

AMSER Rad Path Case of the Month: May 2019

24-year-old woman with new onset hypertension

Anna Zhao, MS3

Harvard Medical School

Shanna Matalon, MD

Department of Radiology

Natalie Rizzo, MD

Department of Pathology

Angela Giardino, MD

Department of Radiology

Brigham & Women's Hospital



Patient Presentation

HPI: 24F G1P0 at 13 weeks of gestation presents with 6 months of **palpitations** and **new hypertension** to 170s/100s

Associated symptoms include lightheadedness, nausea, a flushing sensation, left retrosternal chest pain sometimes triggered by exertion.

1-2 year history of **early satiety** with a 10 lb weight loss over the past month

PMH: **neurofibromatosis type 1**

Medications: none

Social hx: denies tobacco/alcohol use.

Vitals: normal temp, HR, SpO2, RR

Physical exam: unremarkable

Pertinent Workup

- EKG showed normal sinus rhythm, troponins negative
- normal LFTs, no thrombocytopenia, and no proteinuria
- normal TSH
- plasma metanephrine **4.0 nmol/L** (ref: <0.5) and normetanephrine **25 nmol/L** (ref: <0.9)
- 24h urine total metanephrines **18360 mcg/24h** (ref: <1300)
- 24h urine catecholamines: dopamine **1748 mcg/24h** (ref: 65-400), epinephrine **566 mcg/24h** (ref <21), norepinephrine **1424 mcg/24h** (ref: 15-80)

Imaging of Pregnant Patients

- As low as reasonably achievable (ALARA)
- Use alternative exams when appropriate to limit ionizing radiation
 - Deterministic effects: dose threshold must be reached before damage occurs
 - Stochastic effects: no absolute threshold, cancer risk increases with increasing radiation dose
- Medically necessary imaging should not be delayed
- Contrast
 - Iodinated contrast – FDA category B (no adverse effects found in animal studies)
 - Gadolinium – FDA category C (adverse effects found in animal studies)

Fetal Radiation Dose

Type of Examination	Fetal Dose* (mGy)
Very low-dose examinations (<0.1 mGy)	
Cervical spine radiography (anteroposterior and lateral views)	<0.001
Radiography of any extremity	<0.001
Mammography (two views)	0.001–0.01
Chest radiography (two views)	0.0005–0.01
Low- to moderate-dose examinations (0.1–10 mGy)	
Radiography	
Abdominal radiography	0.1–3.0
Lumbar spine radiography	1.0–10
Intravenous pyelography	5–10
Double-contrast barium enema	1.0–20
CT	
Head or neck CT	1.0–10
Chest CT or CT pulmonary angiography	0.01–0.66
Limited CT pelvimetry (single axial section through the femoral heads)	<1
Nuclear medicine	
Low-dose perfusion scintigraphy	0.1–0.5
Technetium-99m bone scintigraphy	4–5
Pulmonary digital subtraction angiography	0.5
Higher-dose examinations (10–50 mGy)	
Abdominal CT	1.3–35
Pelvic CT	10–50
¹⁸ F PET/CT whole-body scintigraphy	10–50



Potential Effects of Radiation Exposure

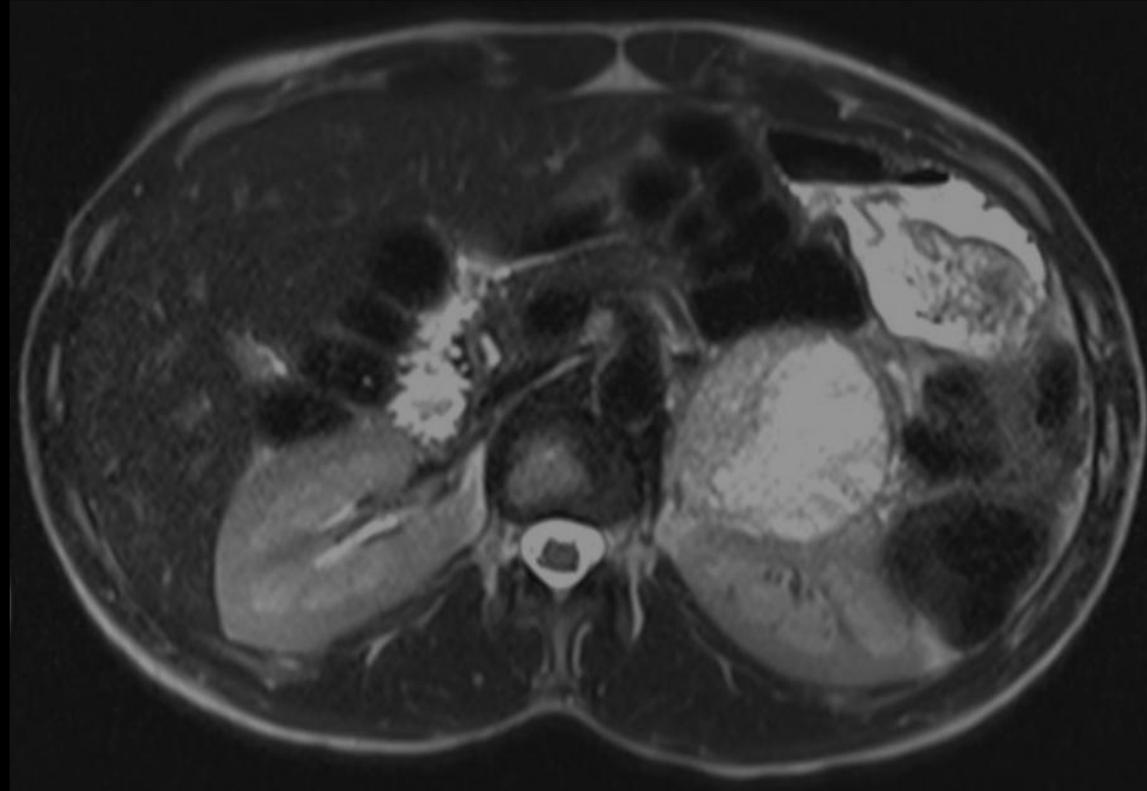
Table 1: Summary of Suspected In Utero Induced Deterministic Radiation Effects*[4,5]

