

AMSER Case of the Month:

35-year-old man with retroperitoneal mass who had been lost to follow up

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Patient Presentation

- HPI:
 - 35 y.o. male presenting with right flank pain.
 - Admits to intermittent symptoms of hot flashes, palpitations, and diaphoresis which occur monthly and last up to 1 hour
 - 15 lbs. weight gain in last 6 months and increased fatigue
- PMH:
 - Essential hypertension
 - Lumbar fusion with chronic pain controlled with opioids
 - Outside medical report of a 3.5 cm retroperitoneal mass in 2015 (images not immediately available)

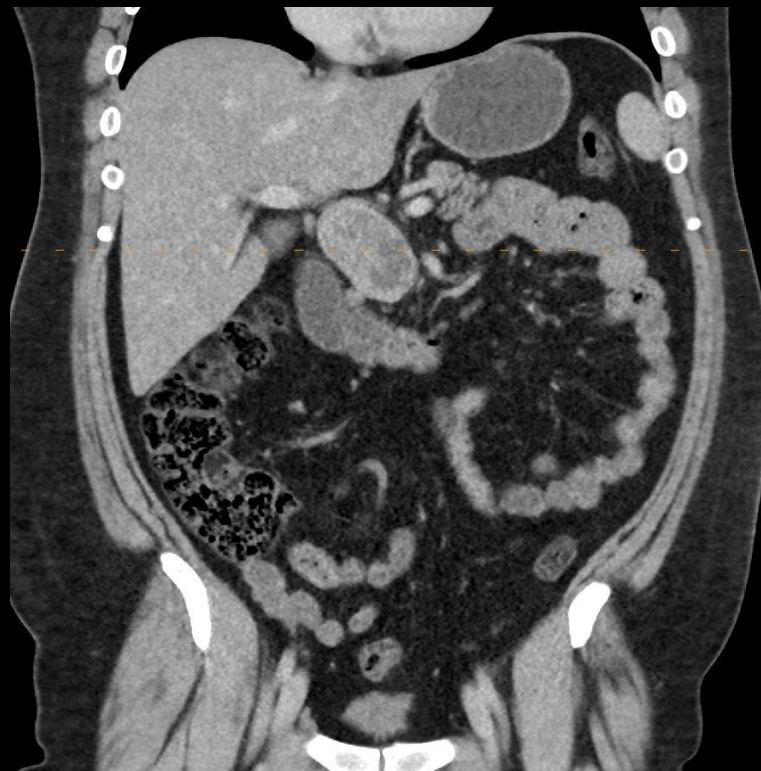
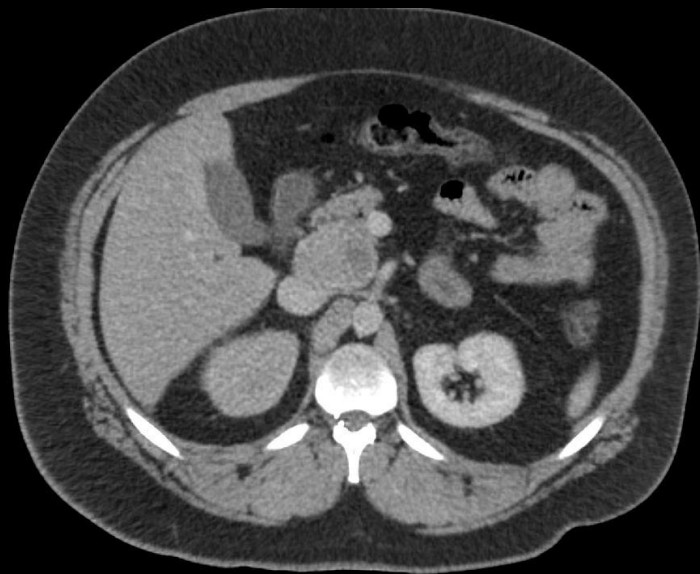
ACR Appropriateness Criteria

American College of Radiology
 ACR Appropriateness Criteria®
 Palpable Abdominal Mass-Suspected Neoplasm

