An Extremely Rare Case Report

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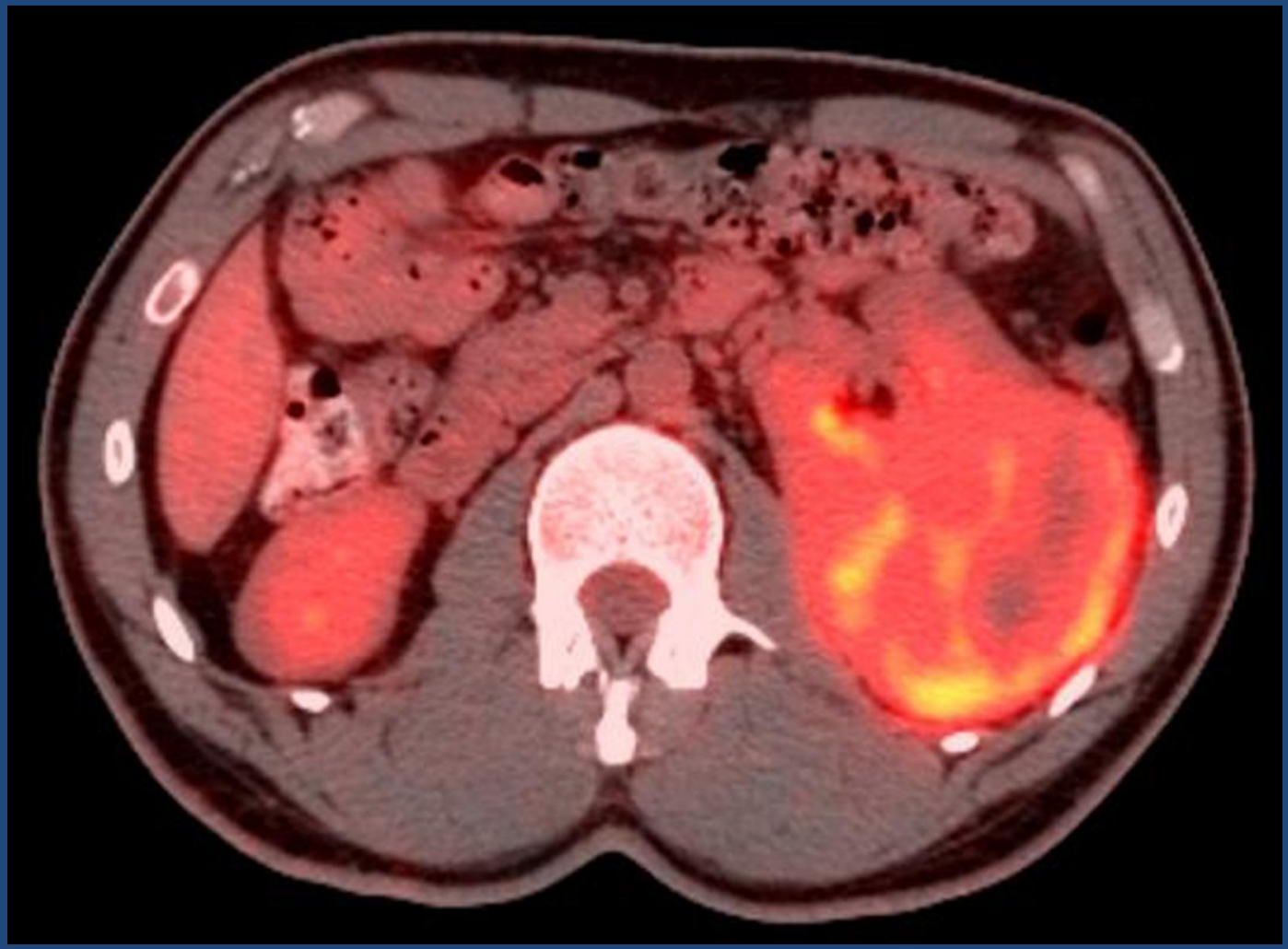
Case Report

- 37-year-old male
- 5-week Hx of left flank pain & early satiety
- PE: left flank — moderately enlarged & tender mass
- All labs normal

