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:
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**
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
- 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
  - Epithelioid cell (30%) neoplasm

- **Immunohistochemistry:**
  - 95% positive to CD-117 (c-kit)
  - ?% positive to CD-34
  - Negative to CK, CD-31, S-100, desmin, vimentin
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 \(\rightarrow\) not applicable to EGIST
  - most common type of mesenchymal tumors
  - <1% of tumors in GI
  - Encourage follow-up with patient
Adrenal incidentoloma:

- 0.6 to 1.3% of all abdominal CT
- Possible DDx:
  - adenoma, myelolipoma, cyst, lipoma, pheochromocytoma, adrenal cancer, metastatic cancer, hyperplasia, TB
• Positive immunoreactivity to CD-117 and CD-34
• Negative immunoreactivity to:
  
  CD-31 – angioma, angiosarcomas,
  S-100 – melanoma, schwannoma, neurofibroma,
  cytokeratin – glandular epithelia tumors
  desmin and vimentin – sarcomas

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

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