

Best Cases of the AIRP

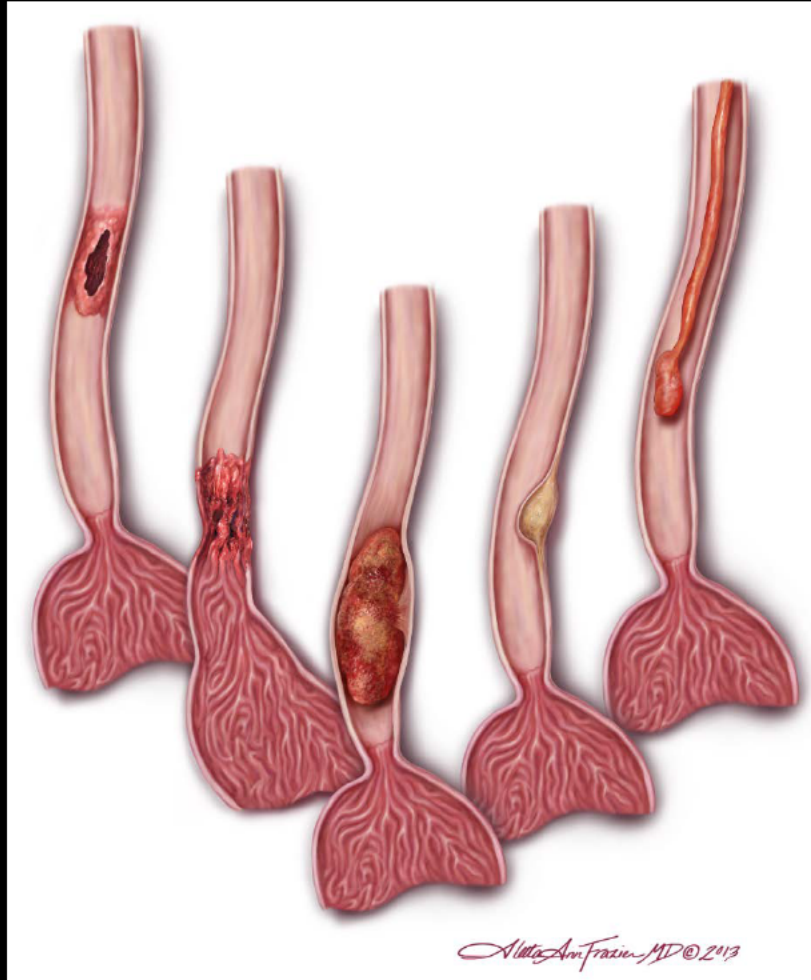
02 2018

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Gastrointestinal Best Case



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Alta AmFrazin MD © 2013

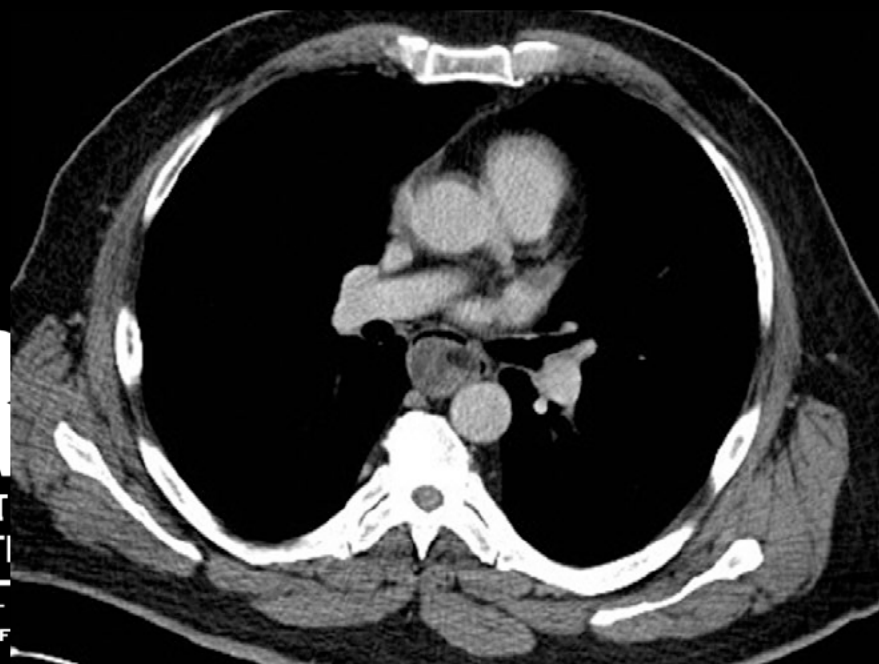
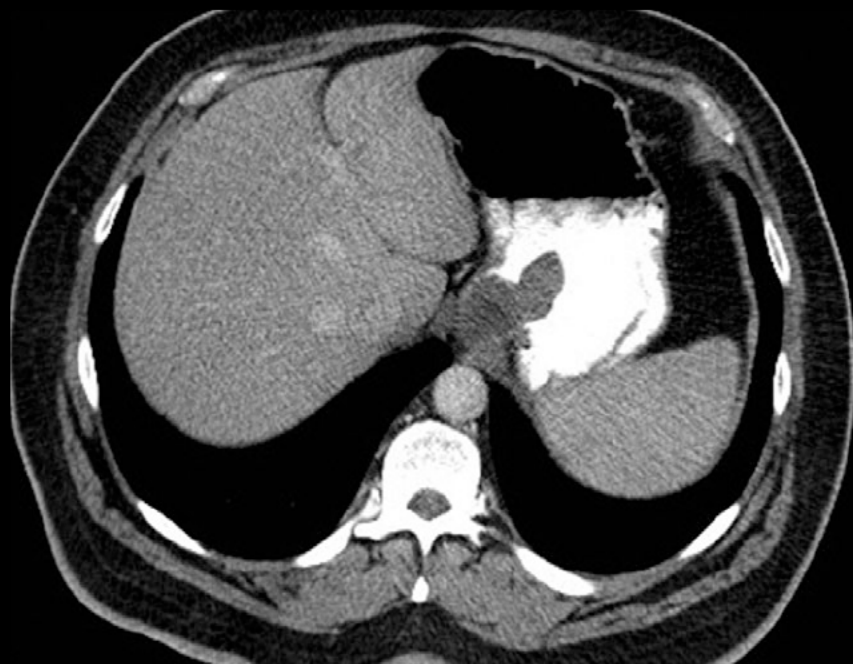
Clinical information

51-year-old white man presents with a vague one year history of mild dysphagia, anemia, weight loss, and abdominal pain.



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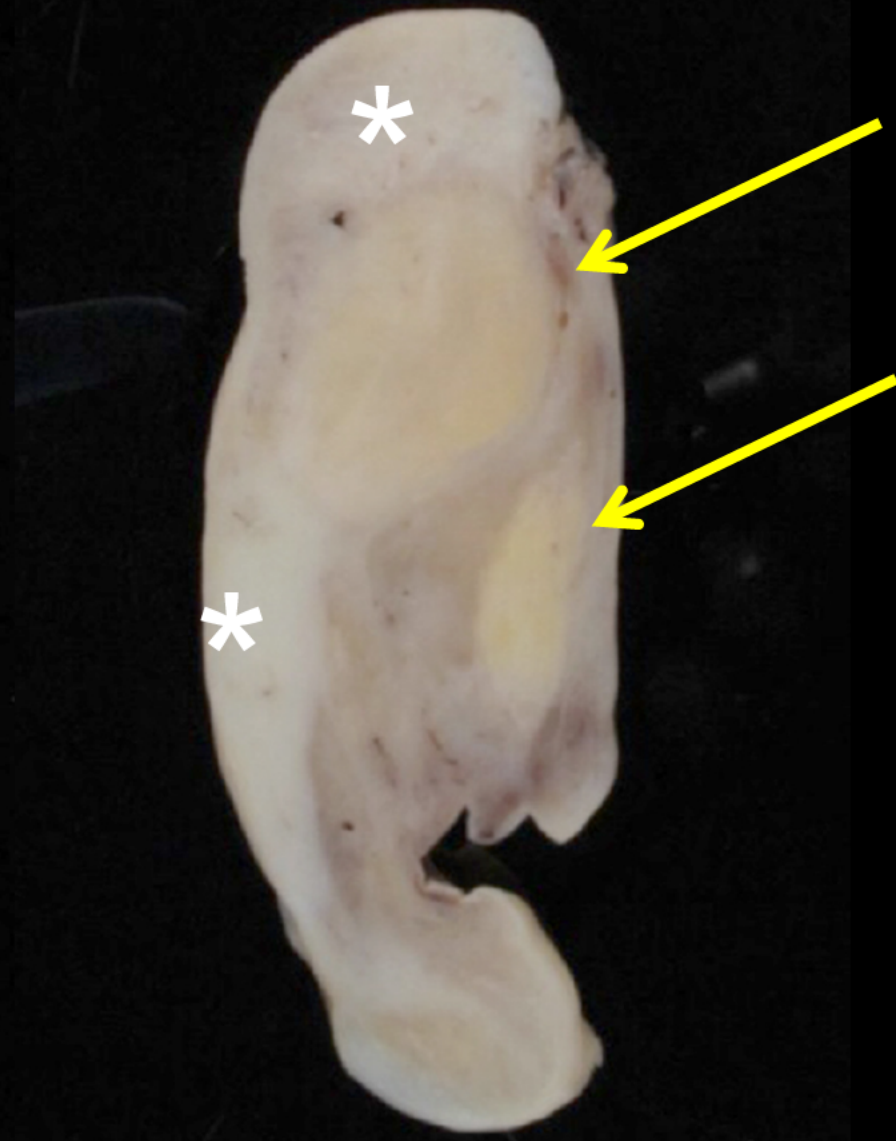
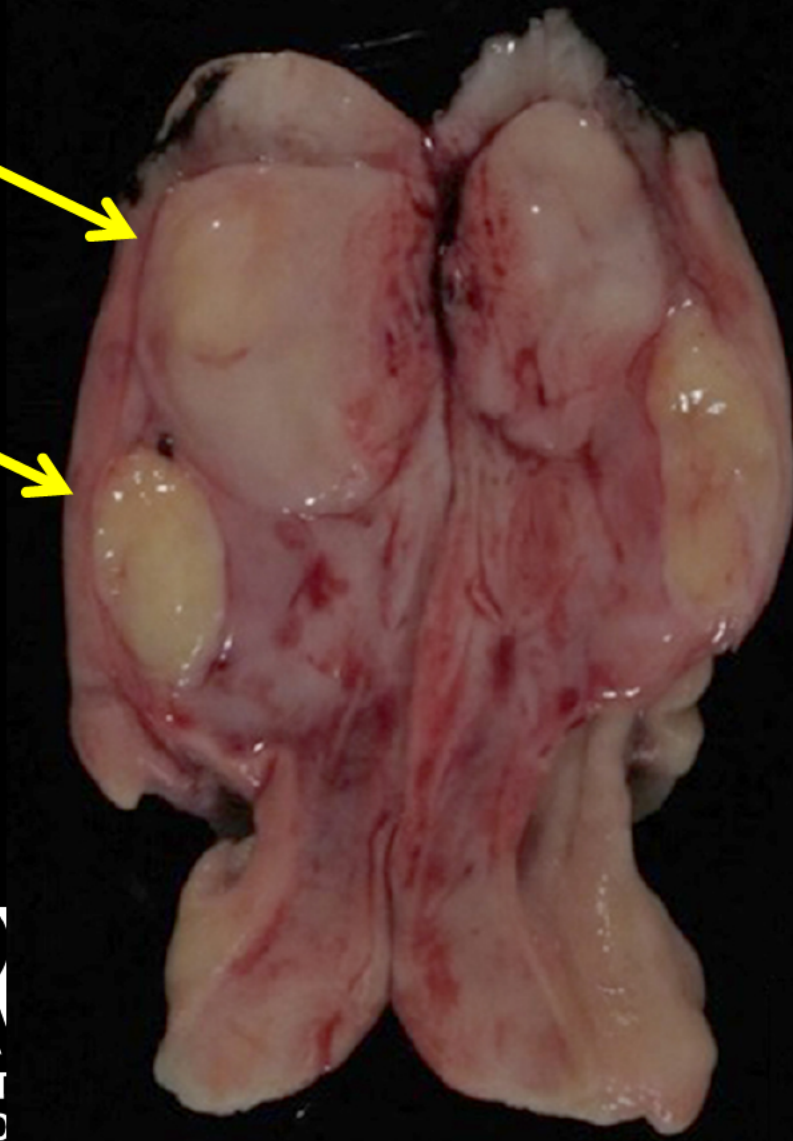