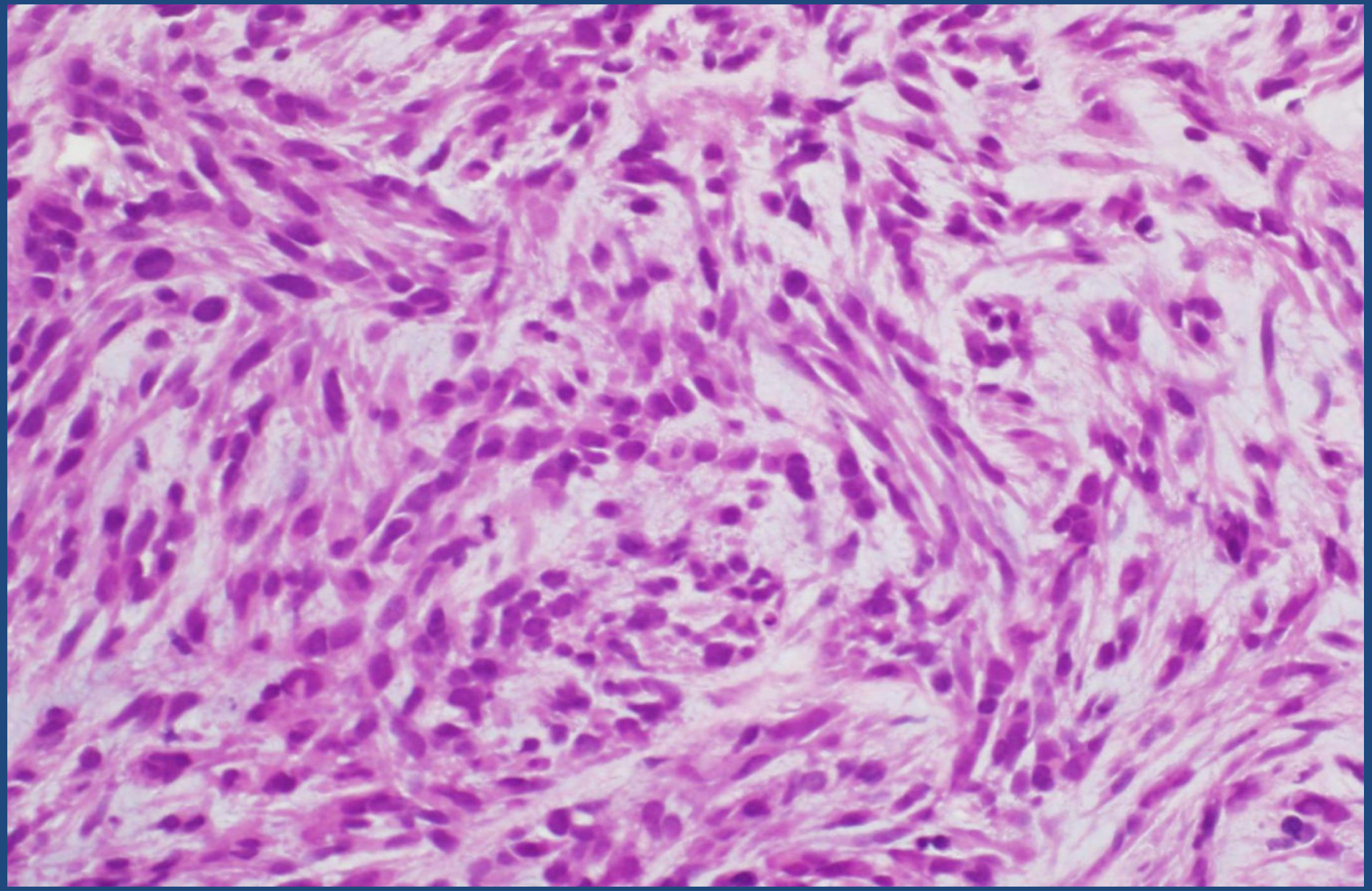
PET–CT Scan



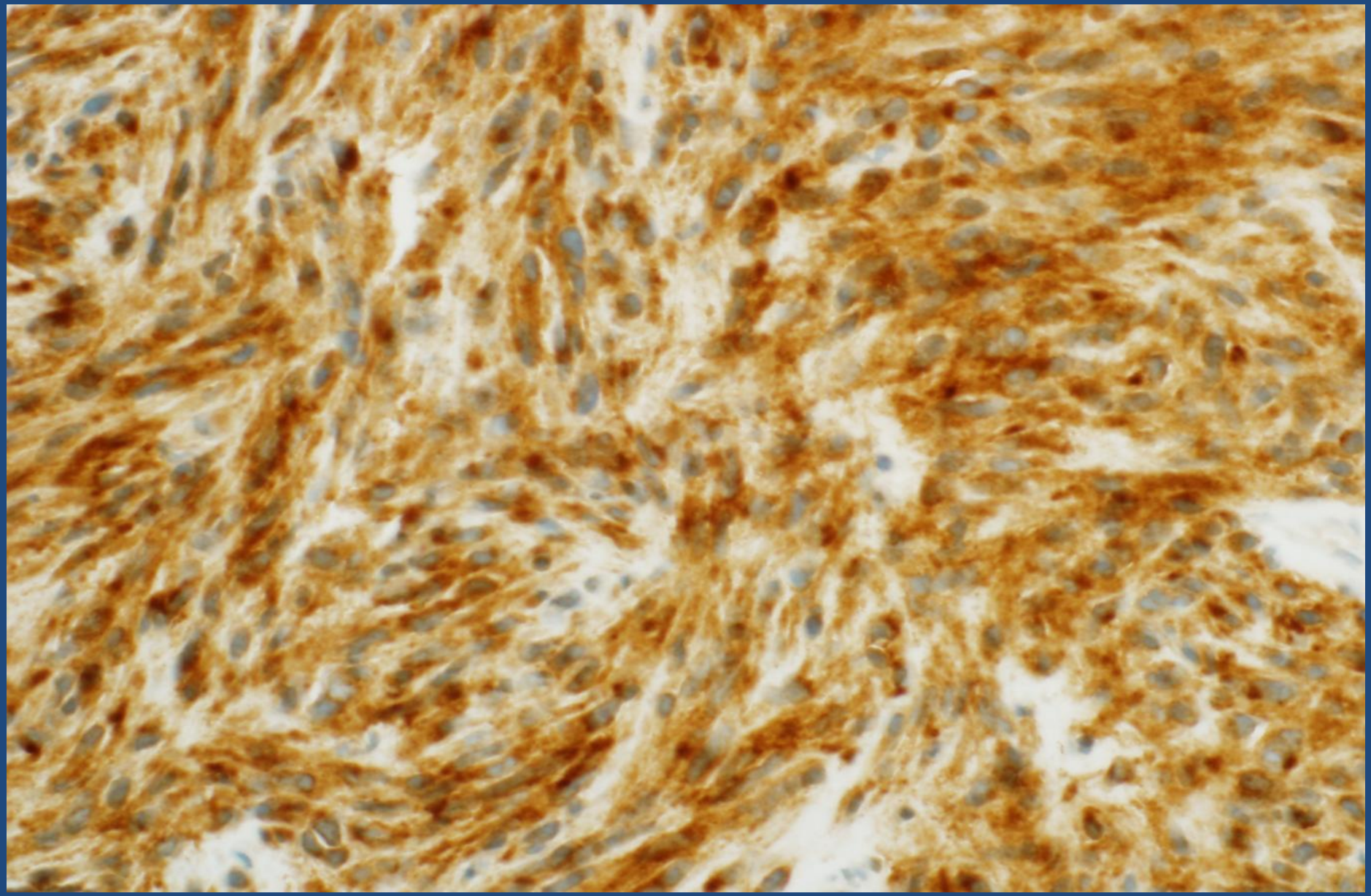
PET–CT Scan:

14 x 11 cm hypermetabolic FDG-avid mass lesion involving the left adrenal gland & dorsal part of the left hemi-diaphragm.