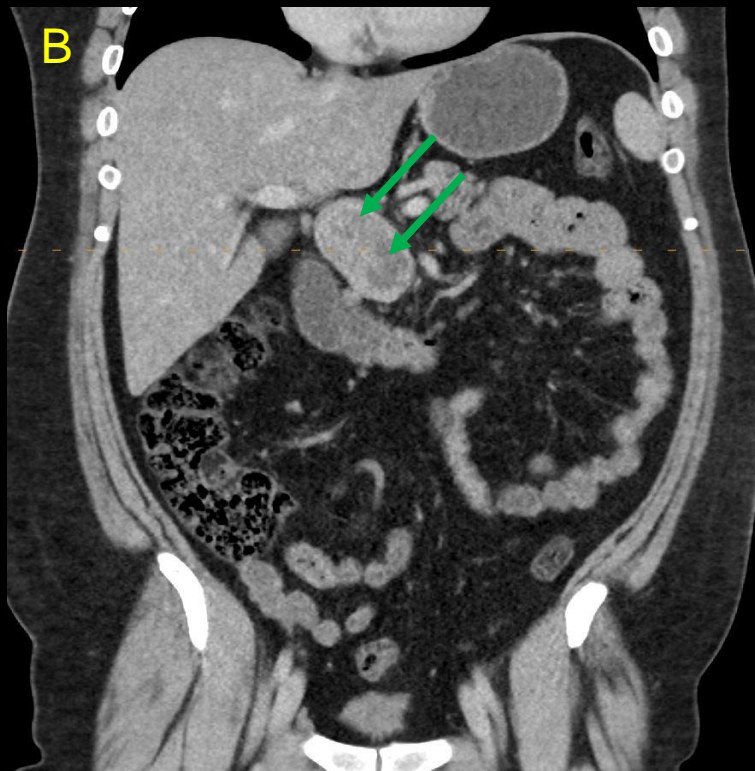
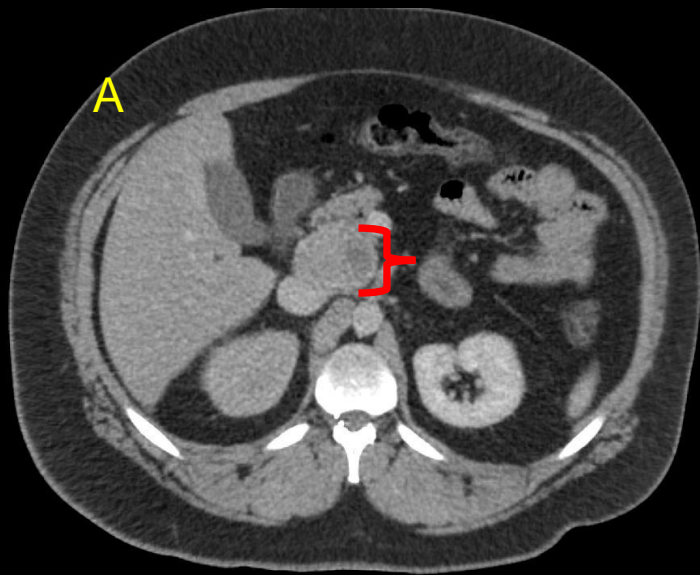
Variant 1: Palpable abdominal mass. Suspected intra-abdominal neoplasm. Initial imaging.

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

Findings: (unlabeled)