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Perim 12.0 mm
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Avg -59.38 HU
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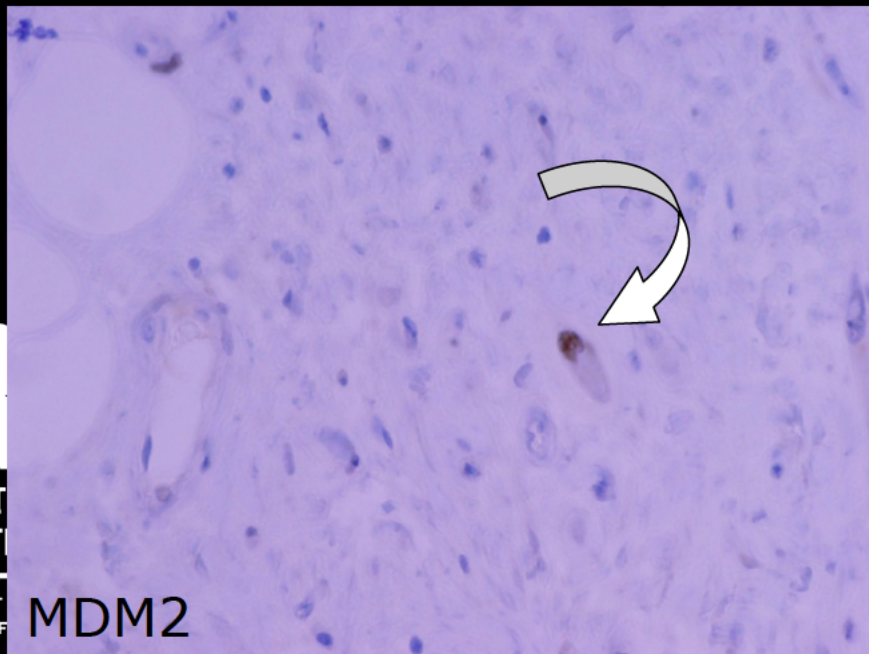
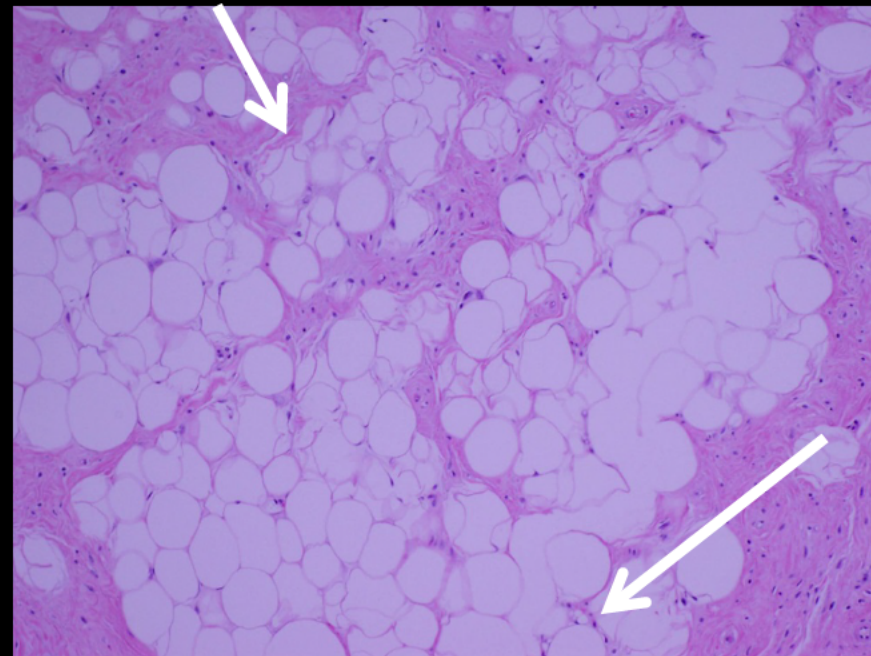
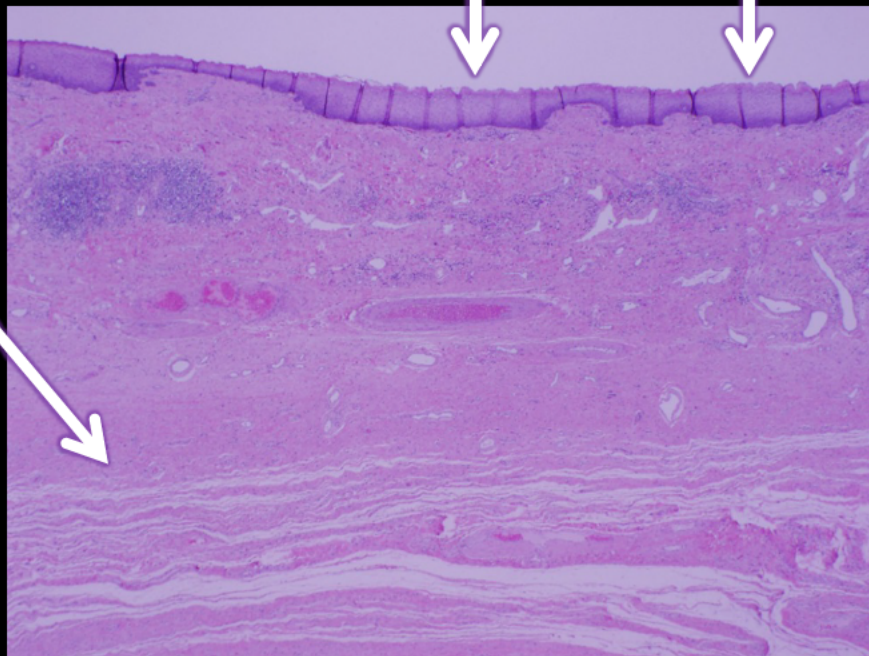
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Well-differentiated liposarcoma/Atypical lipomatous tumor in a giant fibrovascular polyp

Kathleen Eddy

Victoria General Hospital

Halifax, Nova Scotia, Canada

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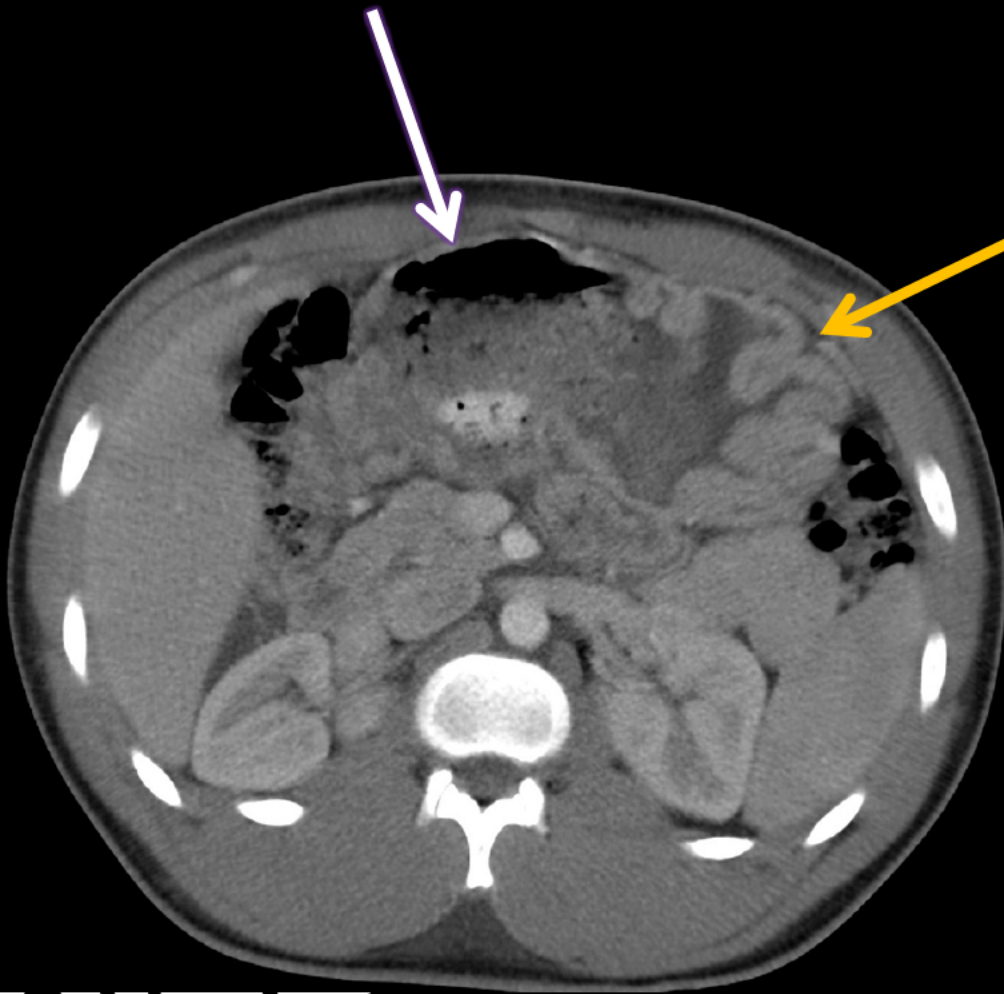
Honorable mention



