60 year old male with history of uncontrolled hypertension

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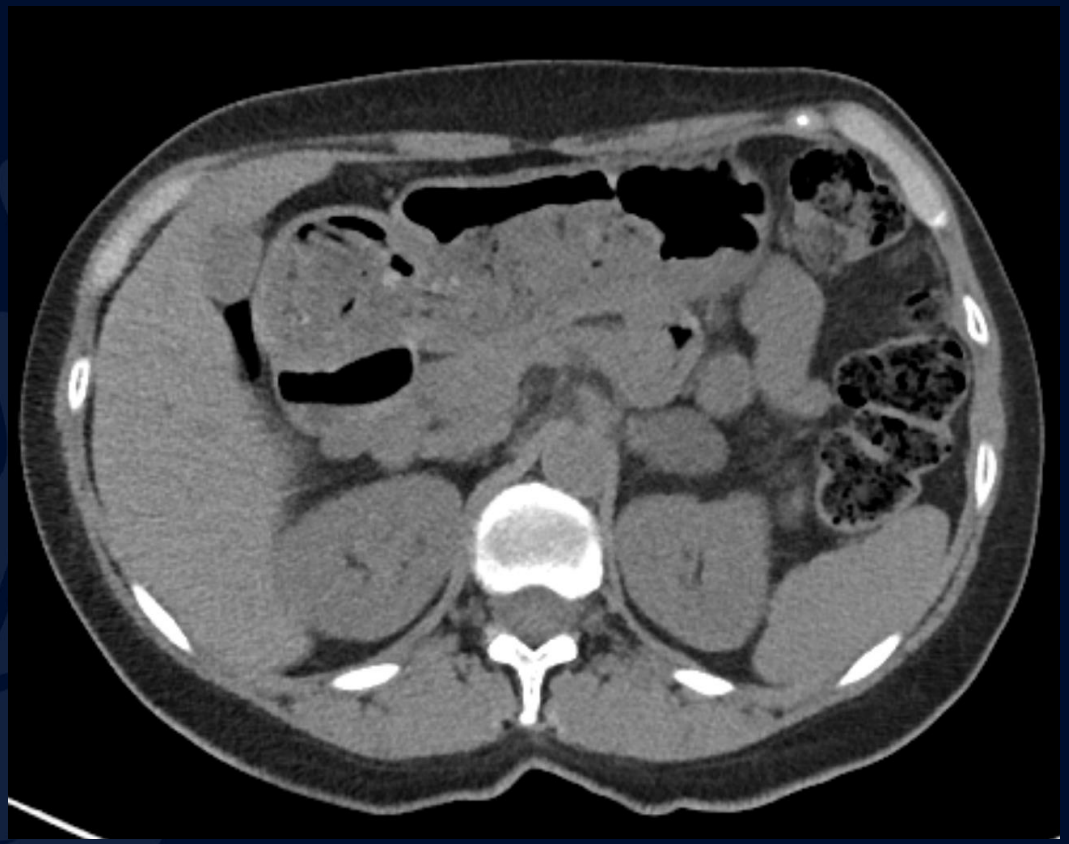