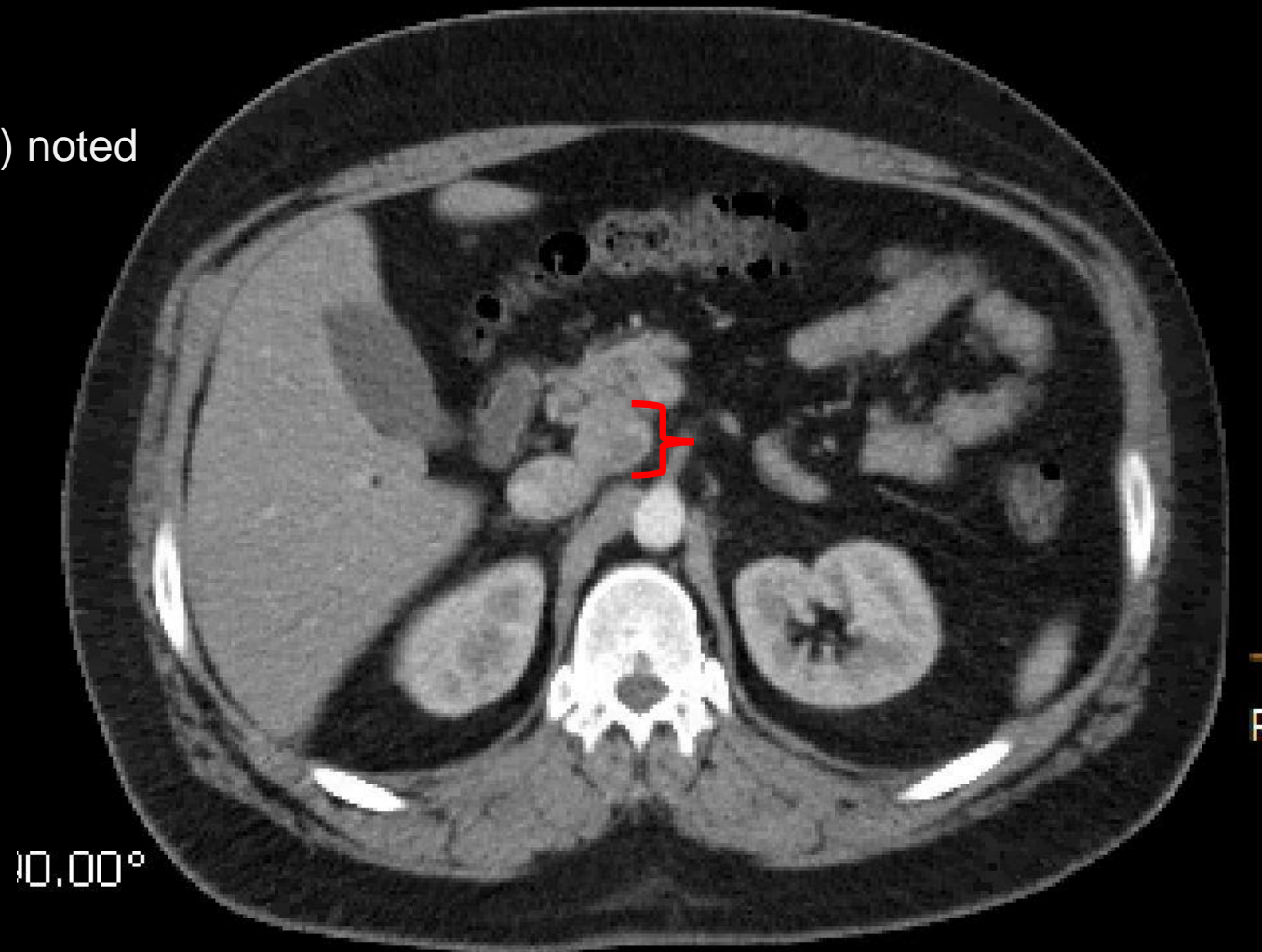
Findings: (labeled)



- Axial CT (A) showing well-defined, 4 cm x 7 cm, heterogeneously enhancing mass in the portocaval region (red bracket)
- Coronal view (B) showing internal hypodense/necrotic areas (green arrows)
- Mild displacement of inferior vena cava (IVC) (blue arrow) on sagittal view (C)
- Mass is separate from right adrenal gland

Findings (4 years prior)

3.5 cm mass (red bracket) noted



DDx (Based on imaging)

- Paraganglioma (PGL)
- Metastatic Tumor
- Lymphoma
- Neuroendocrine Tumor
- Other Retroperitoneal Tumor (Leiomyosarcoma)

Because of concern for pheochromocytoma, the following labs were drawn:

	Ref. Range	6/10/2019 16:05
Epinephrine	Latest Ref Range: 0 - 62 pg/mL	32
Norepinephrine	Latest Ref Range: 0 - 874 pg/mL	1401 (H)
Dopamine, Plasma	Latest Ref Range: 0 - 48 pg/mL	183 (H)