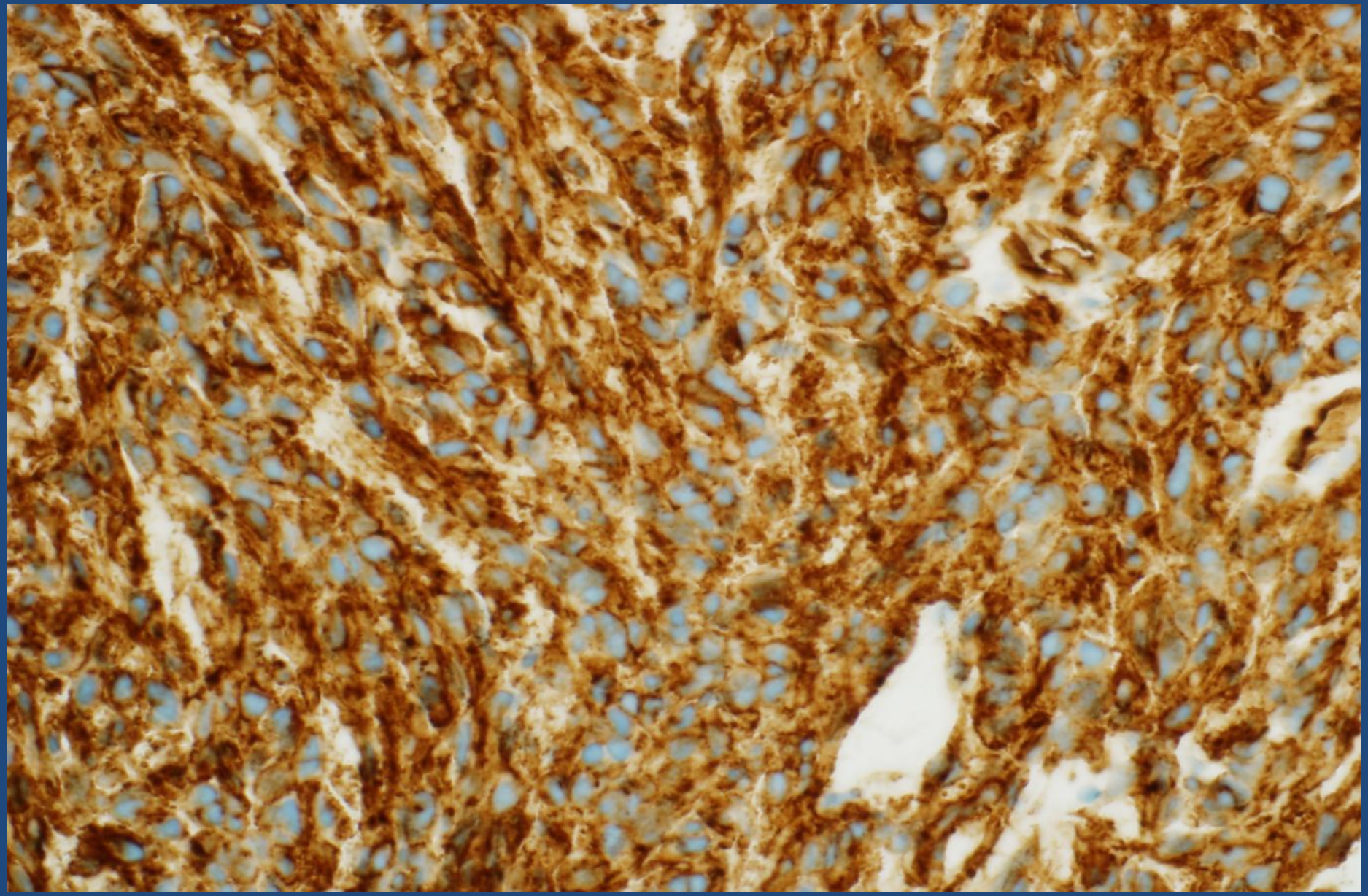
Hematoxylin & Eosin Stain



Immunoreactivity to CD-117 (c-kit)