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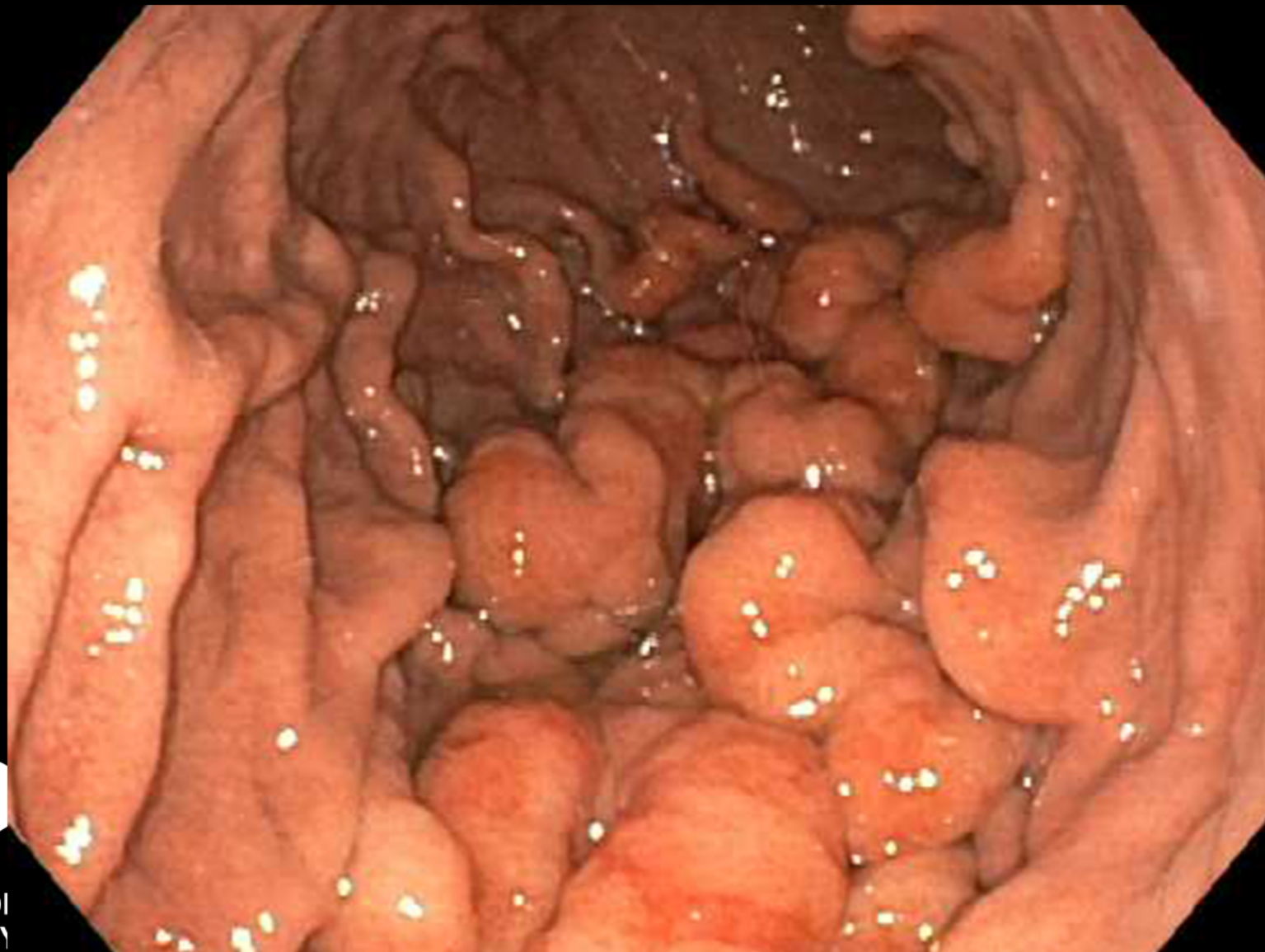
Clinical information

26-year-old man with six days of acute bilateral pitting edema with labs showing long standing hypoalbuminemia.



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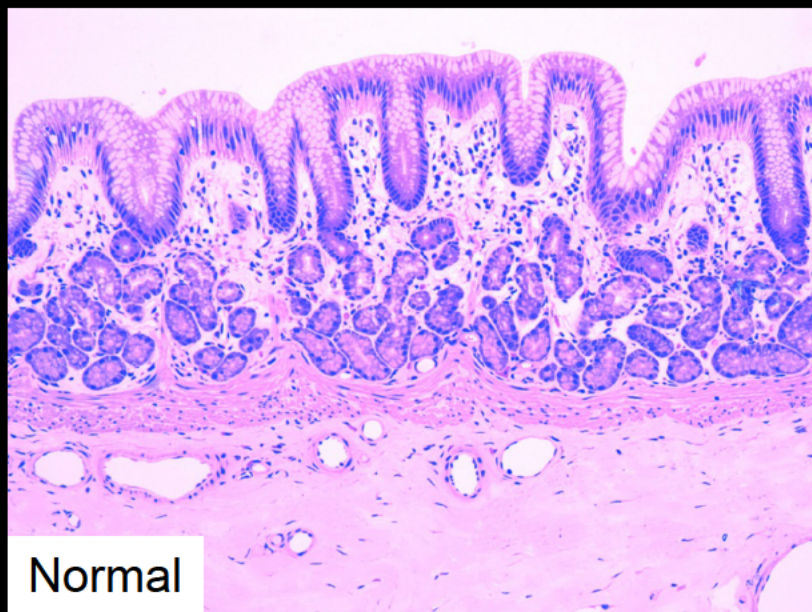
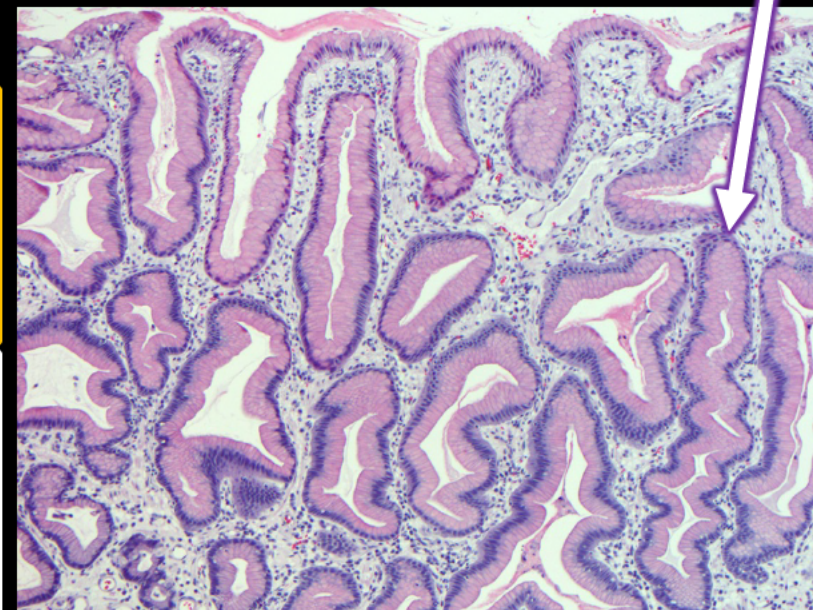
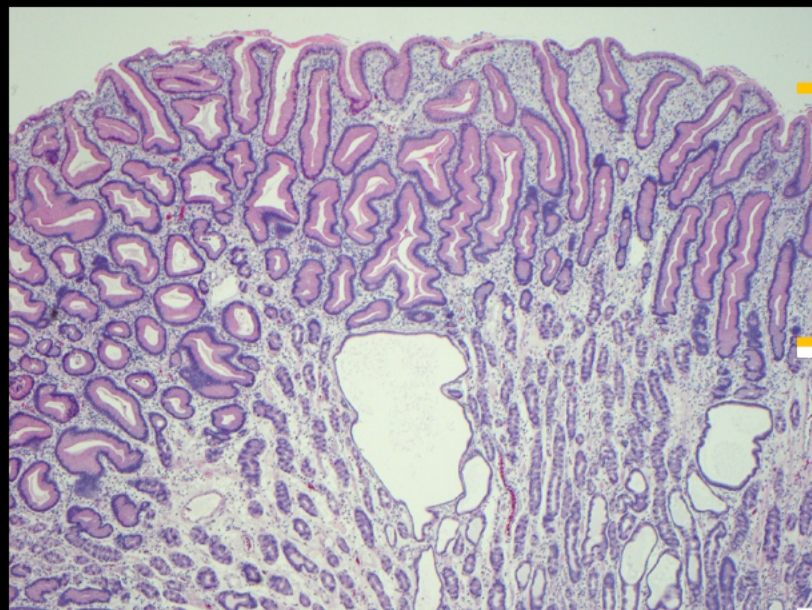
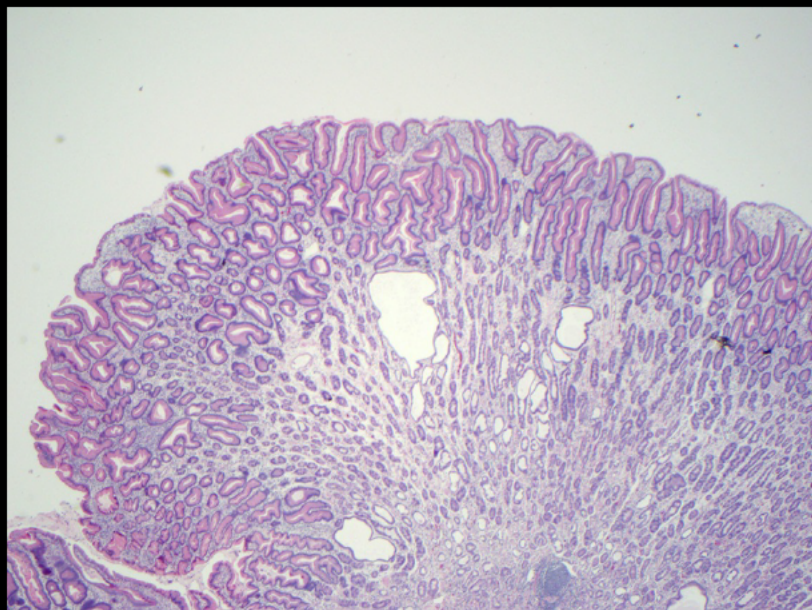
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Normal

Foveolar glands – mucin producing

Oxyntic glands – acid producing

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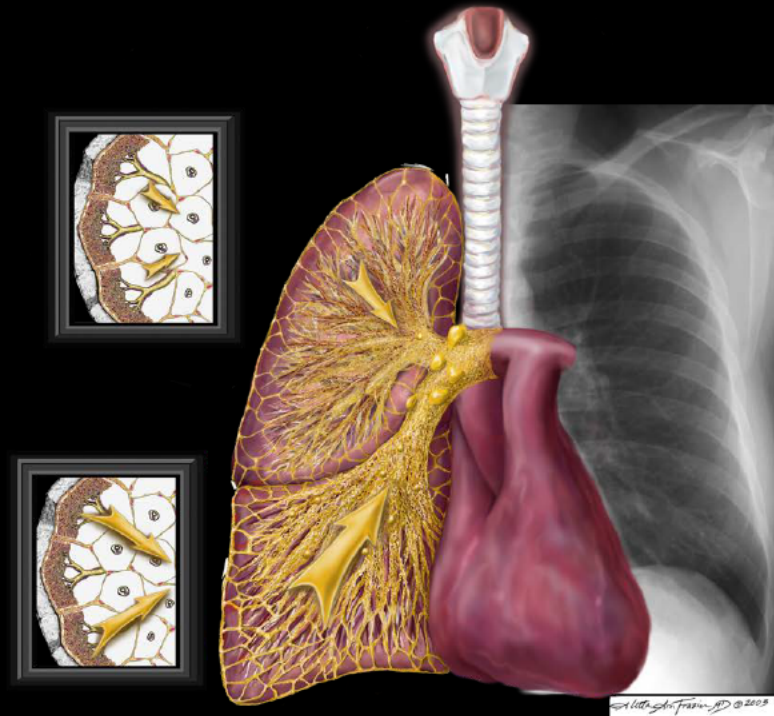
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Menetrier disease