	Ref. Range	6/24/2019 08:30
NORMETAN EPHR., U, 24 H	Latest Ref Range: 82 - 500 ug/24 hr	1292 (H)

	Ref. Range	6/24/2019 08:30
Metanephrine , 24 Hour Urine	Latest Ref Range: 45 - 290 ug/24 hr	153

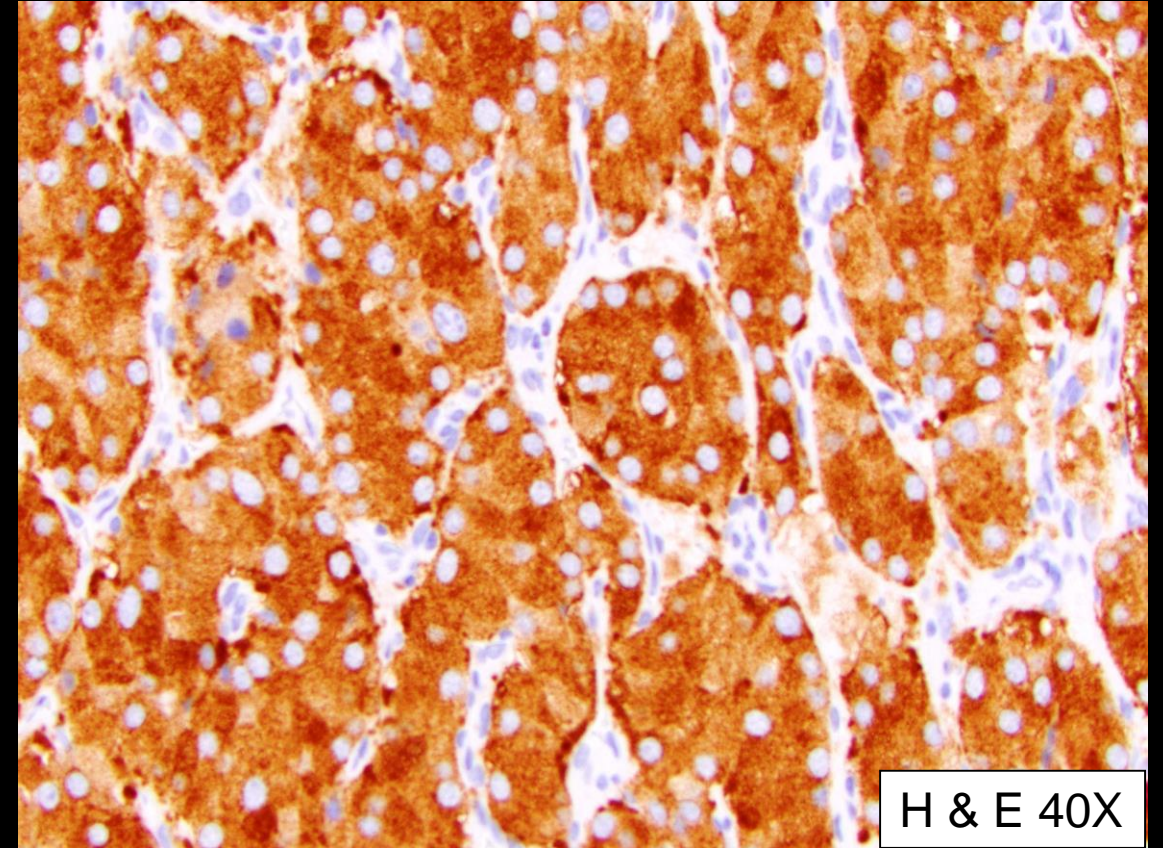
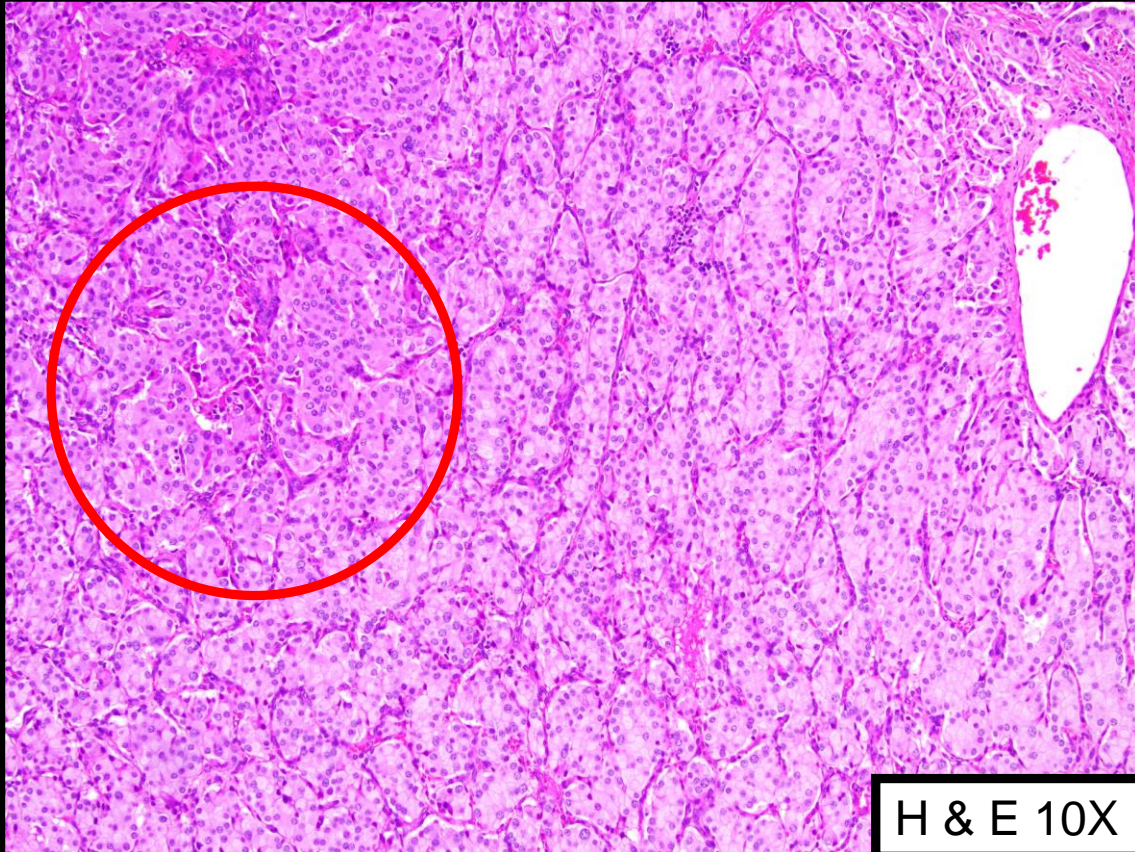
Patient was scheduled for surgery