Immunoreactivity to CD-34

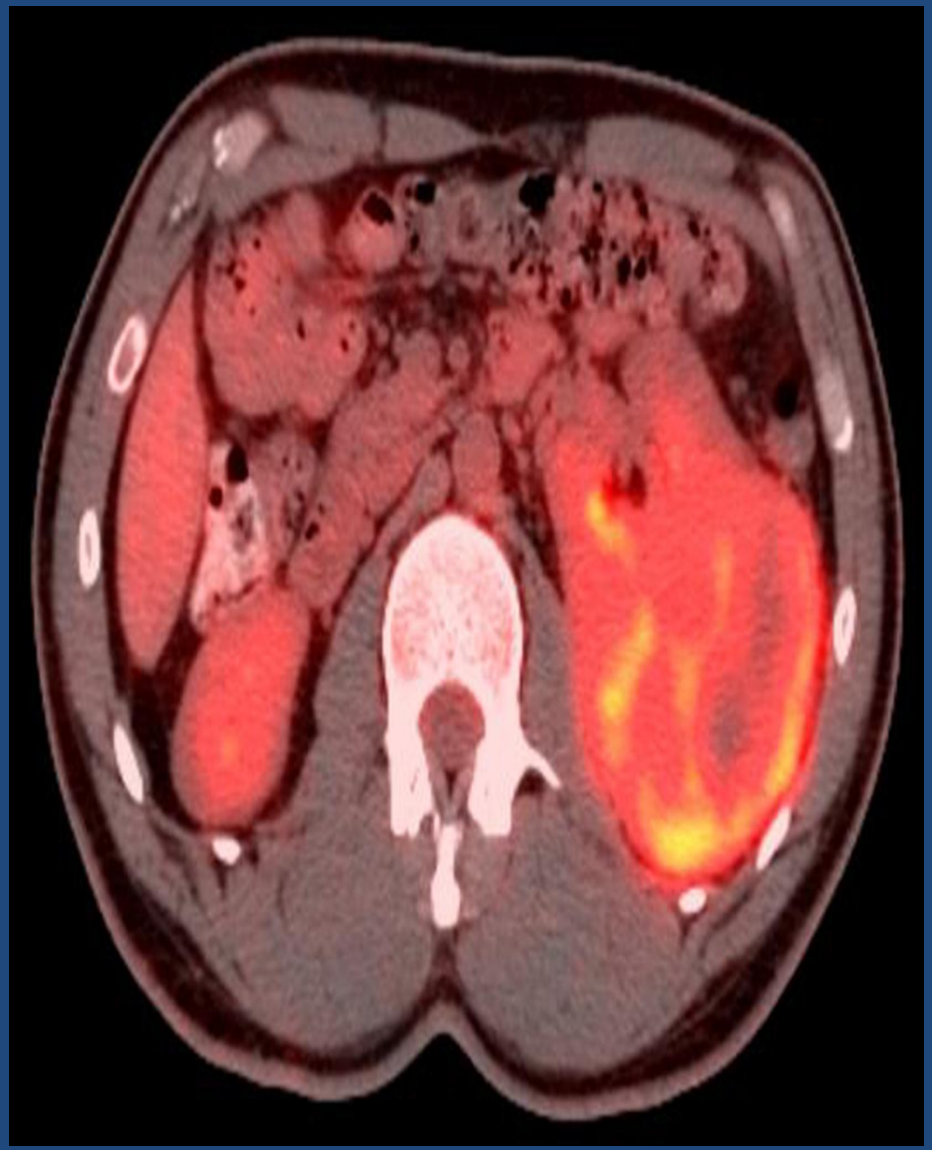


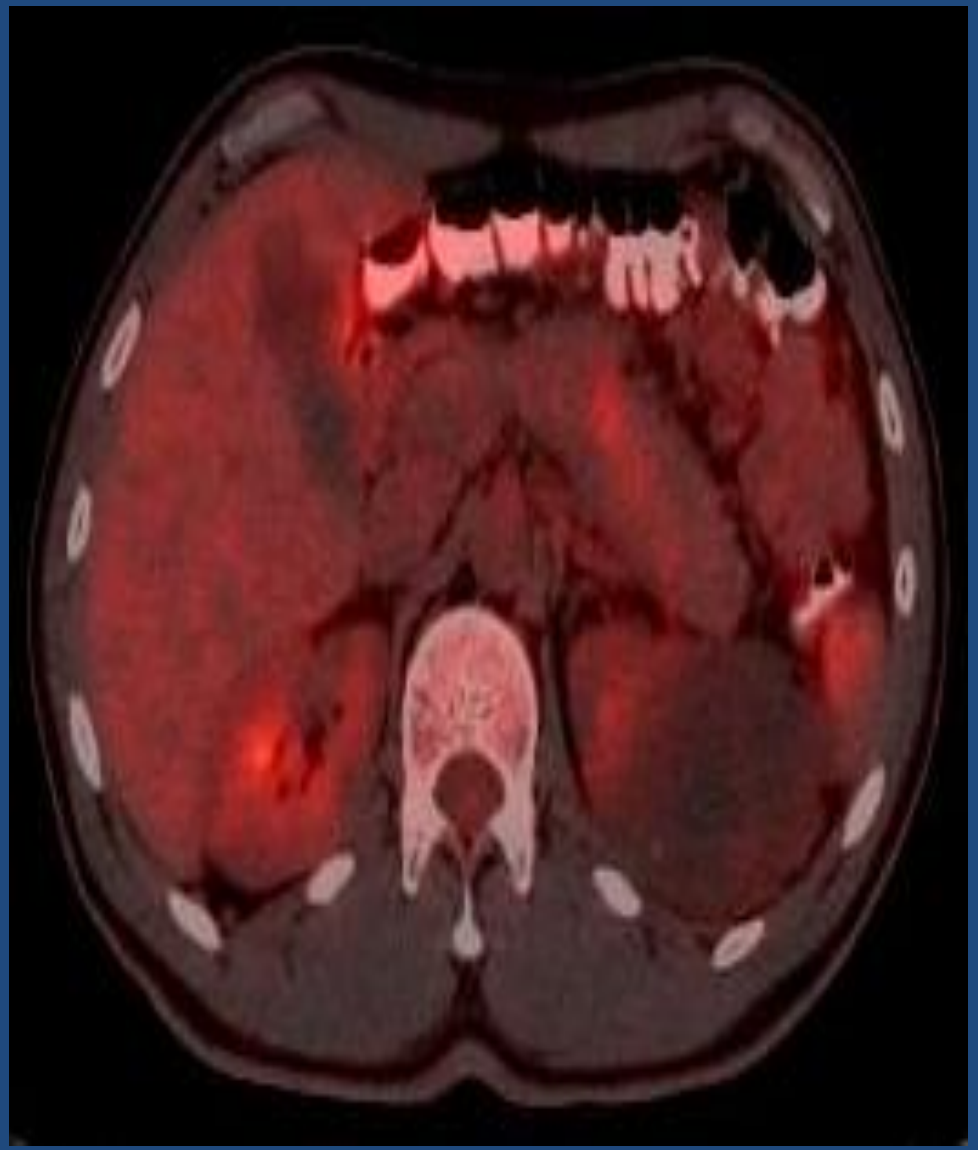
Final Diagnosis

- Primary Gastro-Intestinal Stromal Tumor (GIST) of the left adrenal gland
- Surgery was advised
  - Patient refused
  - Alternative treatment was imatinib

3-month F/U PET–CT

PET—CT: pre and post Tx





GIST (1/8)

- Mesenchymal tumors
- Arise from interstitial cells of Cajal
- c-kit (90%) and PDGFRA mutations
- Common sites:
  - Stomach (40-70%)
  - Small intestine (20-40%)
  - Esophagus, colon & rectum (< 10%)

GIST (2/8)

- Uncommon sites:
  - Adrenal gland
  - Only 1 published “case report”
    - Sereg et al. Gastrointestinal stromal tumor presenting as a hormonally inactive adrenal mass Endocrine. 2011 Feb;39(1):1-5

GIST (3/8)

- 70% of cases are symptomatic
- Common symptoms:
  - Mass effect
  - Abdominal pain, GI bleeding, fatigue, bloating, weight loss
- Radiologic modality:
  - CT Scan
  - PET—CT scan: FDG-avid lesions

GIST (4/8)

- **Histopathology**:
  - Spindle cell (70%) neoplasm
  - Epitheliod cell (30%) neoplasm
- **Immunohistochemistry**:
  - 95% positive to CD-117 (c-kit)
  - ??% positive to CD-34
  - Negative to CK, CD-31, S-100, desmin, vimentin

GIST (5/8)

- **Management**:
  - Surgery: the gold standard
  - Imatinib: alternative therapy
    - Tyrosine kinase inhibitor (TKI)