Menstrual or Gestational age	Conception age	<50 mGy (<5 rad)	50–100 mGy (5–10 rad)	>100 mGy (>10 rad)
0–2 weeks (0–14 days)	Prior to conception	None	None	None
3rd and 4th weeks (15–28 days)	1st–2nd weeks (1–14 days)	None	Probably none	Possible spontaneous abortion.
5th–10th weeks (29–70 days)	3rd–8th weeks (15–56 days)	None	Potential effects are scientifically uncertain and probably too subtle to be clinically detectable.	Possible malformations increasing in likelihood as dose increases.
11th–17th weeks (71–119 days)	9th–15th weeks (57–105 days)	None	Potential effects are scientifically uncertain and probably too subtle to be clinically detectable.	Risk of diminished IQ or of mental retardation, increasing in frequency and severity with increasing dose.
18th–27th weeks (120–189 days)	16th–25th weeks (106–175 days)	None	None	IQ deficits not detectable at diagnostic doses.
>27 weeks (>189 days)	>25 weeks (>175 days)	None	None	None applicable to diagnostic medicine.



**Stochastic risks are suspected, but data are not consistent [6]. For exposure to a newborn child, the lifetime attributable risk of developing cancer is estimated to be 0.4% per 10 mGy (1 rad) dose to the baby. The potential risks in utero for the second and third trimesters and part of the first trimester may be comparable, but the uncertainties in this estimate are considerable.*