Gross Pathology:



Retroperitoneal mass measuring 4.2 x 5.3 x 7.0 cm

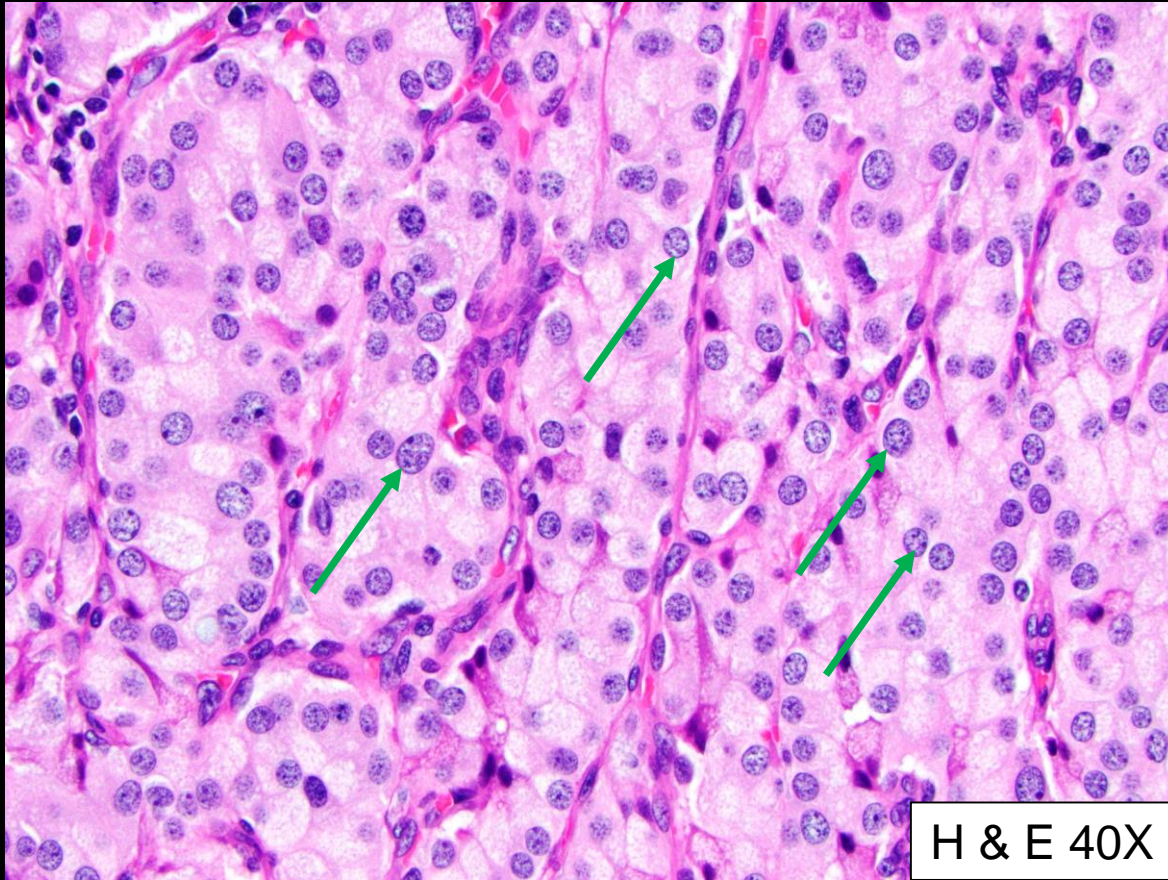
Pathology Findings