    - Decrease tumor size and recurrence rate
    - Safe and well-tolerated
    - Drug resistance in some patients

GIST (6/8)

- **Prognosis:**
  - tumor size and mitotic activity → not applicable to EGIST
  - most common type of mesenchymal tumors
  - <1% of tumors in GI
  - Encourage follow-up with patient

GIST (7/8)

- **Adrenal incidentoloma**:
  - 0.6 to 1.3% of all abdominal CT
  - Possible DDx:
    - adenoma, myelolipoma, cyst, lipoma, pheochromocytoma, adrenal cancer, metastatic cancer, hyperplasia, TB

GIST (8/8)

- Positive immunoreactivity to CD-117 and CD-34
- Negative immunoreactivity to:

  CD-31 –angioma, angiosarcmas,
  S-100 – melanoma, schwanoma, neurofibroma,
  cytokeratin – glandular epithelia tumors
  desmin and vimentin– sarcomas

- GI leiomyoma – Desmin 100% positive
- Fibromatosis – CD117 and CD34 negative

Conclusion (1/2)

- Primary E-GIST arising adrenal gland is extremely rare.
  - Only 1 reported case report in literature
- The proximity of the left adrenal gland to the greater curvature can be misleading

Conclusion (2/2)

- Radiological investigations (PET—CT scan) and immunohistochemical staining (CD-117 and CD-34) are essential to delineate the tumor and confirm diagnosis.
- Imatinib therapy for GISTs & EGISTs has been shown to be promising in resolving tumor and symptoms

Thank You

Riyadh, Saudi Arabia