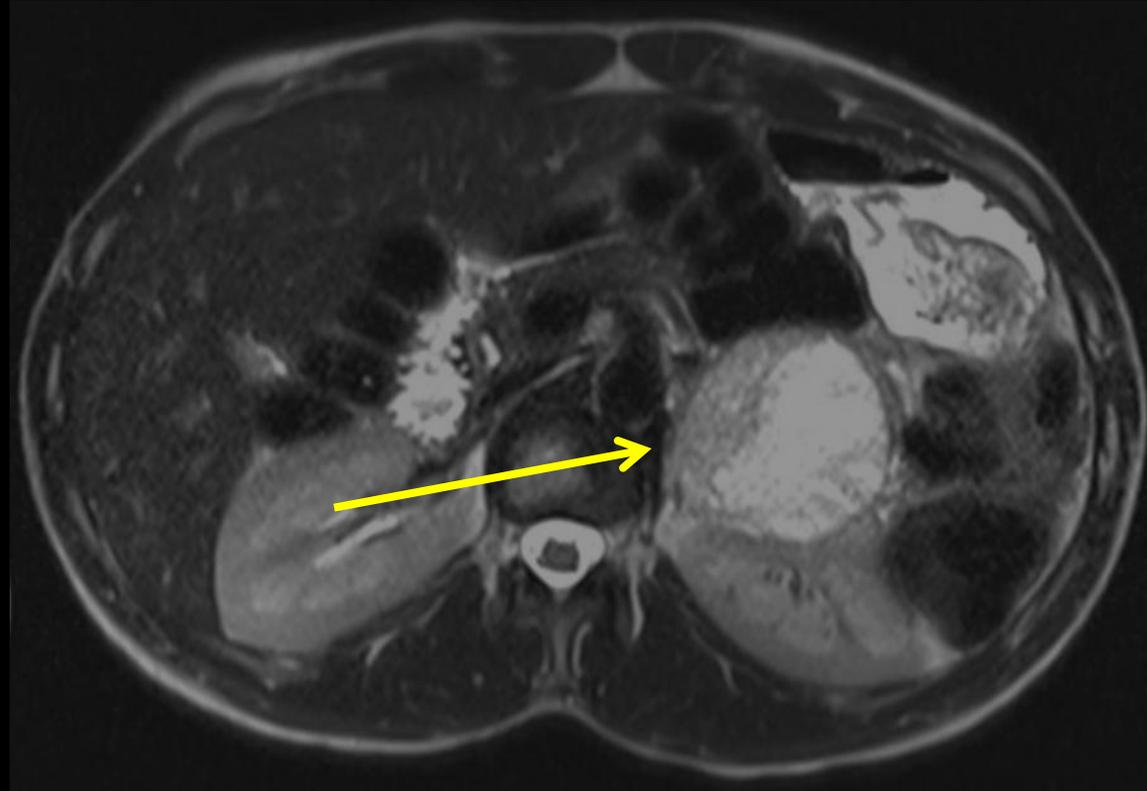
Non-Contrast Axial MRI



T2

Non-Contrast Axial MRI

5.5 cm T2
hyperintense left
adrenal mass

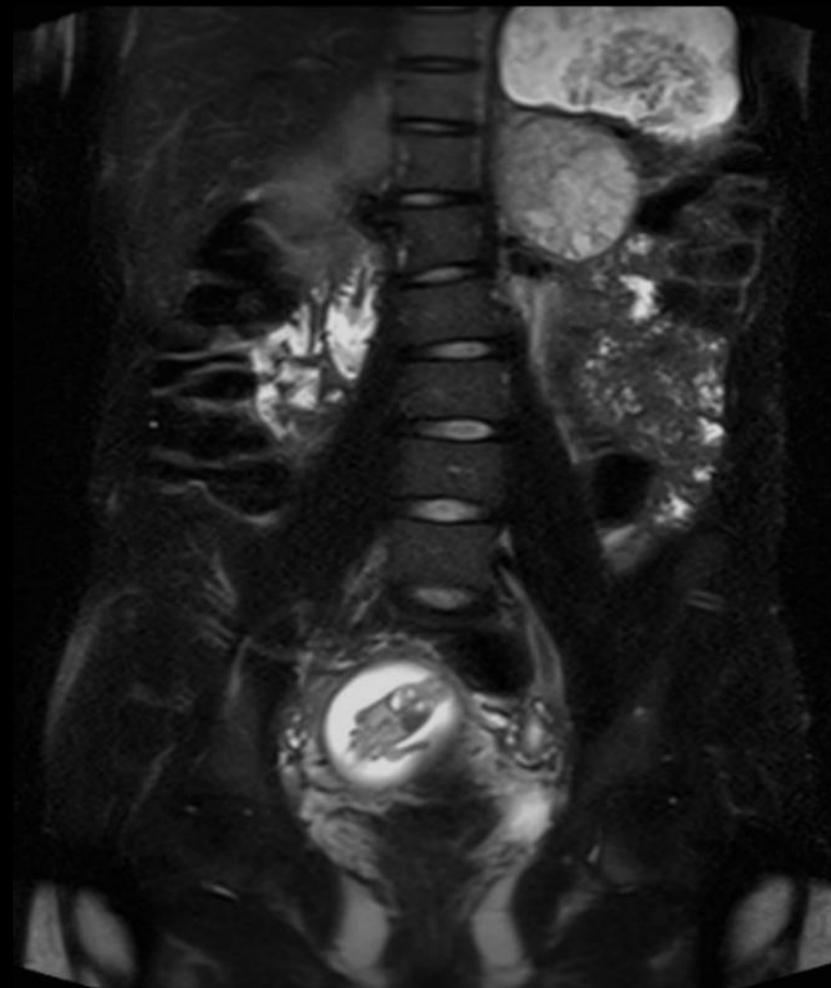


T2

Non-contrast Coronal MRI

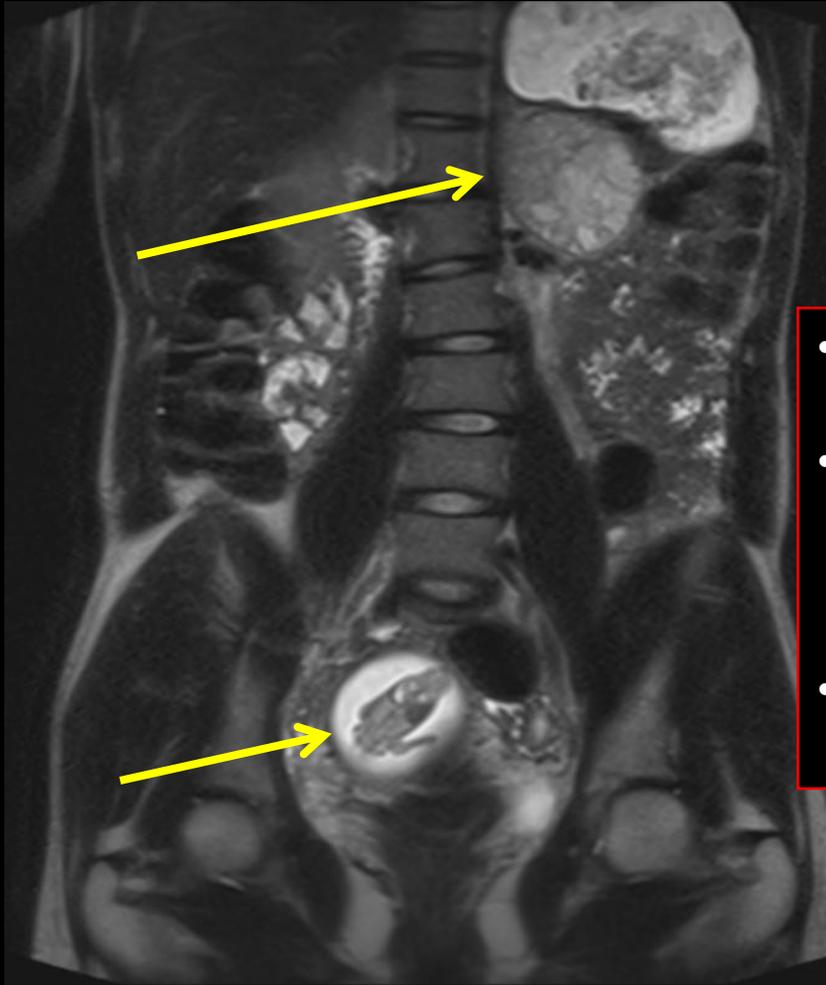


T2