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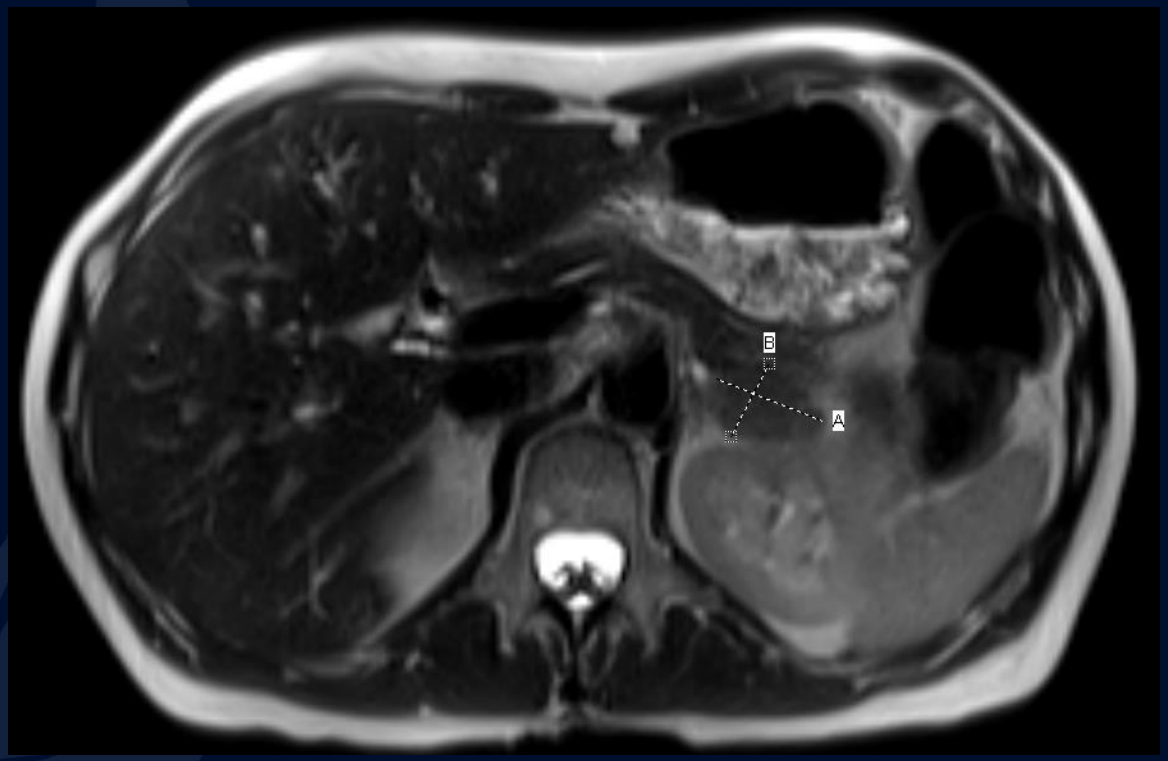

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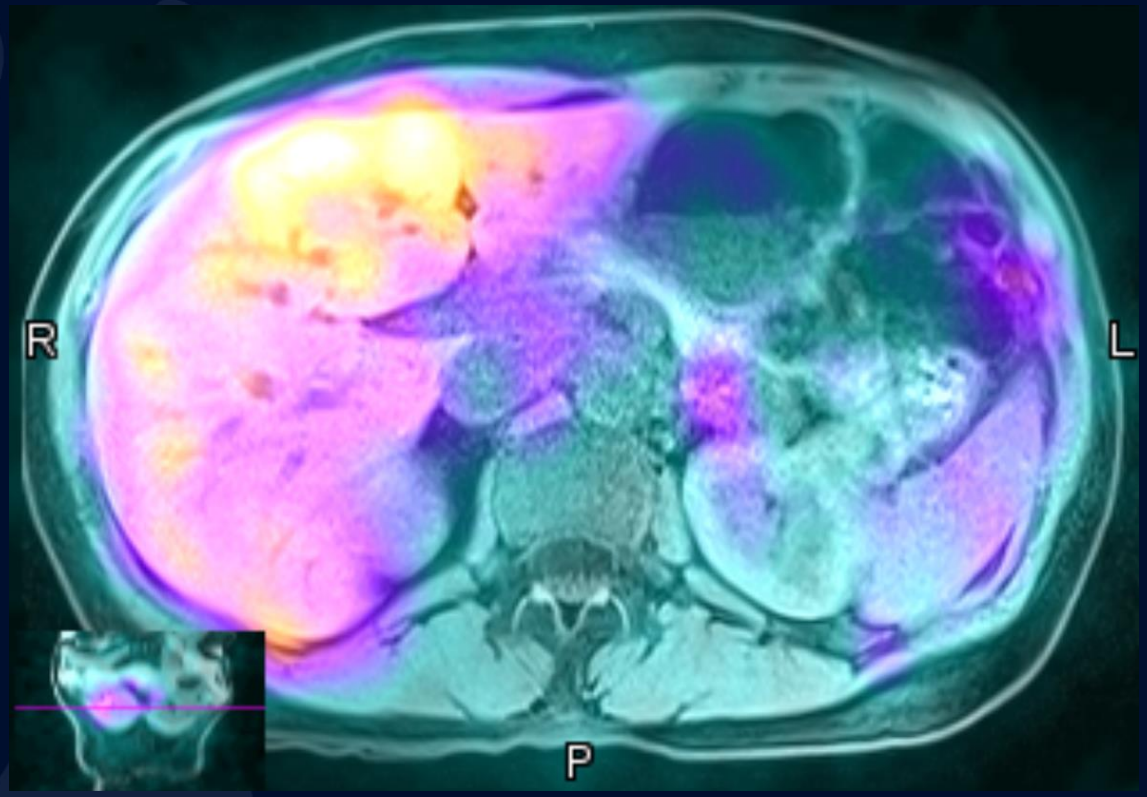

