
</tr>
<tr>
<td>T. Bili</td>
<td>1.8 mg/dL (D. bili 0.4 mg/dL)</td>
</tr>
<tr>
<td>ALT</td>
<td>349 U/L</td>
</tr>
<tr>
<td>AST</td>
<td>179 U/L</td>
</tr>
<tr>
<td>Fe</td>
<td>166 mcg/dL</td>
</tr>
<tr>
<td>TIBC</td>
<td>211 mcg/dL</td>
</tr>
<tr>
<td>Ferritin</td>
<td>274 ng/mL</td>
</tr>
<tr>
<td>Abd x-ray</td>
<td>Dilated colon</td>
</tr>
<tr>
<td>CT</td>
<td>Large amount of stool in colon</td>
</tr>
</tbody>
</table>
Summary

59-year-old Man with Acute Abdominal Pain

Stress
Difficulty sleeping
Fatigue
Nausea
Dysgeusia
Constipation
Edema, varicosities
Muscle and joint pains

Tachycardia
Labile blood pressure
Aminotransferase elevations
Increased iron saturation
Acute anemia
Basophilic stippling
Colonic pseudo-obstruction
Hyponatremia
Diagnosis?

Diagnostic test?
Acute Porphyria

Abdominal pain
Nausea
Stress, restlessness
Constipation, pseudo-obstruction
Pain in extremities
Tachycardia, episodic hypertension
Syndrome of inappropriate ADH
(Red-colored urine)

Not: dysgeusia, basophilic stippling

The Porphyrias

Classification

Acute (Neurovisceral)

Cutaneous
<table>
<thead>
<tr>
<th>Acute (Neurovisceral) Porphyrias</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
</tr>
<tr>
<td><strong>Enzymatic Defect</strong></td>
</tr>
<tr>
<td><strong>Mode of Inheritance</strong></td>
</tr>
<tr>
<td><strong>Usual Age of Onset</strong></td>
</tr>
<tr>
<td><strong>Major Site of Expression</strong></td>
</tr>
<tr>
<td><strong>Major Biochemical Findings</strong></td>
</tr>
<tr>
<td>Acute intermittent porphyria</td>
</tr>
<tr>
<td>PBG deaminase</td>
</tr>
<tr>
<td>Autosomal dominant</td>
</tr>
<tr>
<td>Adulthood</td>
</tr>
<tr>
<td>Liver</td>
</tr>
<tr>
<td>Urine: ALA &lt; PBG</td>
</tr>
<tr>
<td>Plumboporphyria</td>
</tr>
<tr>
<td>ALA dehydratase</td>
</tr>
<tr>
<td>Autosomal recessive</td>
</tr>
<tr>
<td>Childhood</td>
</tr>
<tr>
<td>Liver</td>
</tr>
<tr>
<td>Urine: ALA</td>
</tr>
<tr>
<td>Hereditary coproporphyria</td>
</tr>
<tr>
<td>Coproporphyrinogen oxidase</td>
</tr>
<tr>
<td>Autosomal dominant</td>
</tr>
<tr>
<td>Adulthood</td>
</tr>
<tr>
<td>Liver</td>
</tr>
<tr>
<td>Urine: ALA &gt; PBG, coproporphyrin</td>
</tr>
<tr>
<td>Stool: coproporphyrin</td>
</tr>
<tr>
<td>Variegate porphyria</td>
</tr>
<tr>
<td>Protoporphyrinogen oxidase</td>
</tr>
<tr>
<td>Autosomal dominant</td>
</tr>
<tr>
<td>Adulthood</td>
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<tr>
<td>Liver</td>
</tr>
<tr>
<td>Urine: ALA &gt; PBG, coproporphyrin</td>
</tr>
<tr>
<td>Stool: coproporphyrin, protoporphyrinogen</td>
</tr>
</tbody>
</table>
Heme Synthesis Pathway

Glycine + Succinyl CoA

\[ \text{ALA synthase} \]

5-aminolevulinic acid

\[ \text{ALA dehydratase} \]

Porphobilinogen

\[ \text{PBG deaminase} \]

Hydroxymethylbilane

\[ \text{Uroporphyrinogen III co synthase} \]

Uroporphyrinogen III

\[ \text{Uroporphyrinogen III decarboxylase} \]

Coproprophyrinogen III

\[ \text{Coproporphyrinogen oxidase} \]

Protoporphyrinogen IX

\[ \text{Protoporphyrinogen oxidase} \]

Protoporphyrin IX

\[ \text{Ferrochelatase} \]

Heme

ALAs, 5-aminolevulinic acid; PBC, porphobilinogen
Acute Porphyria

Triggers

Starvation, negative energy balance
Drugs*
Alcohol
Smoking
Infections
“Stress”

* Not pirbuterol, per the American Porphyria Foundation (www.porphyriafoundation.org)
Features Inconsistent with Acute Porphyria

Dysgeusia

Basophilic stippling

(First attack age 59)
Dysgeusia
Altered sense of taste

Chemotherapeutic agents
- Cyclophosphamide
- Cisplatin

Pesticides and other toxins
- Lead
- Zinc deficiency

Other drugs
- Albuterol, pirbuterol
- Histamine H1-receptor antagonist
- D-penicillamine
- Metronidazole
- Boceprevir

Xerostomia
Basophilic Stippling

Sideroblastic anemia
Lead poisoning
Arsenic poisoning*
Thalassemia
Erythrocyte 5′-nucleotidase deficiency
Thrombotic thrombocytopenic purpura

*Associated with garlicky odor in breath, not true dysgeusia, and with severe diarrhea and pulmonary toxicity
Lead Poisoning

Features

Abdominal pain (“lead colic”)  Hypertension
Nausea  Acute anemia
Dysgeusia  Basophilic stippling
Constipation, pseudo-obstruction  SIADH
Joint and muscle pains  Fanconi-type syndrome
Neuropsychiatric effects

(Aminotransferase elevations may be seen)

Lead Poisoning can be Confused with Acute Porphyria

Lead inhibits ALA dehydratase
Overproduction of ALA in both
Plumboporphyria = deficiency of ALA dehydratase
(children; one case in a 63-year-old man with polycythemia vera)

PBG also elevated in other porphyrias

(ALA and PBG are elevated during an attack)

Lead Poisoning
Signs

“Lead lines”
Bluish pigmentation at the gum-tooth line

Deposition of lead in bone
Blood Lead Levels

Elevated

≥10 mcg/dL

Acute lead poisoning

>100 mcg/dL

Inhibition of heme synthesis

~55 mcg/dL

(ALA dehydratase)

↑ Free erythrocyte protoporphyrin

↑ Zinc protoporphyrin
What was the source of lead exposure?
Lead Poisoning

Sources

Work place exposures

Lead smelting and refinement

Coal combustion

Manufacture:  Batteries  Solder
              Paints     Tin cans
              Pigments  Lead glazing
              Car radiators  Ammunition
              Cable and wire  Cosmetics

Lead Poisoning

Sources

Home exposures

Lead paint
Home renovations
Natural disasters
Moonshine
Lead glass (leaching)
“Laced” marijuana

Curious Features of Case

Patient’s “distress because of personal issues”

No exposure or occupation history

Partner
Phobic anxiety of lead poisoning?

or

A stressful relationship?
A Game of Clue???

The partner,
In the kitchen,
With lead!
Diagnosis

Lead poisoning
Lead level
91 mcg/dL

Zinc protoporphyrin
425 µmol/mole hgb