T2 Fat Sat

Non-contrast Coronal MRI



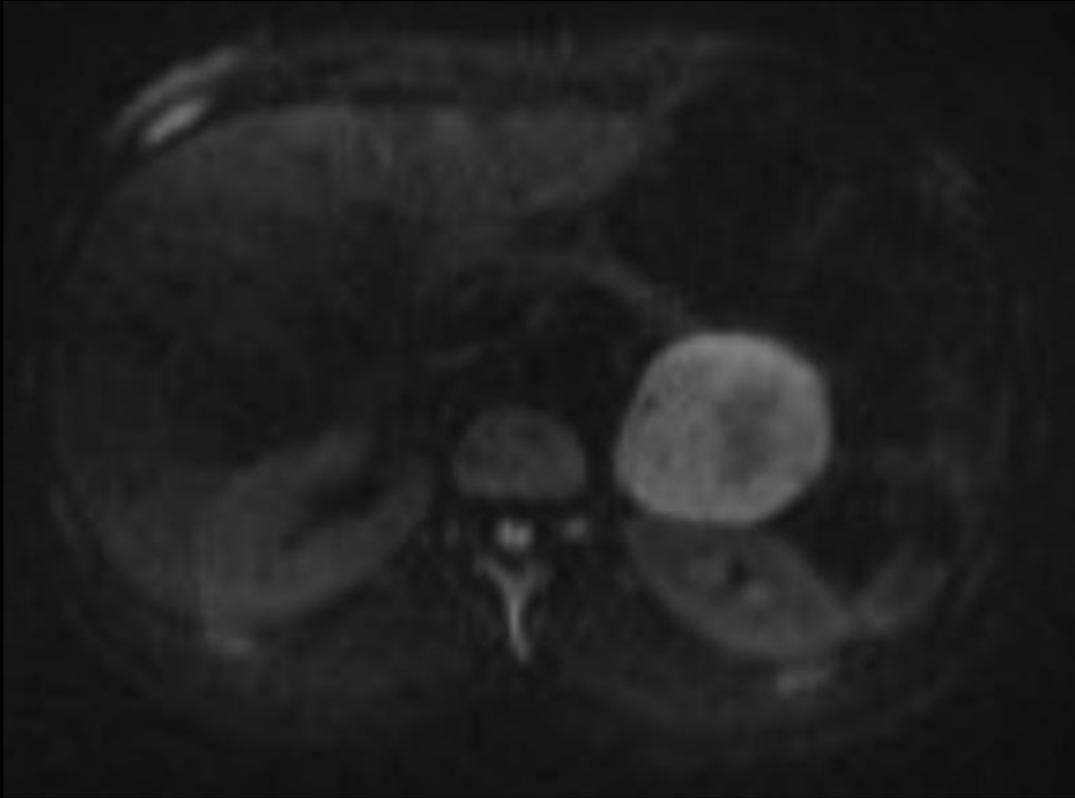
T2

- 5.5 cm T2 hyperintense left adrenal mass
- No fat suppression on T2 fat saturation indicates there is no macroscopic fat
- Single intrauterine gestation is visualized

