

# AMSER Case of the Month

## November 2025

53-year-old woman with abdominal distension and  
nausea

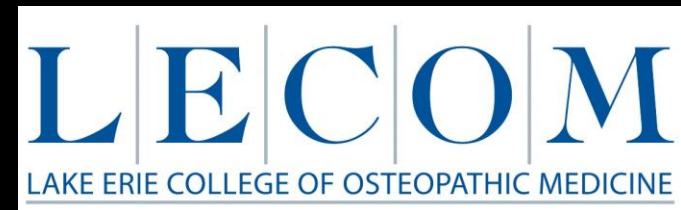
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**ROCHESTER**  
REGIONAL HEALTH



# Patient Presentation

- **HPI:** 53-year-old woman with past medical history of anxiety, uterine fibroids, pancreatitis, and polysubstance abuse presented to the emergency department with nausea, bloating and epigastric discomfort.
- **Physical Exam:** Positive for distension. Bowel sounds normal. No abdominal tenderness, guarding, or rebound.
- **Pertinent Labs:** CBC, WBC, BUN and Cr within normal limits

What Imaging Should We Order?

# Select the applicable ACR Appropriateness Criteria

**Variant 4:** Acute nonlocalized abdominal pain. Not otherwise specified. Initial imaging.

| Procedure   | Appropriateness Category | Relative Radiation Level |
|---|--------------------------|--------------------------|
| CT abdomen and pelvis with IV contrast                      | Usually Appropriate      | ⊕⊕⊕                      |
| CT abdomen and pelvis without IV contrast                   | Usually Appropriate      | ⊕⊕⊕                      |
| MRI abdomen and pelvis without and with IV contrast         | Usually Appropriate      | ○                        |
| US abdomen  | May Be Appropriate       | ○                        |
| MRI abdomen and pelvis without IV contrast                  | May Be Appropriate       | ○                        |
| CT abdomen and pelvis without and with IV contrast          | May Be Appropriate       | ⊕⊕⊕⊕                     |
| Radiography abdomen   | May Be Appropriate       | ⊕⊕                       |
| FDG-PET/CT skull base to mid-thigh                          | Usually Not Appropriate  | ⊕⊕⊕⊕                     |
| WBC scan abdomen and pelvis                                 | Usually Not Appropriate  | ⊕⊕⊕⊕                     |
| Nuclear medicine scan gallbladder                           | Usually Not Appropriate  | ⊕⊕                       |
| Fluoroscopy upper GI series with small bowel follow-through | Usually Not Appropriate  | ⊕⊕⊕                      |
| Fluoroscopy contrast enema                                  | Usually Not Appropriate  | ⊕⊕⊕                      |