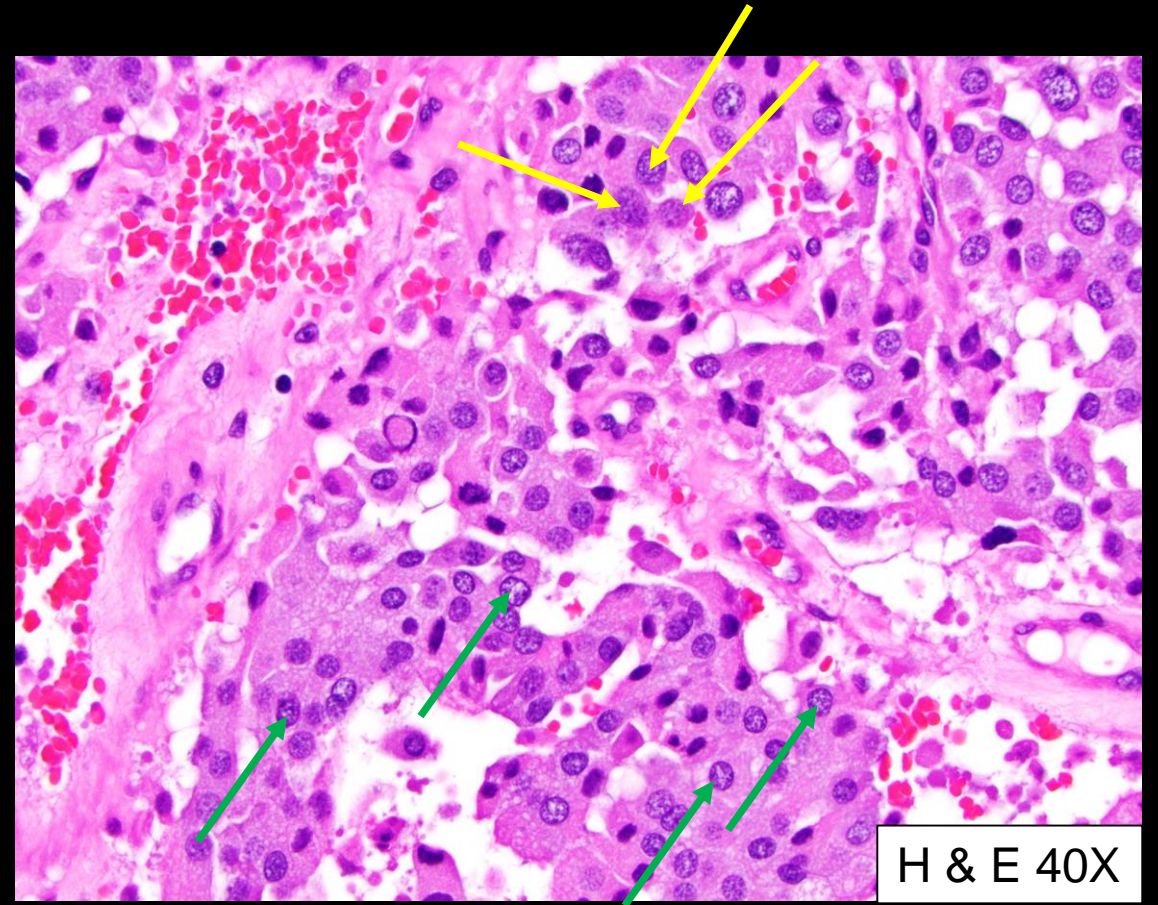
- Tumor cells arranged in solid nests with increased number of mitoses (red circle)
 - Zellballen pattern
 - Diagnostic for PGL/Pheo