Nicholas DiGeorge

**Naval Medical Center Portsmouth
Portsmouth, Virginia**

Pulmonary and Mediastinal Best Case



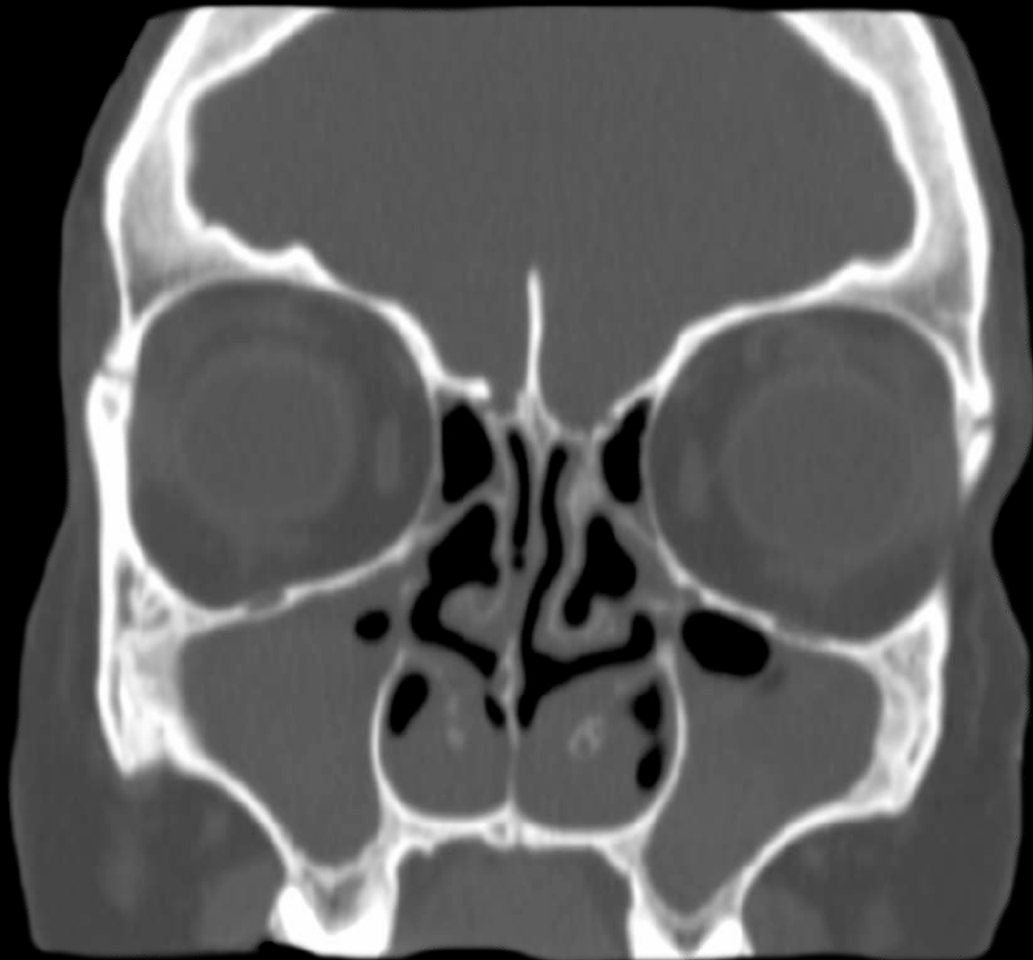
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- **47 year old female with a history of chronic cough with recurrent episodes of bronchitis and sinus infections**

Sinus disease



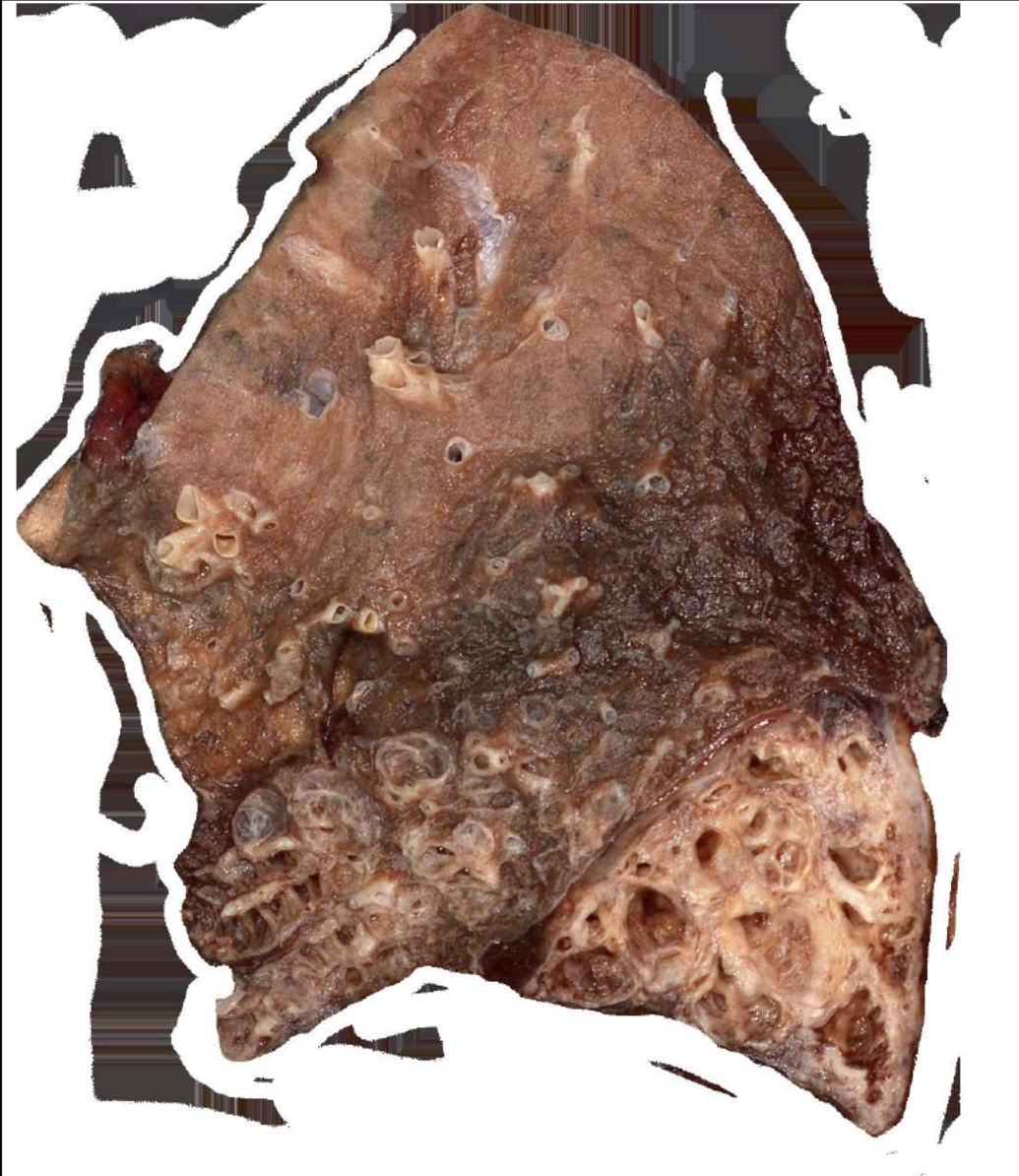
Situs inversus and lower lobe bronchiectasis



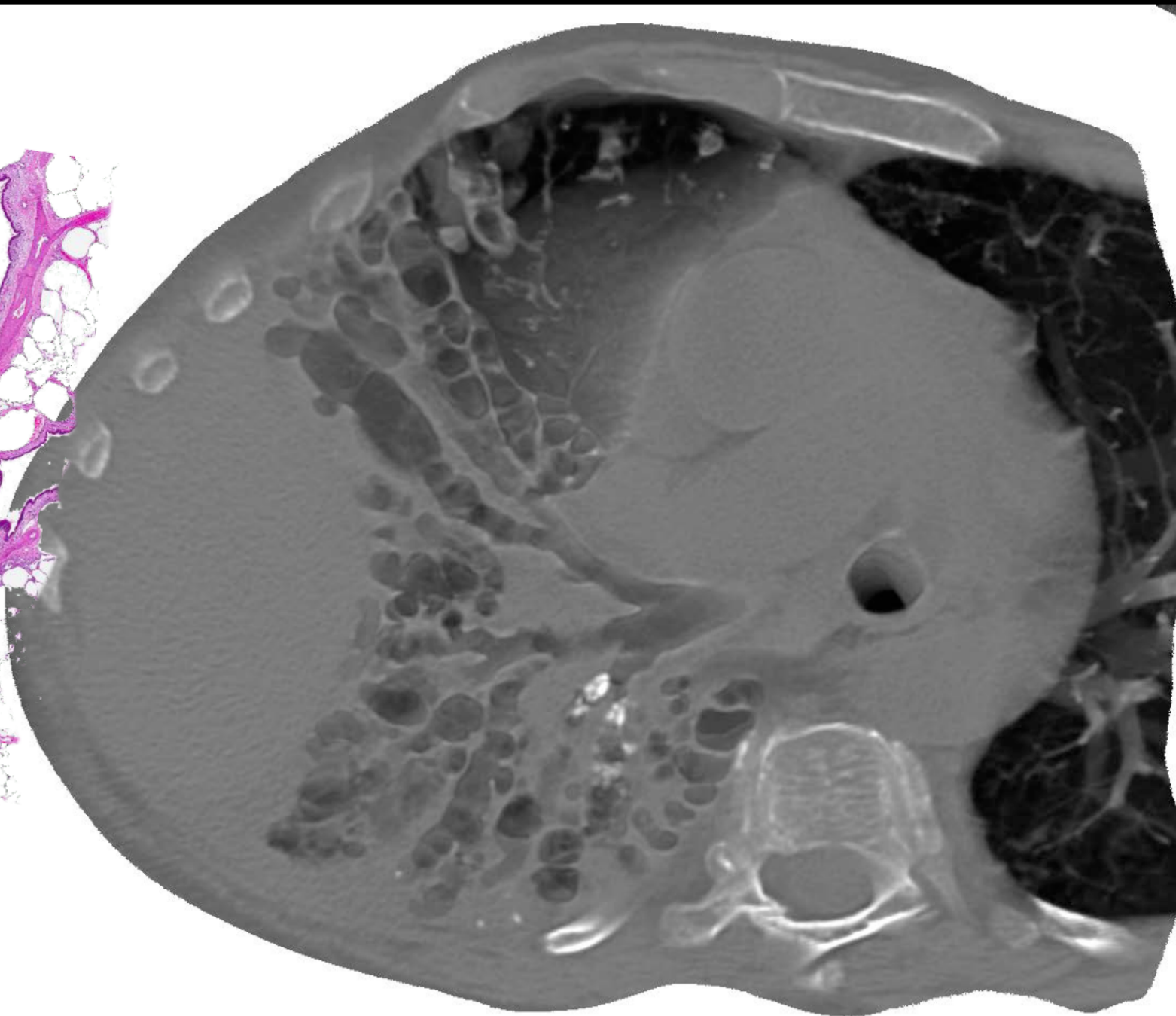
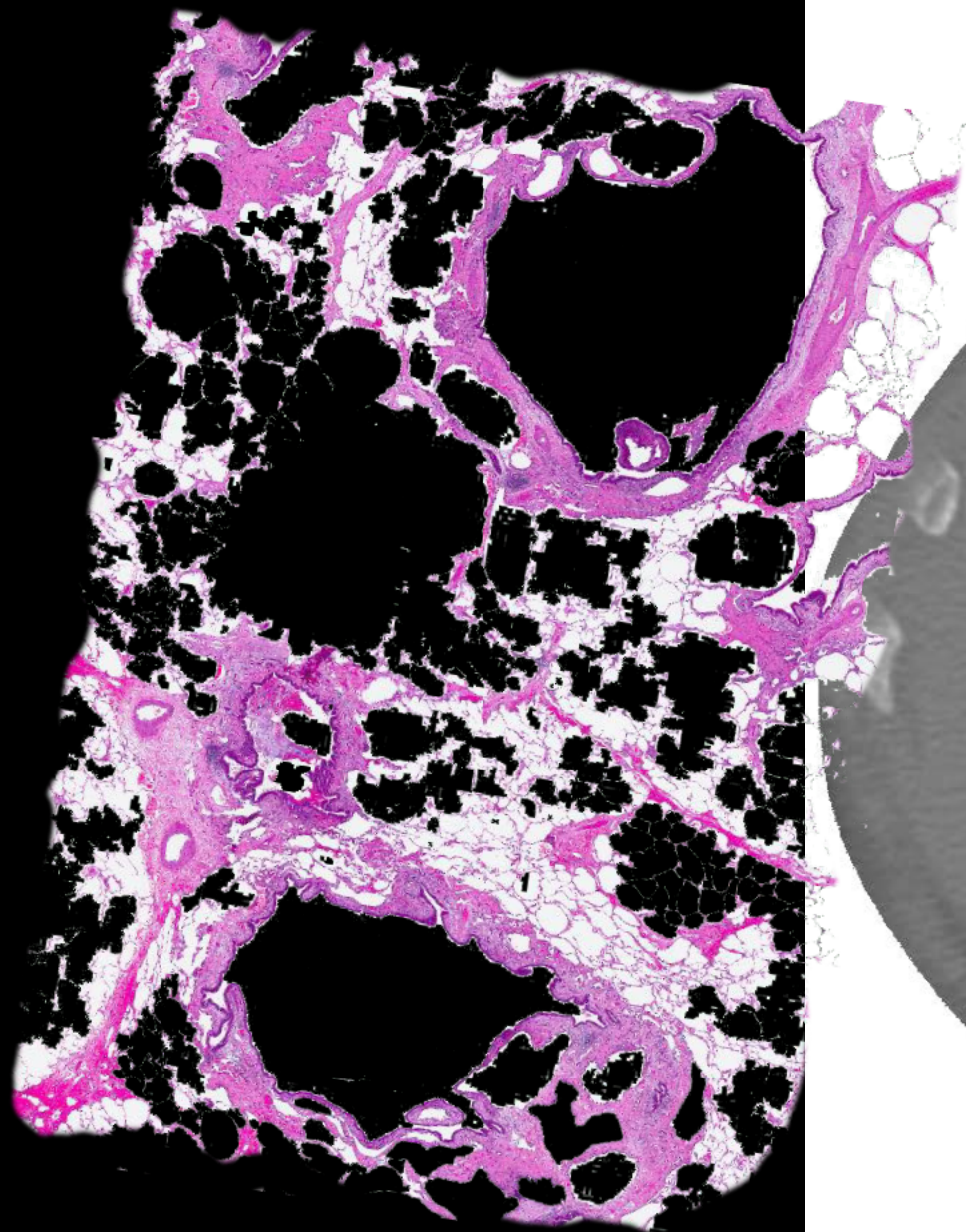
Situs inversus and lower lobe bronchiectasis