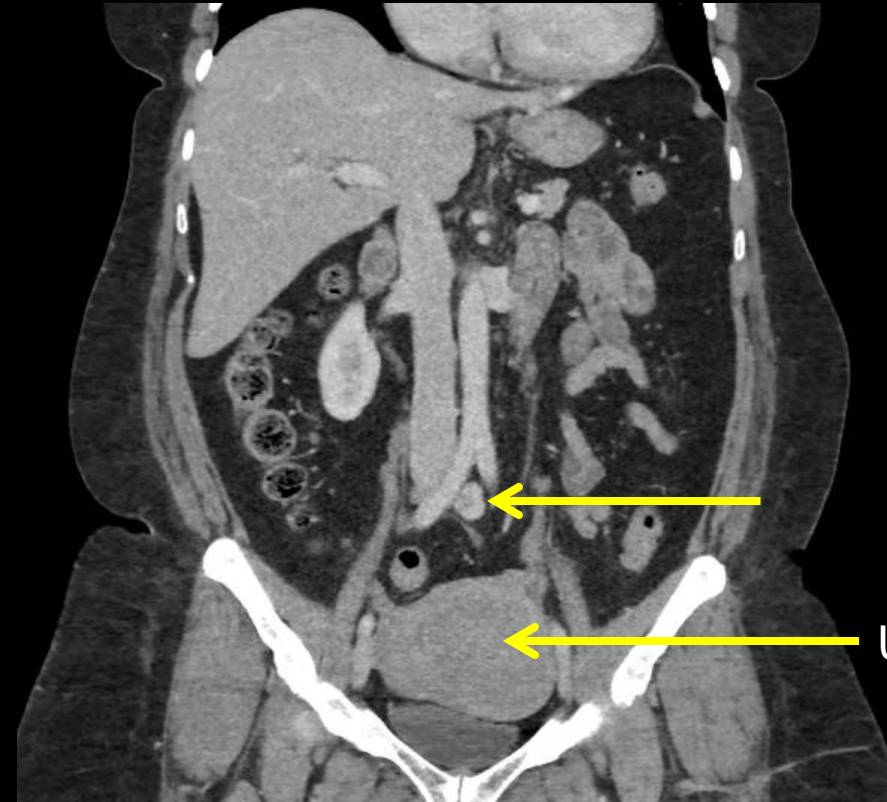
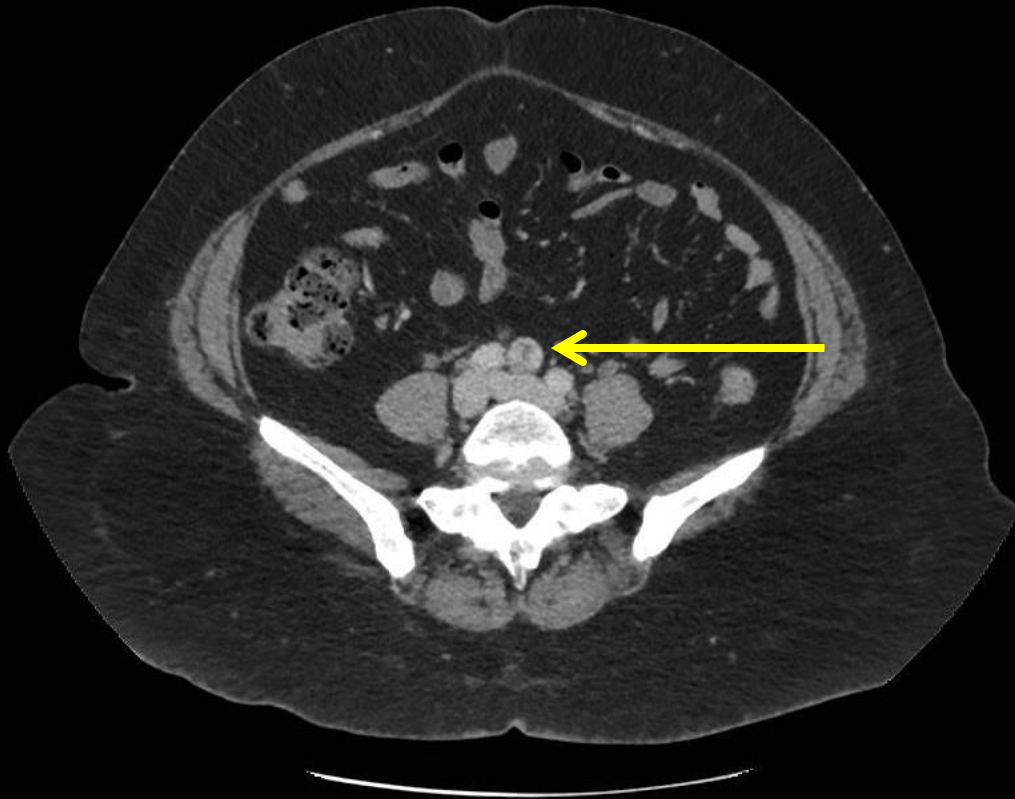
This imaging modality was ordered by the ER physician

# Findings (unlabeled)



CT Abdomen & Pelvis with IV Contrast

# Findings: (labeled)



CT Abdomen & Pelvis with IV Contrast

**Findings:** 1.7 x 1.6 cm soft tissue density at the aortic bifurcation

Final Dx:

Non-functioning paraganglioma of the organ of  
Zuckerkindl (benign)

# Case Discussion

## Paraganglia Basics:

- Clusters of neuroendocrine cells derived from neural crest cells<sup>1</sup>
- Widely distributed throughout the body: adrenal medulla (largest cluster), paravertebral space, head/neck region<sup>1</sup>
- Paragangliomas are rare neuroendocrine tumors
  - Potentially fatal, often neglected- mimic anxiety, panic disorder, thyrotoxicosis, and hypoglycemia<sup>2</sup>
  - However, approximately 10–30% of paragangliomas are discovered incidentally<sup>3</sup>
    - In one series of abdominal paragangliomas, the rate of incidental findings was about 11%<sup>4</sup>

# Case Discussion

## The “rule of 10s”:

- The "rule of 10s" is a historical concept used to describe the characteristics of pheochromocytoma and paraganglioma (PPGLs)<sup>5,6</sup>:
  - **10% Extra-adrenal:** These tumors occur outside the adrenal glands.
  - **10% Bilateral:** Both adrenal glands are affected.
  - **10% Malignant:** The tumors have metastasized.
  - **10% Familial:** These tumors are hereditary and associated with genetic syndromes.
  - **10% Asymptomatic:** These tumors are not associated with symptoms like hypertension.
  - **10% Occur in Children:** This was a traditional part of the rule.
- However, newer data suggests larger proportions of these tumors are asymptomatic, malignant, or carry a germ-line mutation.<sup>7</sup>