- Tumor cells positive for synaptophysin and chromogranin A
 - * Also S100 positive (not shown)

Pathology Findings:



- “Salt & pepper” nuclei (green arrows) representing granular chromatin



- Some nuclear irregularity (small yellow arrows)

Final Dx:

Extra Adrenal Paraganglioma

Case Discussion

- Paragangliomas are rare tumors (benign) that arise from extra-adrenal chromaffin cells
 - Originate from paraganglia at a number of anatomical sites
 - Head, neck, thorax and **abdomen**
- Retroperitoneal paraganglioma represents between 21.5 and 87% of all extra-adrenal paragangliomas
- Characterized by secretions of excessive catecholamines
 - Epinephrine
 - Norepinephrine
 - Dopamine

Case Discussion

- Clinical symptoms include
 - Episodic hypertension
 - Tachycardia
 - Diaphoresis
 - Headache
- 40-50% of paragangliomas are non-functional/potentially functional
 - Misdiagnosis prior to removal problematic
 - Possible sudden release of catecholamines
 - Give alpha blocker prior to surgery (phenoxybenzamine, prazosin)
 - Phentolamine intraoperatively for hypertensive crisis

Case Discussion

- Hereditary Syndrome Associations:
 - Multiple Endocrine Neoplasia Type 2 (MEN-2) → RET mutation
 - von Hippel Lindau → VHL mutation
 - Hereditary PGL/Pheochromocytoma Syndromes → SDHx mutation
 - Neurofibromatosis Type 1 → NF1 mutation (Rare)
- Surgery is treatment of choice
 - Usually curative

References

- Ji XK, Zheng XW, Wu XL, et al. Diagnosis and surgical treatment of retroperitoneal paraganglioma: A single-institution experience of 34 cases. *Oncol Lett*. 2017;14(2):2268–2280. doi:10.3892/ol.2017.6468
- Wen J, Li HZ, Ji ZG, Mao QZ, Shi BB, Yan WG. A decade of clinical experience with extra-adrenal paragangliomas of retroperitoneum: Report of 67 cases and a literature review. *Urol Ann*. 2010;2(1):12–16. doi:10.4103/0974-7796.62919
- Kapetanakis S, Chourmouzi D, Gkardaris G, Katsaridis V, Eleftheriadis E, Givissis P. Functional extra-adrenal paraganglioma of the retroperitoneum giving thoracolumbar spine metastases after a five-year disease-free follow-up: a rare malignant condition with challenging management. *Pan Afr Med J*. 2017;28:94. Published 2017 Sep 29. doi:10.11604/pamj.2017.28.94.13783