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Pheochromocytoma

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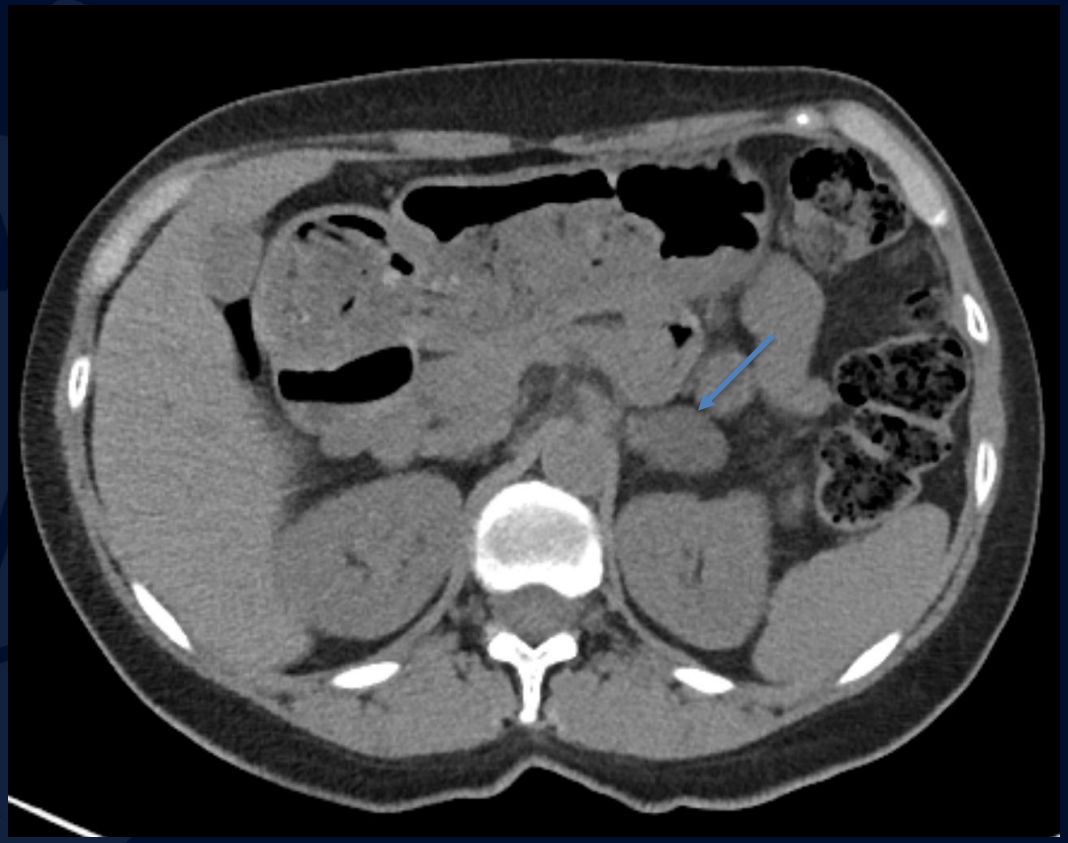
CT scan



Low-attenuation left adrenal gland mass measuring 2.9 x 2.0 cm.