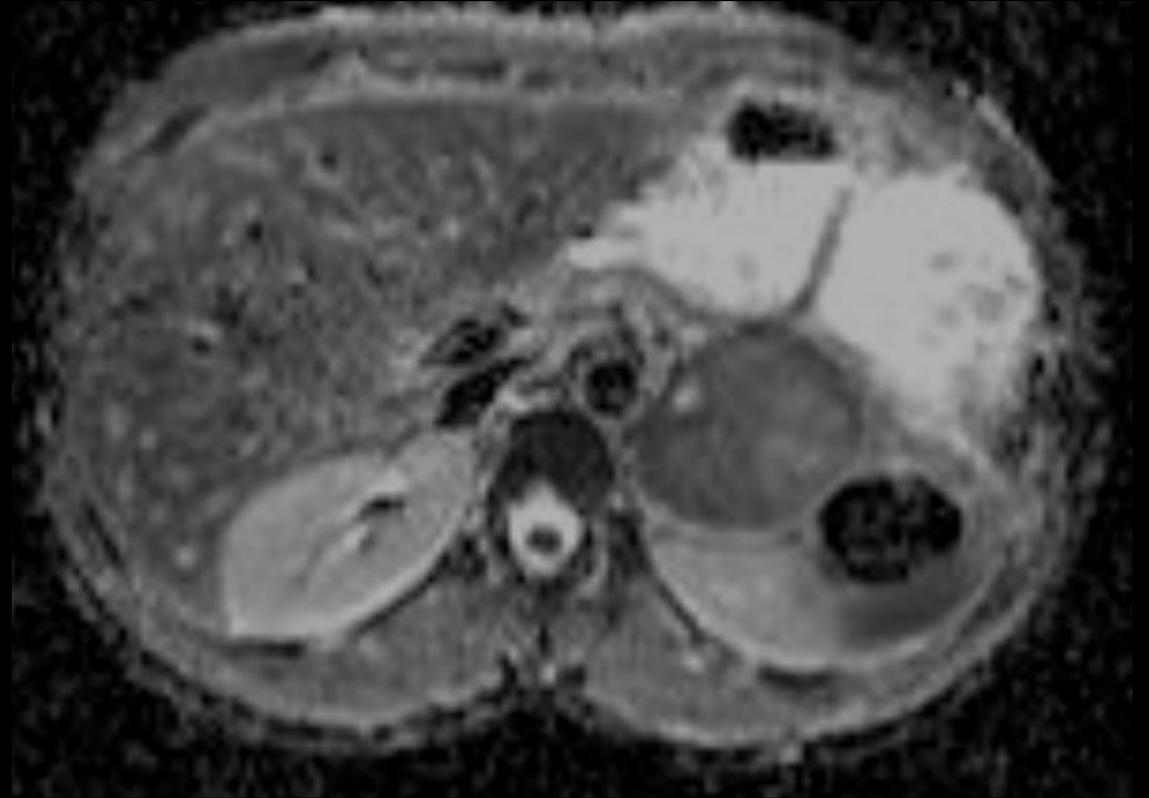


T2 Fat Sat

Axial Diffusion Weighted Imaging



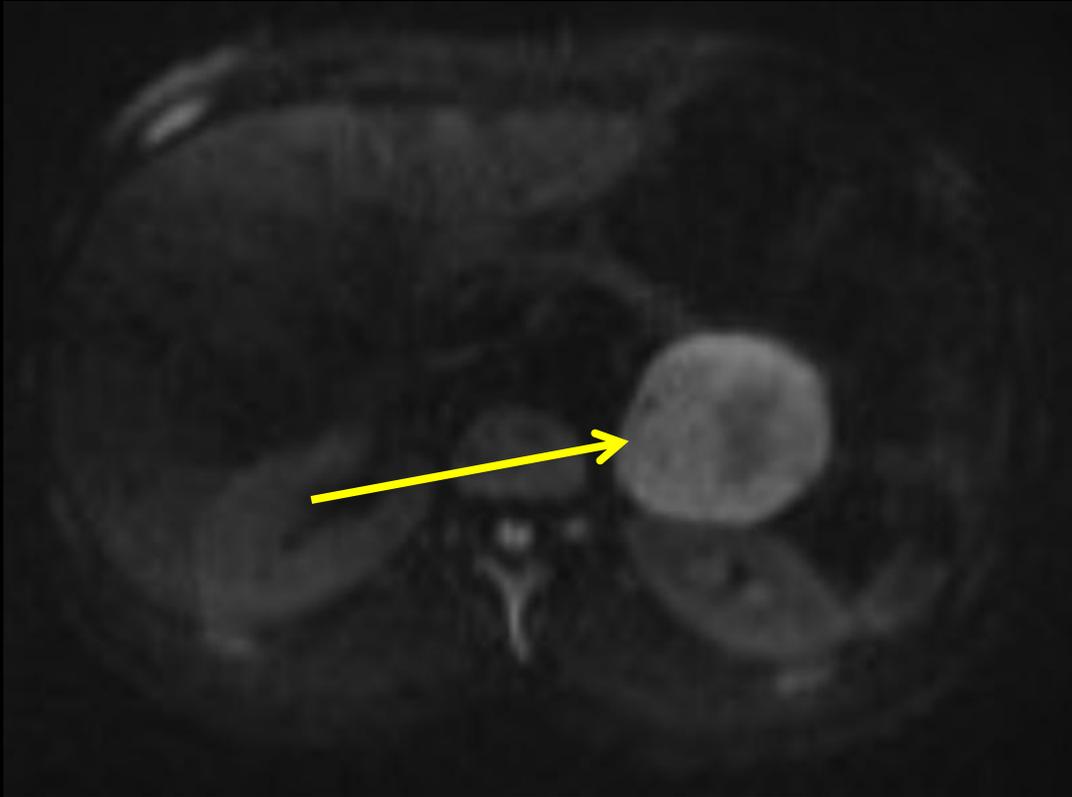
DWI



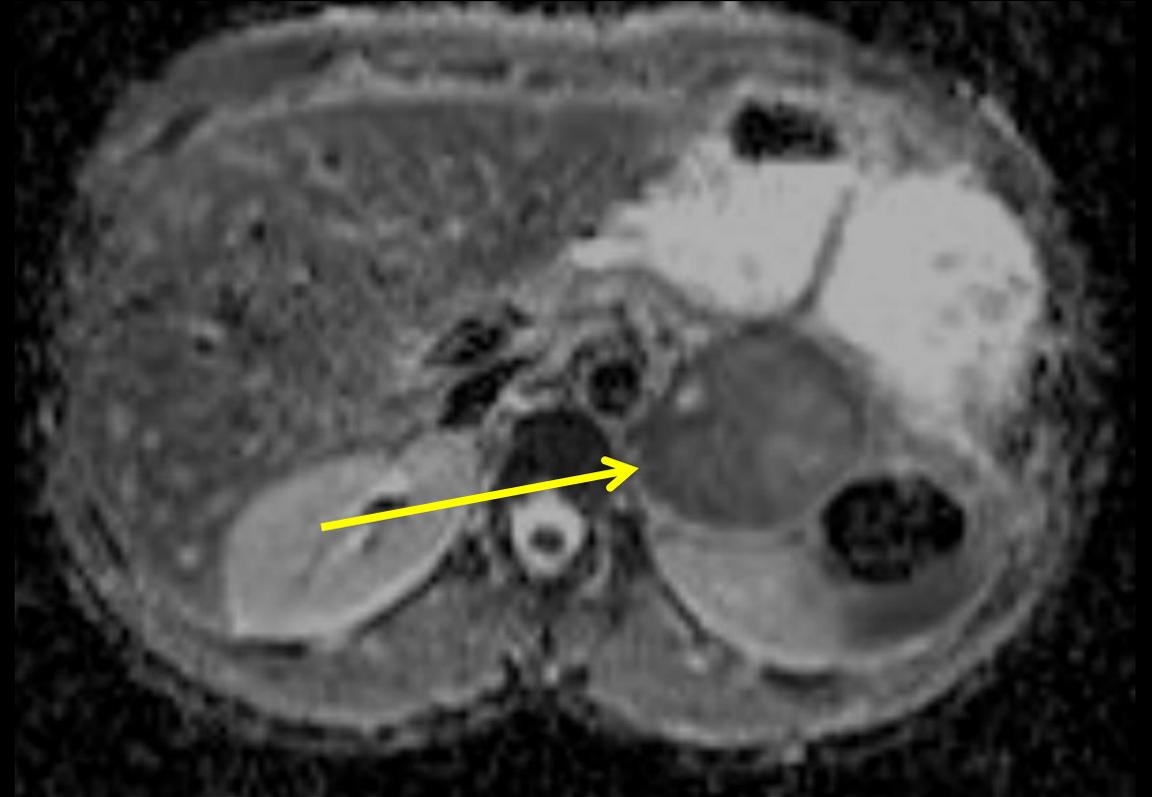
ADC

Axial Diffusion Weighted Imaging

The mass demonstrates restricted diffusion, with increased signal on DWI and decreased signal on ADC