# Case Discussion

## Types of Paragangliomas:

- Parasympathetic (primarily in the head and neck and usually nonfunctional)<sup>8</sup>
- Sympathetic (primarily in the chest and abdomen)<sup>8</sup>
  - Can occur anywhere along the sympathetic chain, from the skull base down to the pelvic organs<sup>8</sup>
  - Most common sites: IVC and left renal vein junction and at the aortic bifurcation near the origin of inferior mesenteric artery (the organ of Zuckerkandl)<sup>8</sup>
  - **In this case, paraganglioma of the organ of Zuckerkandl was found:**
    - The organ of Zuckerkandl is a small cluster of chromaffin cells (neural crest origin) that originates from just above the SMA/renal arteries to the aortic bifurcation or slightly beyond<sup>9</sup>

# Case Discussion

## Pathophysiology:

- Catecholamine excess → episodic hypertension, palpitations, tremor, anxiety<sup>10</sup>
  - Catecholamines include dopamine, norepinephrine, and epinephrine<sup>10</sup>
- Mass effect → compression/bony erosion due to mass effect<sup>10</sup>
- Paragangliomas can be sporadic or hereditary:
  - Sporadic: diagnosed between the age 30 to 50 with a female-to-male ratio of 3:1<sup>1</sup>
  - Hereditary: younger onset with an equal male-to-female ratio<sup>1</sup>
    - Associated syndromes:<sup>2</sup>
      - von Hippel-Lindau Syndrome (VHL)
      - Multiple endocrine neoplasia 2A (MEN2A)
      - Neurofibromatosis type 1 (NF1)

# Case Discussion

## Diagnosis

- Anatomical and functional imaging is required for diagnosis and staging:
  - Anatomic Imaging: CT and MRI<sup>10</sup>
  - Functional imaging: <sup>68</sup>Ga-DOTATATE PET, <sup>18</sup>F-FDA PET, <sup>18</sup>F-DOPA PET, <sup>18</sup>F-FDG PET<sup>10</sup>
- Presumptive diagnosis can be made through measurement of urine and plasma metanephrines<sup>10</sup>
- Precautions:
  - If catecholamine excess has not been ruled out, avoid contrast CT unless alpha blockade has been given<sup>10</sup>
  - Biopsy contraindicated until biochemical screening is negative for catecholamine excess<sup>10</sup>
- Definitive diagnosis requires confirmation by histology<sup>10</sup>
  - Early identification with complete surgical resection is often curative with a good prognosis, **as in this case**<sup>8</sup>

# Case Discussion

## Anatomical Imaging Findings<sup>11</sup>

### Contrast Enhanced CT

- Morphology: **variable**, can mimic other conditions
- Appearance:
  - **Heterogeneous** with cystic, necrotic, or calcified areas
  - **Solid and homogeneous**
  - Typically **well-circumscribed, encapsulated**
- Imaging appearance **not predictive** of malignancy
- Highly vascular → prone to **hemorrhage/necrosis**, even if benign
- May show **capsular or vascular invasion** (e.g., IVC) in benign tumors

### MRI

#### T1WI:

- Usually **hypointense** to adrenal gland
  - May be **heterogeneous** (necrosis/hemorrhage)
- **No signal loss** on opposed-phase (no microscopic fat)

#### T2WI:

- Can be **markedly hyperintense**, though ~1/3 are not

#### Post-contrast T1WI:

- Heterogeneous enhancement (similar to CECT)
- **No washout**

# Case Discussion

## Functional Imaging Findings<sup>11</sup>

### DOTATATE PET/CT

- DOTATATE is a somatostatin analogue that binds **somatostatin receptors (SSTRs)**
  - High affinity for **SSTR2**
    - Sensitive for PPGLs due to SSTRs receptor overexpression
  - Available tracers: **Cu-64** and **Ga-68 DOTATATE**
- **Higher uptake** than background in tumor locations
- **Physiologic adrenal uptake** can complicate adrenal lesion diagnosis
- Whole-body scan can detect **synchronous/multiple tumors**

### <sup>18</sup>F-FDG PET

- Uptake reflects **increased glucose utilization** by tumors
- Useful for small adrenal masses (less affected by adrenal physiologic uptake)
- **<sup>18</sup>F-FDG uptake > liver uptake** → suspicious for pheochromocytoma
- Paraganglioma = variable uptake
  - Evaluate for **low- to high-level uptake** in expected sites of paraganglioma
- Whole-body imaging can detect **synchronous/multiple tumors**

# Patient Course

- Initial presentation (nausea, bloating, epigastric discomfort) attributed to uterine fibroids
- Incidental para-aortic soft tissue density warranted further investigation prior to surgery for removal of uterine fibroids
  - Patient later recalled worsening anxiety, profuse sweating, and a racing heart for "a couple months"
  - Endocrinology referral: plasma and urine metanephrines returned within normal limits
- Ultimately, complete excision of a 1.9 cm nodule was removed at the same time as gynecology performed a total abdominal hysterectomy and bilateral salpingo-oophorectomy
- Pathology results confirmed the diagnosis of a benign paraganglioma of the organ of Zuckerkandl
  - Immunoprofile positive stains for synaptophysin, CD56, S100 supported the diagnosis

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