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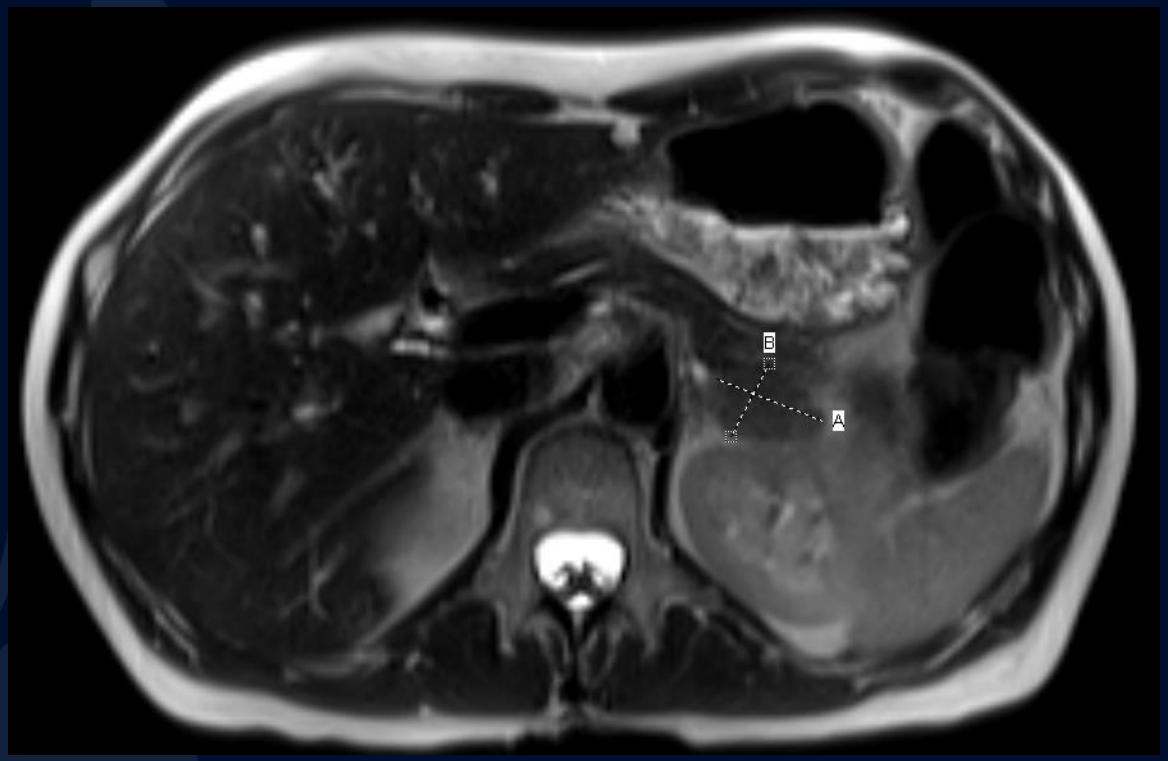
MRI abdomen



Left adrenal mass with homogeneous HASTE characteristics measuring up to 2.9 x 2.0 cm.

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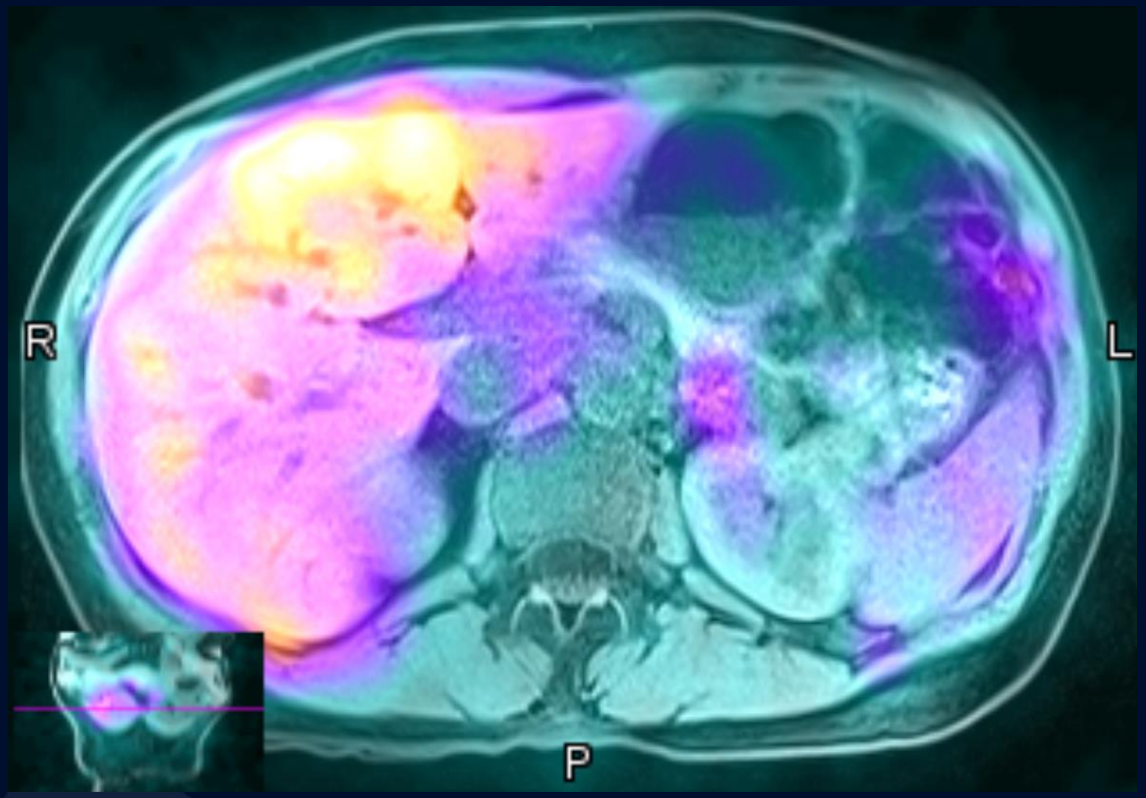
MIBG