DWI

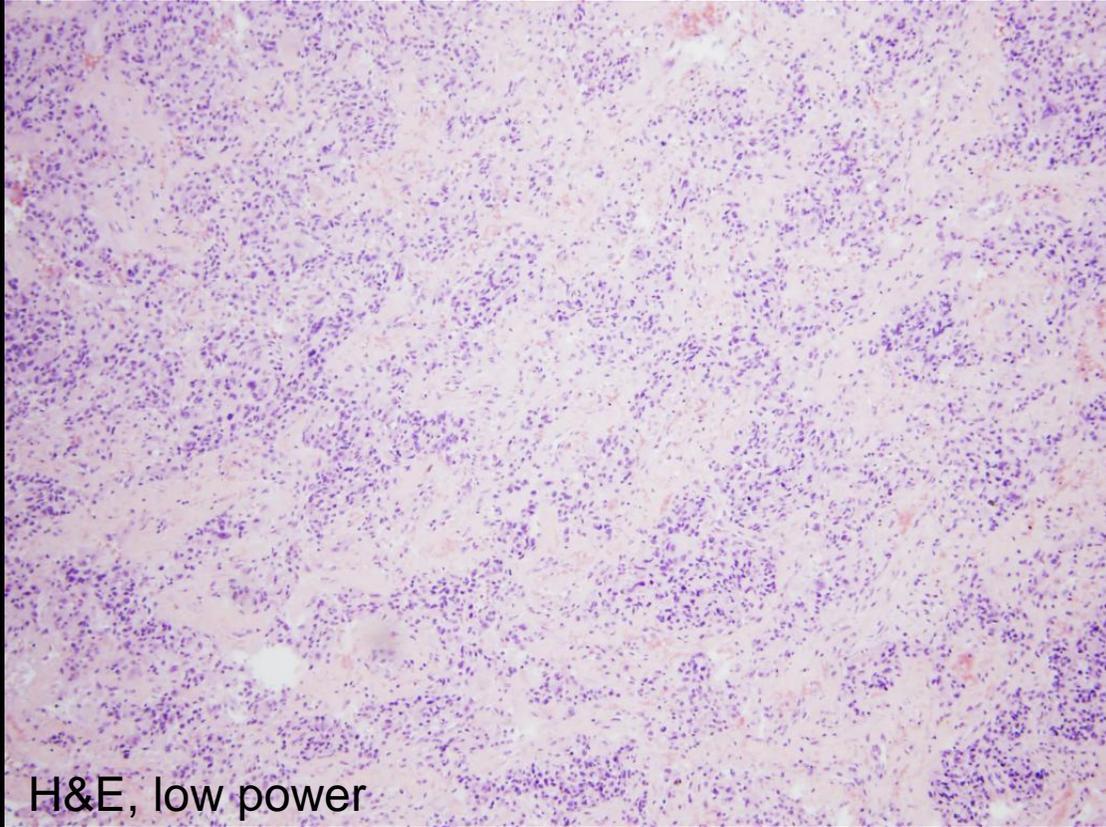


ADC

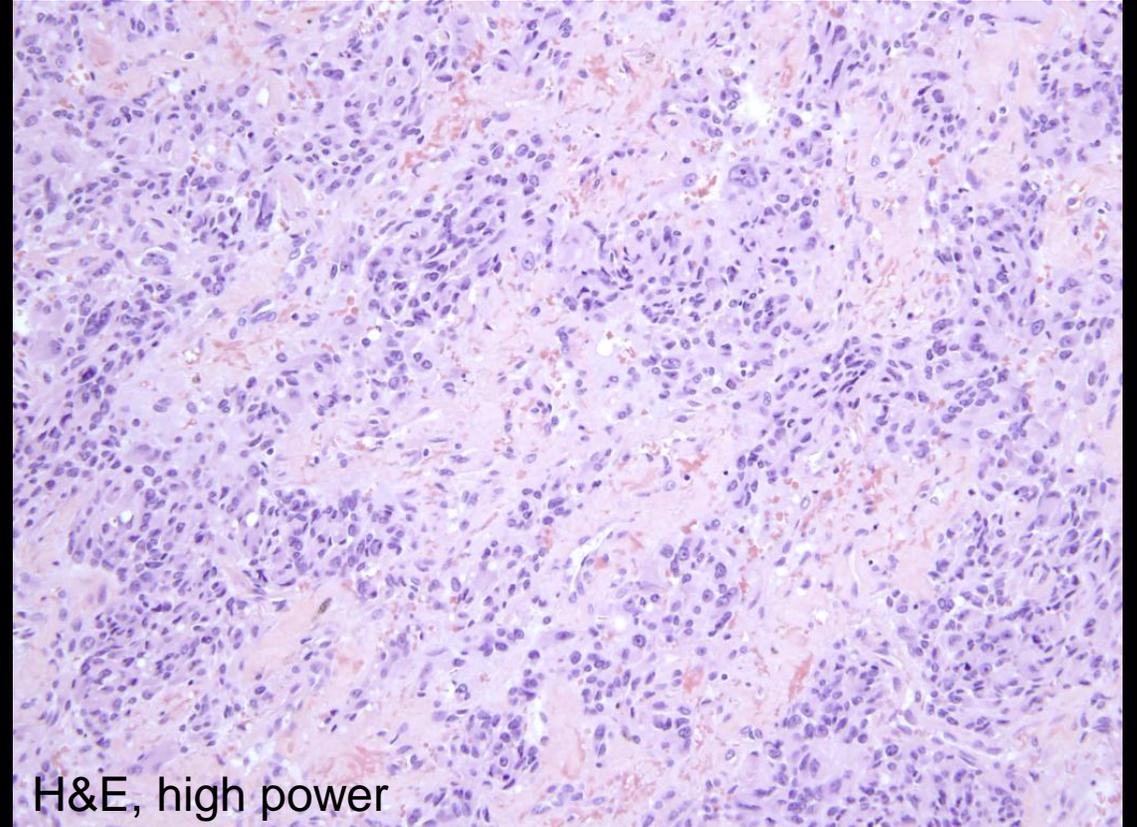
DDX for Adrenal Mass

- Adenoma
- Primary adrenocortical carcinoma
- Metastases
- Myelolipoma
- Pheochromocytoma
- Hemorrhage
- Cyst