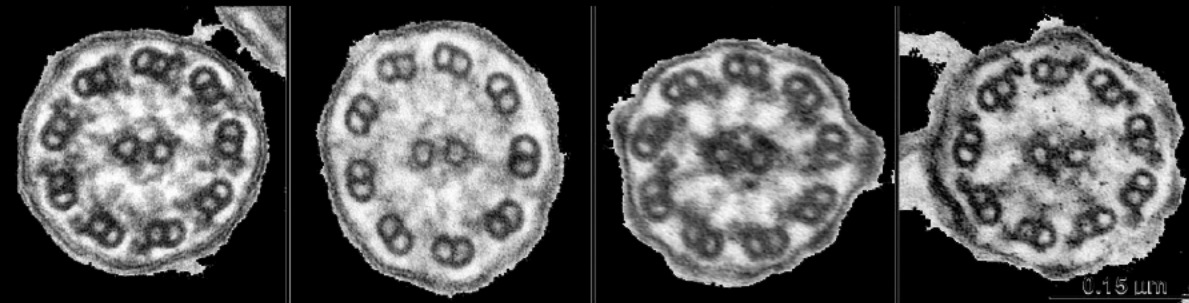
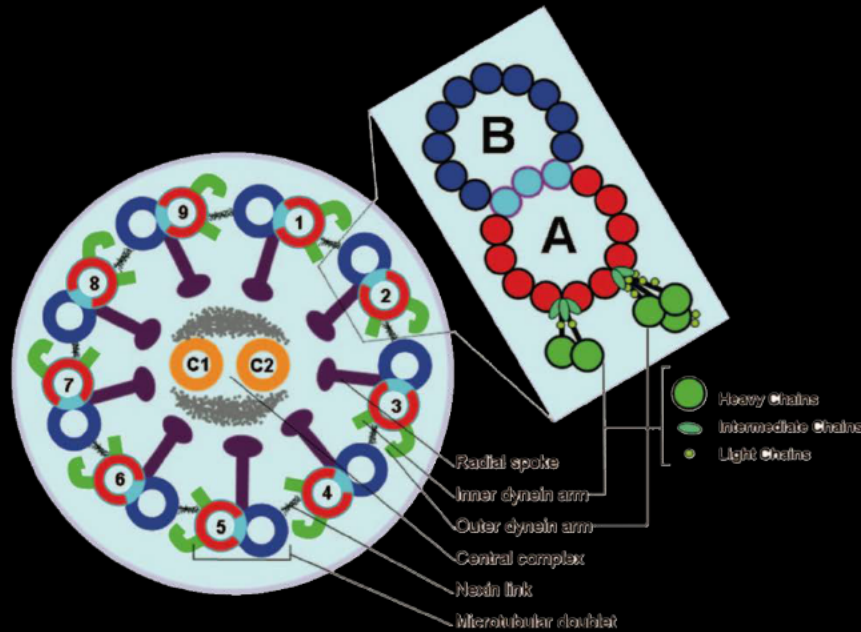
Situs inversus and lower lobe bronchiectasis



bronchiectasi



Primary ciliary dyskinesia



Normal

Both
Arms
Absent

Absent
ODA

Absent
IDA

REVIEW

Clinical and genetic aspects of primary ciliary dyskinesia/Kartagener syndrome

Margaret W. Leigh, MD¹, Jessica E. Pittman, MD¹, Johnny L. Carson, PhD¹, Thomas W. Ferkol, MD², Sharon D. Dell, MD³, Stephanie D. Davis, MD¹, Michael R. Knowles, MD⁴, and Maimoona A. Zariwala, PhD⁵

Abstract: Primary ciliary dyskinesia is a genetically heterogeneous disorder of motile cilia. Most of the disease-causing mutations identified to date involve the heavy (*dynein axonemal heavy chain 5*) or intermediate (*dynein axonemal intermediate chain 1*) chain dynein genes in ciliary outer dynein arms, although a few mutations have been noted in other genes. Clinical molecular genetic testing for primary ciliary dyskinesia is available for the most common mutations. The respiratory manifestations of primary ciliary dyskinesia (chronic bronchitis leading to bronchiectasis, chronic rhino-sinusitis, and chronic otitis media) reflect impaired mucociliary clearance owing to defective axonemal structure. Ciliary ultrastructural analysis in most patients (>80%) reveals defective dynein arms, although defects in other axonemal components have also been observed. Approximately 50% of patients with primary ciliary dyskinesia have laterality defects (including situs inversus totalis and, less commonly, heterotaxy, and congenital heart disease), reflecting dysfunction of embryological nodal cilia. Male infertility is common and reflects defects in sperm tail axonemes. Most patients with primary ciliary dyskinesia have a history of neonatal respiratory distress, suggesting that motile cilia play a role in fluid clearance during the transition from a fetal to neonatal lung. Ciliopathies involving sensory cilia, including autosomal dominant or recessive polycystic kidney disease, Bardet-Biedl syndrome, and Alstrom syndrome, may have chronic respiratory symptoms and even bronchiectasis suggesting clinical overlap with primary ciliary dyskinesia. *Genet Med* 2009;11(7):473–487.

Key Words: primary ciliary dyskinesia, PCD, Kartagener syndrome, situs inversus, dynein

OVERVIEW

Primary ciliary dyskinesia (PCD) (MIM no. 244400) is a genetically heterogeneous, typically autosomal recessive, disorder characterized by ciliary dysfunction and impaired mucociliary clearance, resulting in an array of clinical manifestations,

including chronic bronchitis leading to bronchiectasis, chronic rhino-sinusitis, chronic otitis media, situs inversus (in approximately 50% of cases), and male infertility. The incidence of PCD is estimated at 1/16,000 births based on prevalence of situs inversus and bronchiectasis.^{1,2} However, few patients with PCD carry a well-established diagnosis, which reflects the limited ability to diagnose this disorder.

The first case, reported in the early 1900s, and characterized by a triad of symptoms that included chronic sinusitis, bronchiectasis, and situs inversus,³ became known as Kartagener syndrome. Subsequently, patients with Kartagener syndrome, as well as other patients with chronic sinusitis and bronchiectasis, were noted to have “immotile” cilia and defects in the ultrastructural organization of cilia.^{4–6} Initially, the term “immotile cilia syndrome” was used to describe this disorder; however, later studies showed that most cilia were motile, but exhibited a stiff, uncoordinated, and/or ineffective beat. The name was changed to “primary ciliary dyskinesia” to more appropriately describe its heterogeneous genetic base and the ciliary dysfunction, and to distinguish it from the secondary ciliary defects acquired after multiple causes of epithelial injury.

The “gold-standard” diagnostic test for PCD has been electron microscopic ultrastructural analysis of respiratory cilia obtained by nasal scrape or bronchial brush biopsy. Recent studies have identified mutations in several genes encoding structural and/or functional proteins in cilia. Limited clinical genetic testing is currently available, but a multicenter, international collaboration is focused on defining additional PCD-specific gene mutations to expand PCD genetic testing. Recently, nasal nitric oxide (NO) measurement has been used as a screening test for PCD, because nasal NO is extremely low (10–20% of normal) in patients with PCD.^{7–10} As an adjunct test, nasal NO measurement can identify individuals with probable PCD (even if ciliary ultrastructure appears normal) to target for genetic testing. We anticipate that genetic testing for PCD will soon become the gold standard diagnostic test in a growing number of cases.