Focus of abnormal MIBG uptake in the left adrenal region, compatible with pheochromocytoma. SPECT images were fused with the abdominal MRI on a separate workstation. Fusion images reveal good correlation between this focus of uptake and the medial portion of the LEFT adrenal nodule.

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Pheochromocytoma

- **Definition**:
  - Uncommon tumor of the adrenal gland with a 10% rule:
    - 10% are extra-adrenal
    - 10% are bilateral
    - 10% are malignant
    - 10% are found in children
    - 10% are familial
    - 10% are not associated with hypertension

- **Epidemiology**:
  - Hypertensive adults: 0.1 - 0.6%.
  - General population ~ 0.05%

- **Associations**:
  - Majority are sporadic.
  - 5-10% of cases, a pheochromocytoma is manifestation of an underlying condition:
    - Multiple Endocrine Neoplasia type II (MEN IIa and MEN IIb)
    - Von Hippel-Lindau disease
    - von Recklinghausen disease (Neurofibromatosis type I)
    - Sturge-Weber syndrome
    - Tuberous sclerosis

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Pheochromocytoma

- **Clinical presentation**:

Uncontrolled secondary hypertension, with superimposed paroxysmal hypertensive crises.

- **Pathology**:

Catecholamine-secreting tumors derived from chromaffin cells. On microscopy they show well-defined clusters of tumor cells containing eosinophilic cytoplasm separated by fibrovascular stroma.

- **Location**:

Adrenal: Arise from the chromaffin cells of the adrenal medulla.

Extra-adrenal locations: ~10% are not located in the adrenals

- Sympathetic chain
- Urinary bladder
- Organ of Zuckerkandl.
- Thoracic paragangliomas

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Pheochromocytoma

- **CT findings**:
  - Non-contrast CT: Attenuation ranges from low density to soft tissue attenuation and almost always > 10 HU.
  - Contrast CT: Heterogeneous enhancement due to tissue necrosis, cystic degeneration, hemorrhage.

- **MRI findings**:
  - T1WI: Iso-intense to muscle and hypo-intense to liver.
  - T2WI: Markedly hyper-intense ("light bulb").

- **Nuclear Medicine findings**:
  - I-123 Metaiodobenzylguanidine (MIBG) Scan.
  - Most common and available technique.
  - MIBG is norepinephrine analogue: Uptake proportional to number of neuro-secretory granules within lesion.
  - I-123 sensitivity: 77-90%; specificity: 95-100%.
  - Particularly useful for extra-adrenal paraganglioma detection.

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Pheochromocytoma

Differential Diagnosis:

- Adrenal Adenoma
- Adrenal Carcinoma
- Adrenal Metastases and Lymphoma
- Adrenal Myelolipoma
- Adrenal Hemorrhage
- Adrenal Tuberculosis and or fungal infection

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- 1. Ros PR, Mortele KJ. CT and MRI of the abdomen and pelvis, a teaching file. Lippincott Williams & Wilkins. (2006) ISBN:0781772370.
- 2. Hoegerle S, Nitzsche E, Altehoefer C et-al. Pheochromocytomas: detection with 18F DOPA whole body PET--initial results. Radiology. 2002;222 (2): 507-12.
- 3. Pacak K, Eisenhofer G, Lenders JW. Pheochromocytoma, Diagnosis, Localization, and Treatment. Wiley-Blackwell. (2007) ISBN:1405149507.

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