Pathology

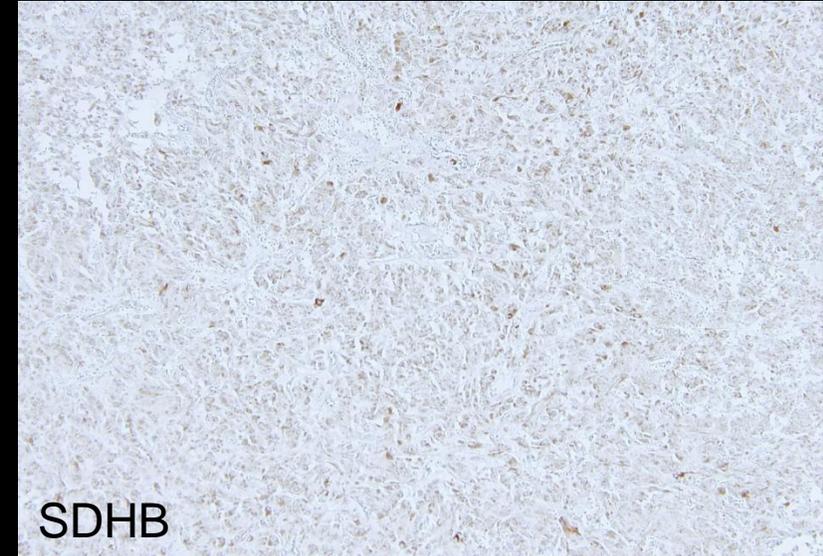
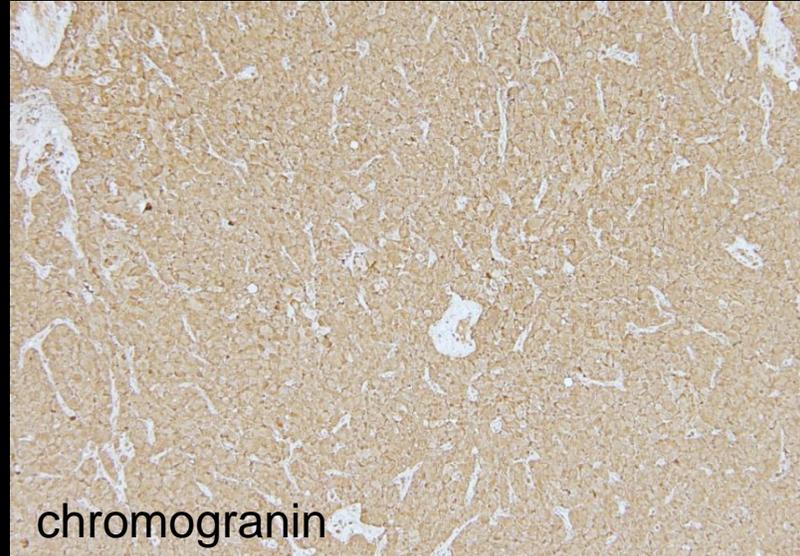
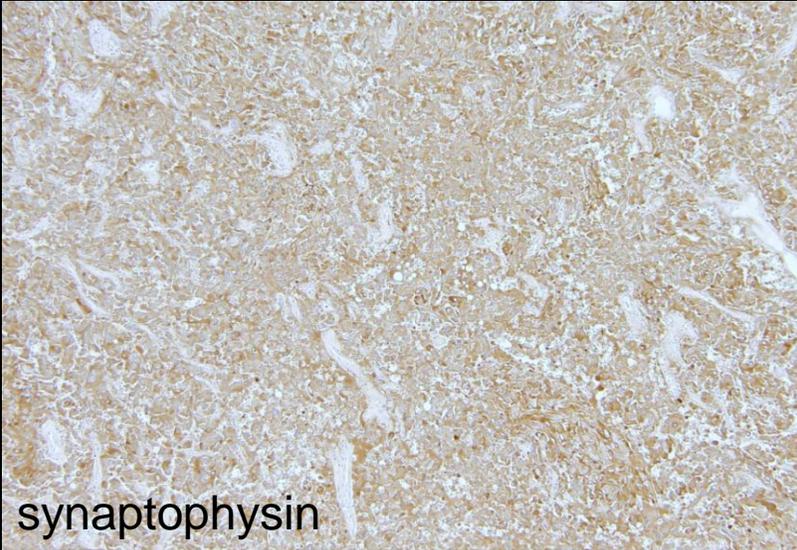


Disorganized nests of cells in classic “zellballen” appearance, with intervening vasculature



Nuclear pleomorphism, increased nuclear:cytoplasmic ratio, prominent nucleoli

Pathology



Positive immunohistochemical staining for synaptophysin and chromogranin, which is characteristic of pheochromocytoma

SDHB expression is retained (wild type). SDHB mutations are associated with hereditary pheochromocytoma/paraganglioma syndromes.

Final Dx:

Adrenal Pheochromocytoma

Case Discussion: Pheochromocytoma

- catecholamine-secreting tumor arising from chromaffin cells
- presents with classic triad of headache, sweating, and tachycardia
- increased catecholamines and metanephrines found in urine and plasma, labs should be obtained prior to imaging
- CT or MRI will detect most symptomatic pheochromocytomas (typically 3-5 cm)
- classic MRI appearance is a T2 hyperintense mass
- ± hemorrhage, necrosis, calcification

Case Discussion: Pheochromocytoma

“Rule of 10’s”

- 10% are extra-adrenal
 - i.e. paraganglioma
 - organ of Zuckerkandl (aortic bifurcation), bladder (may present with post-micturation syncope), head and neck
- 10% are bilateral
- 10% are malignant
- 10% are associated with a genetic syndrome
 - Multiple endocrine neoplasia (MEN) 2A and 2B
 - von Hippel–Lindau
 - Neurofibromatosis type 1

References

- American College of Radiology. ACR practice guideline for imaging pregnant or potentially pregnant adolescents and women with ionizing radiation. Reston, VA: American College of Radiology, 2008.
- Tremblay E (2012). “Quality Initiatives: Guidelines for Use of Medical Imaging during Pregnancy and Lactation,” *Radiographics*; 32(3):897-911.
- Mandel J (2013). *Core Radiology: A Visual Approach to Diagnostic Imaging 1st Edition*. New York, NY: Cambridge University Press.
- Young WF (2018). “Clinical presentation and diagnosis of pheochromocytoma.” In: *UpToDate*, Post TW (Ed), UpToDate, Waltham, MA.
- Young WF (2018). “Pheochromocytoma in genetic disorders.” In: *UpToDate*, Post TW (Ed), UpToDate, Waltham, MA.