In summary, we are in the midst of a revolution for the diagnosis, and understanding of genotype/phenotype correlations in PCD. This effort has greatly benefited from an National Institutes of Health-sponsored Rare Disease Network (<http://rarediseasesnetwork.epi.usf.edu/>), and a consortium focused on studying genetic disorders of mucociliary clearance, including PCD (<http://rarediseasesnetwork.epi.usf.edu/gdmcc/index.htm>).

CILIARY STRUCTURE AND FUNCTION

Normal ultrastructure of motile cilia

Cilia and flagella are evolutionarily ancient organelles whose structure and function have been rigidly conserved across the phylogenetic spectrum. Historically recognized for their role in cell motility and transport of fluids over mucosal surfaces, cilia have recently been recognized to have a sensory function that modulates elements of development and cell function. Both

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Disclosure: The authors declare no conflict of interest.

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Kartagener Syndrome

Tyson Tragon

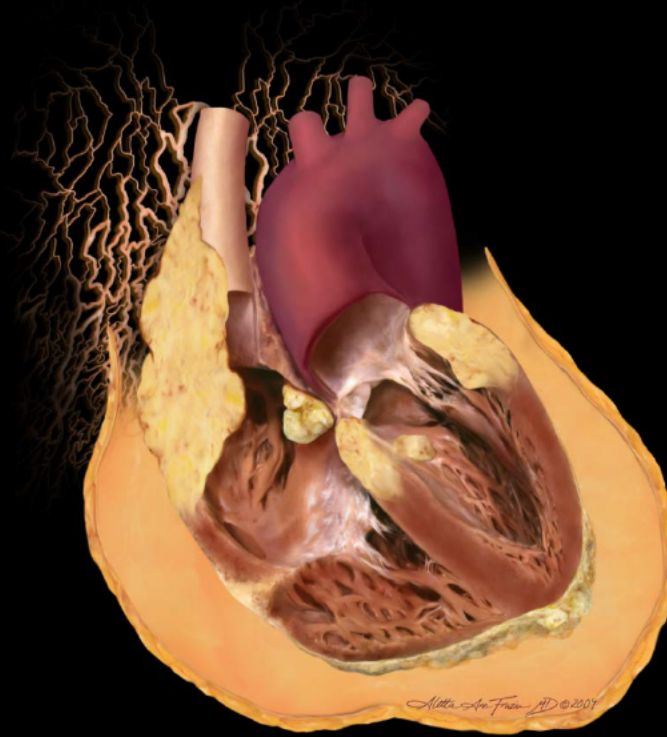
University of Pittsburgh Medical Center
Pittsburgh, Pennsylvania



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Cardiovascular Best Case



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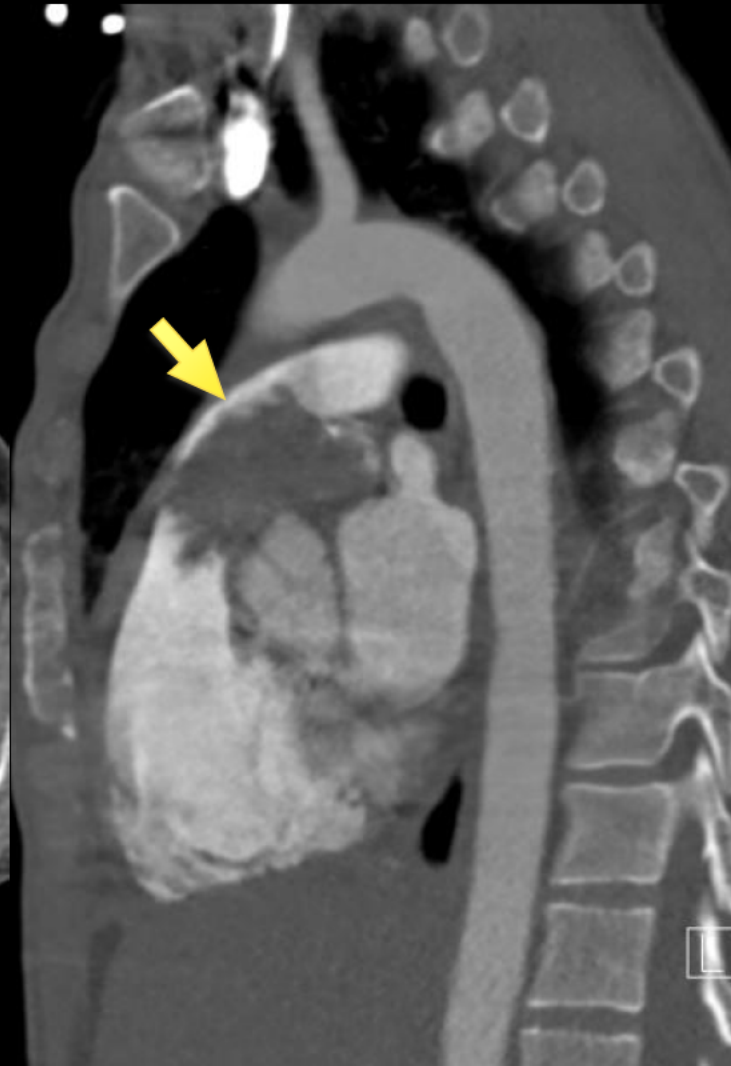
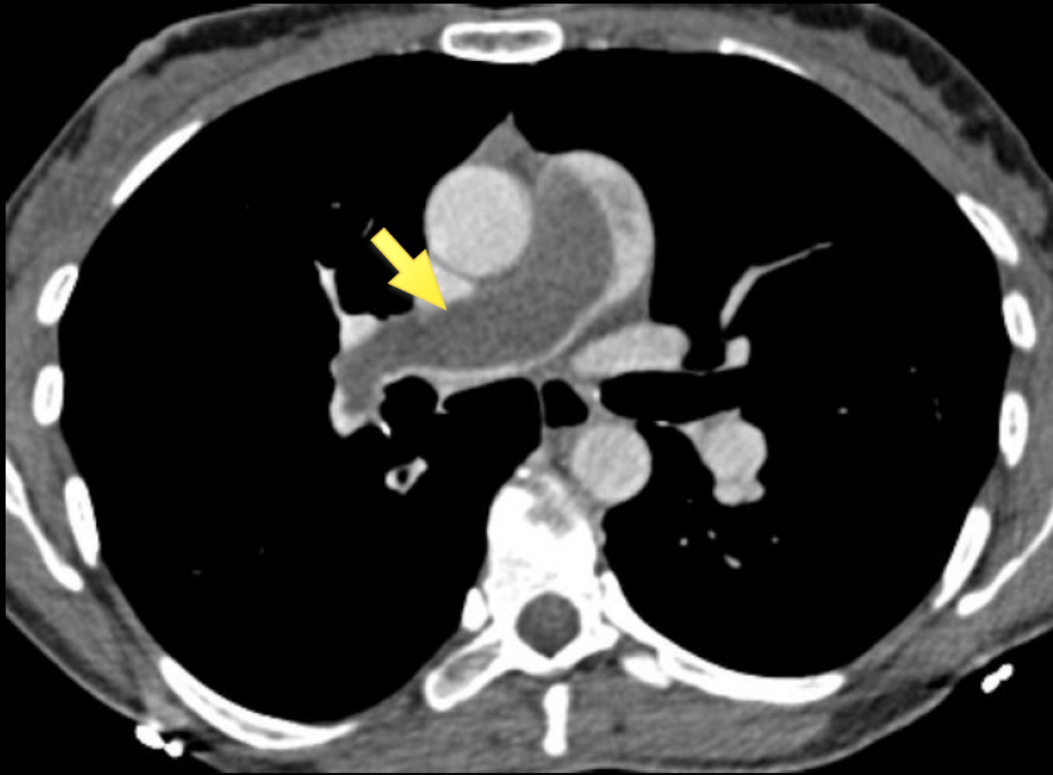
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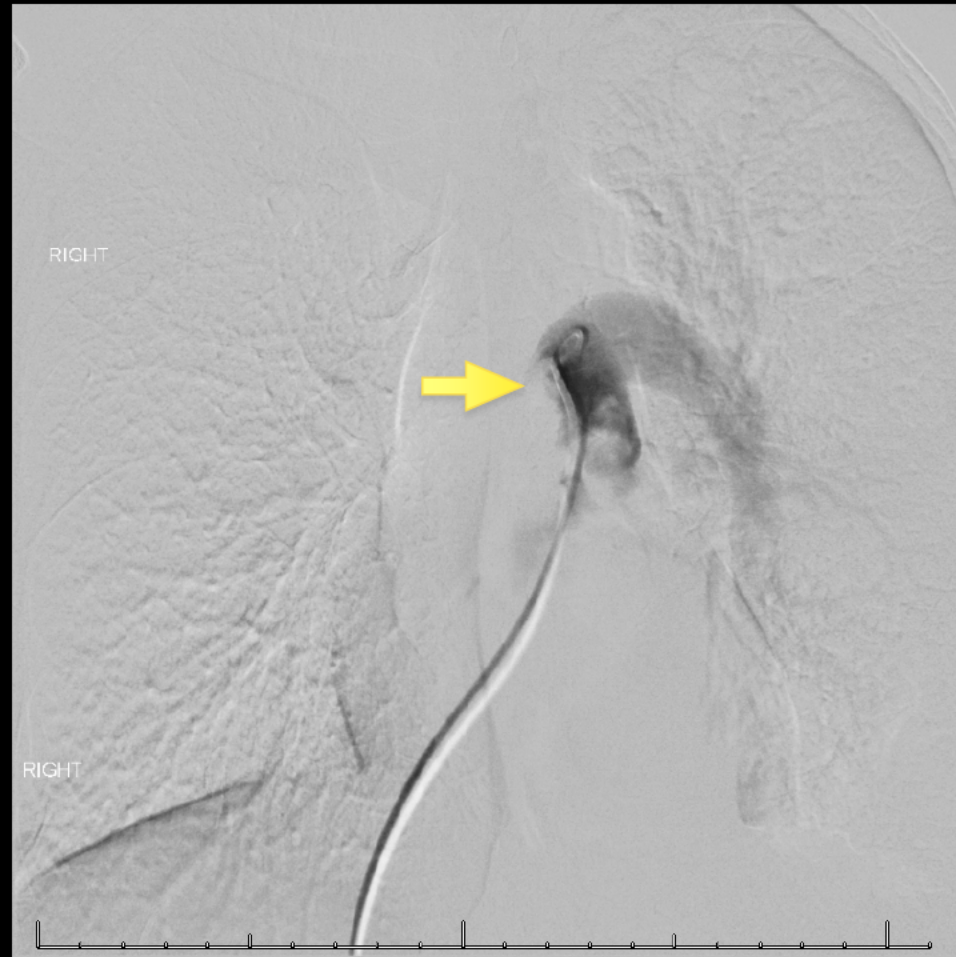
48 year old female with four-month history of dyspnea and non-productive cough.

Initial workup revealed mild tachycardia, elevated D-dimer, and CTA with multiple intravascular filling defects in the pulmonary arteries.

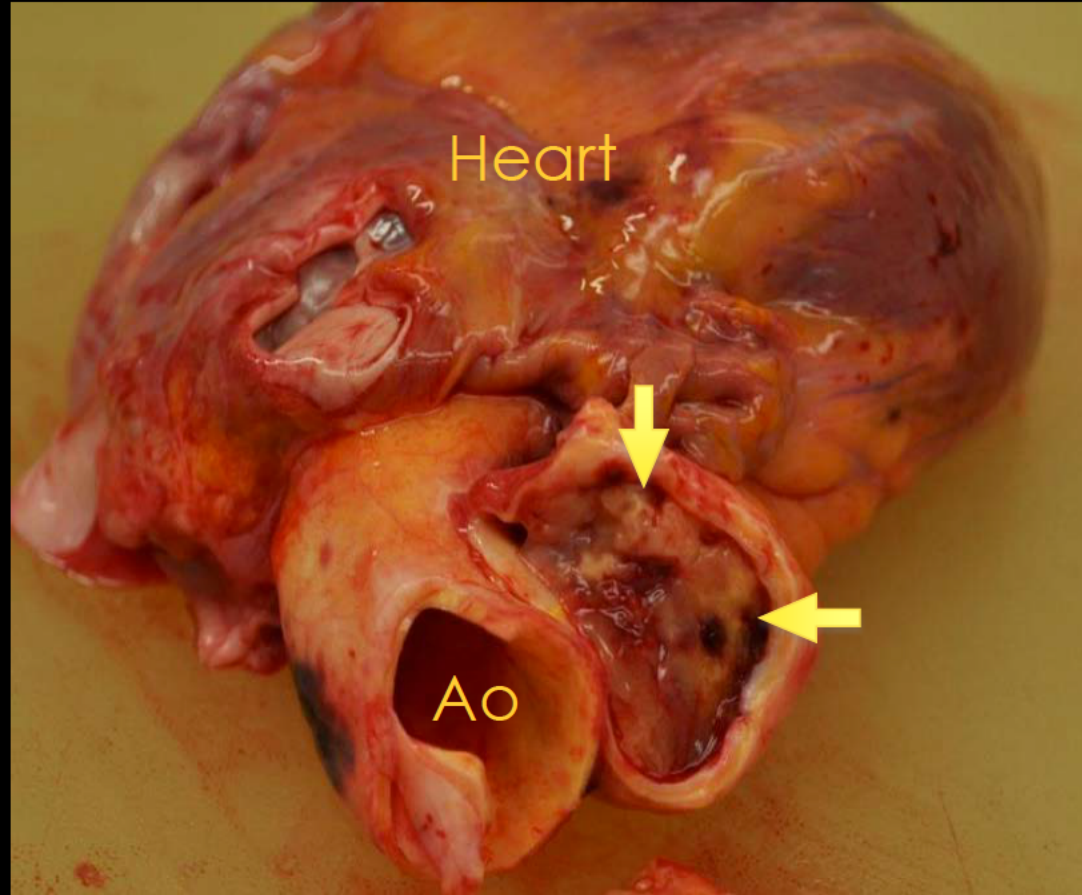
She was discharged on Lovenox, but returned 10 days later with significant worsening of symptoms.



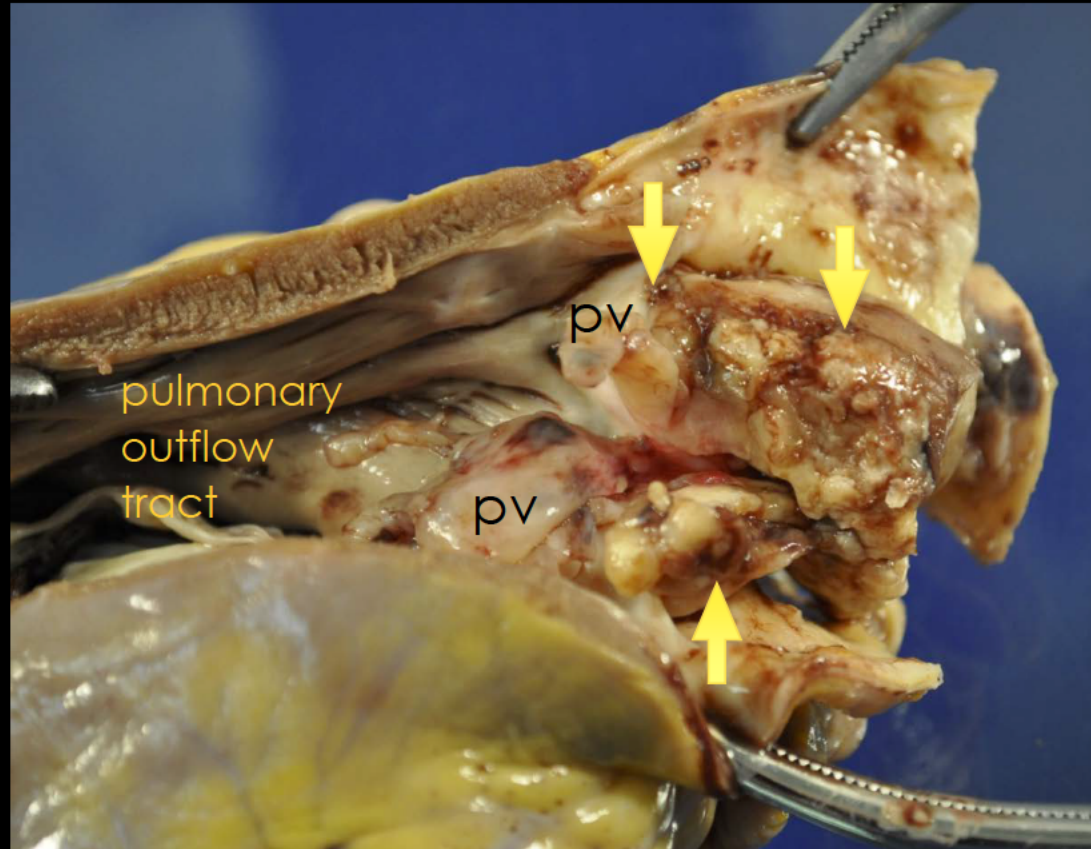
CTA: large broad-based filling defect, right side of main PA extending into right PA. (Shaggy margins, possible peripheral enhancement?)



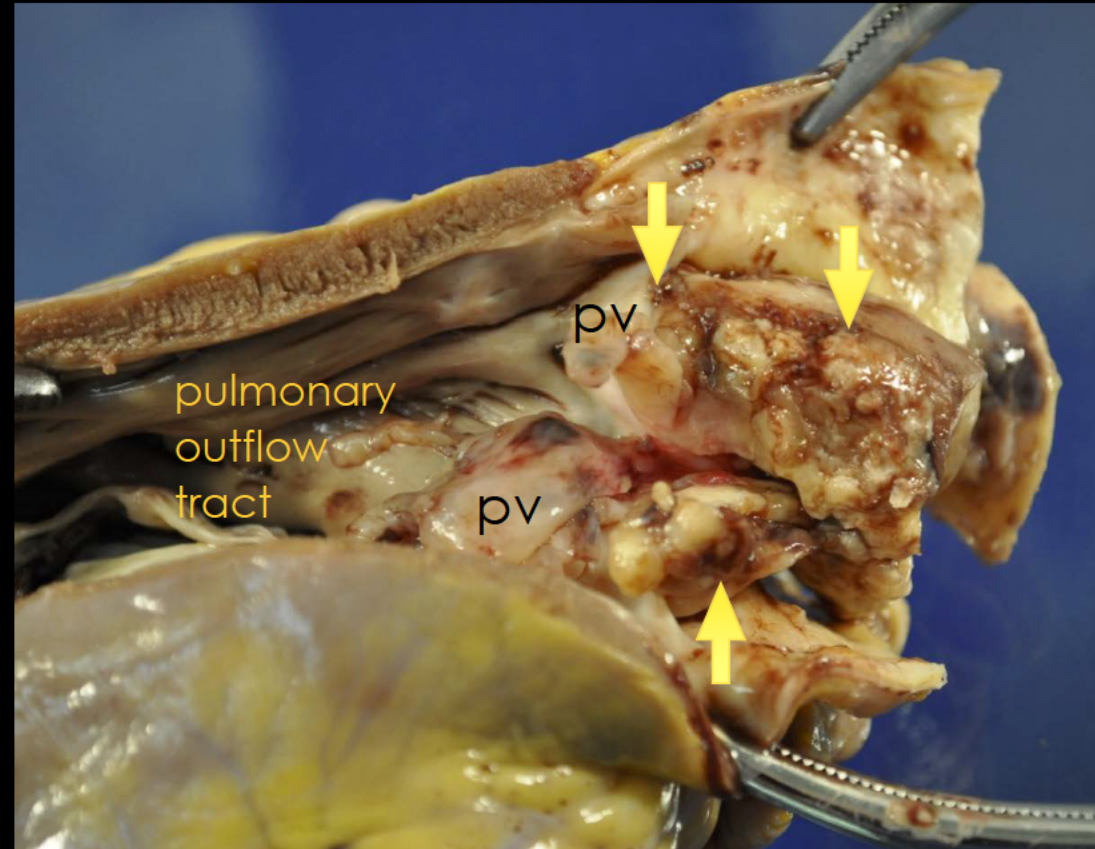
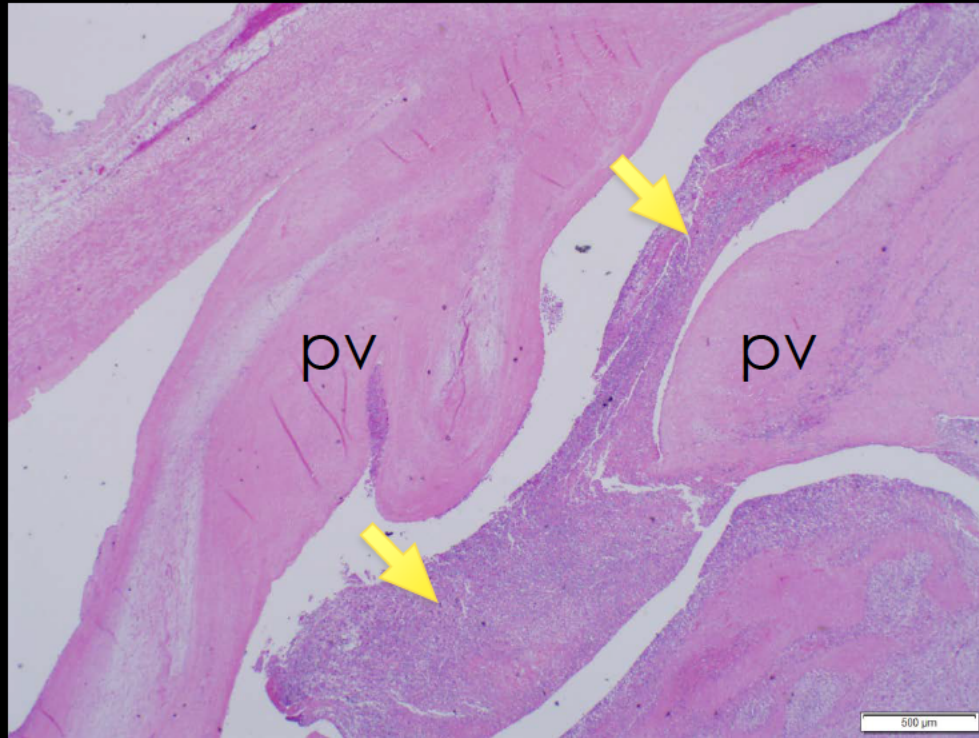
Pulmonary angiogram: cut-off of right main PA



Autopsy performed after cardiac arrest.
Heterogeneous-appearing soft tissue fills main PA (arrows). Adjacent aortic lumen (Ao) is clear.



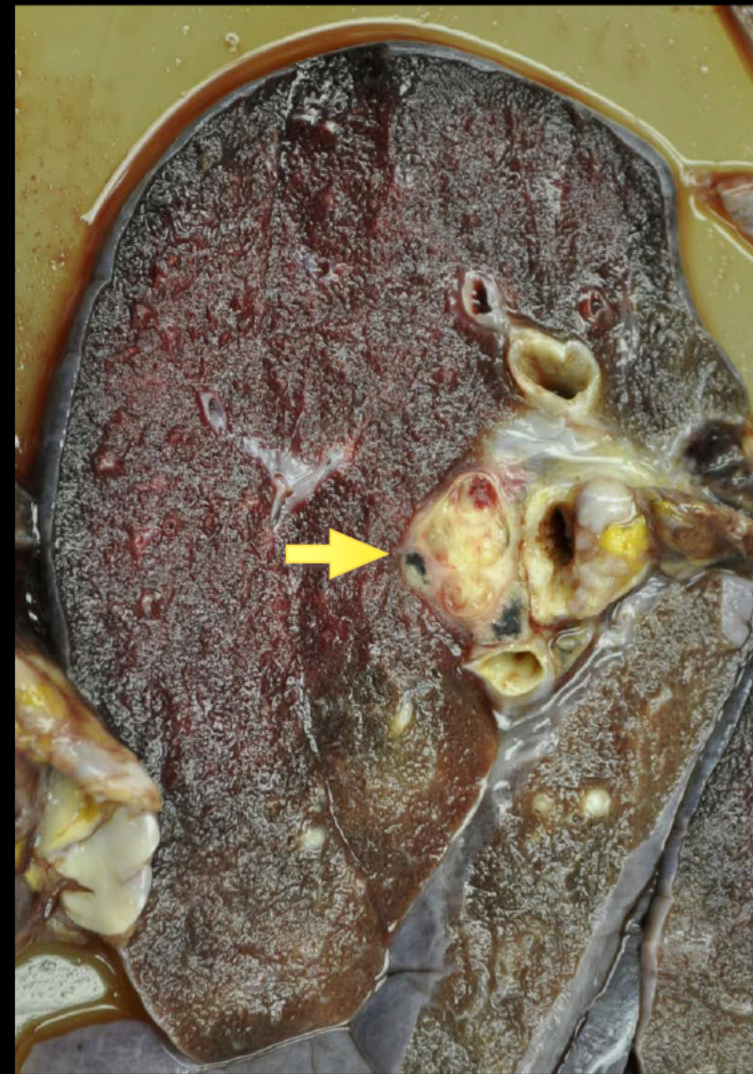
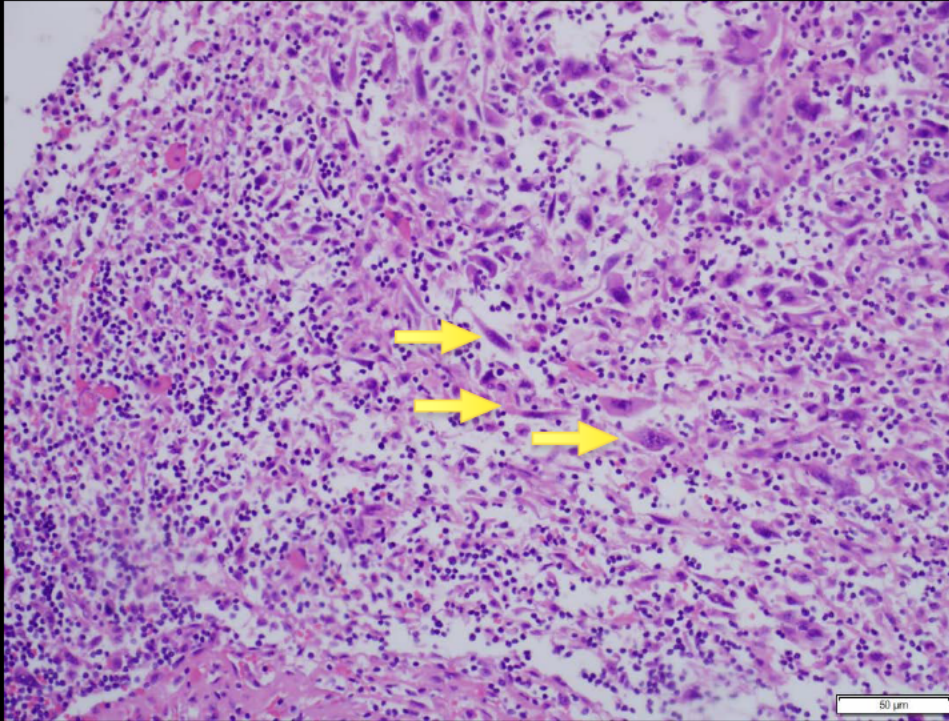
Lobulated soft tissue mass with frond-like margins (arrows) also invades the pulmonary valve (pv).



H&E stained tissue confirms sheets of tumor (arrows) coating pulmonary valve leaflets (pv).



Gross right lung shows central intravascular soft tissue (arrow) which was also found extending into distal lobar vessels.



H&E stained tissue shows dense field of pleomorphic spindle cells (arrows).

Pulmonary Artery Intimal Sarcoma

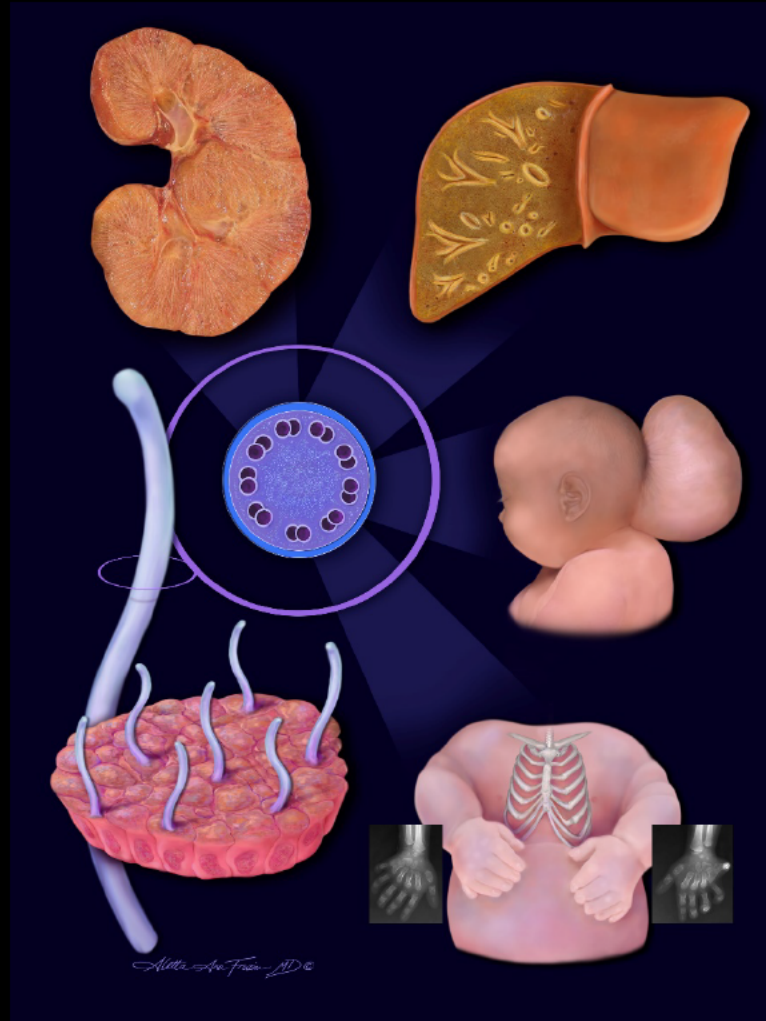
Jessica Chan, MD
University of Utah
Salt Lake City, Utah

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Pediatric Best Case



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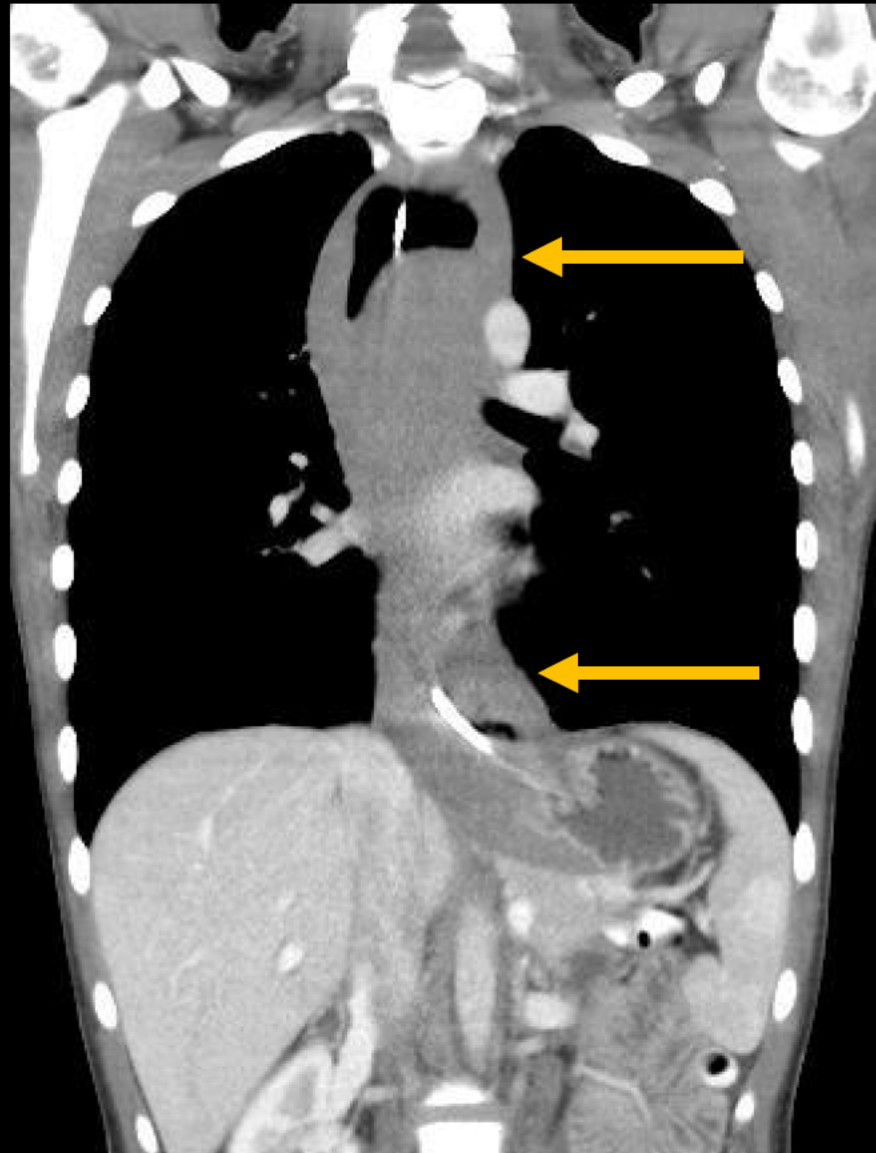
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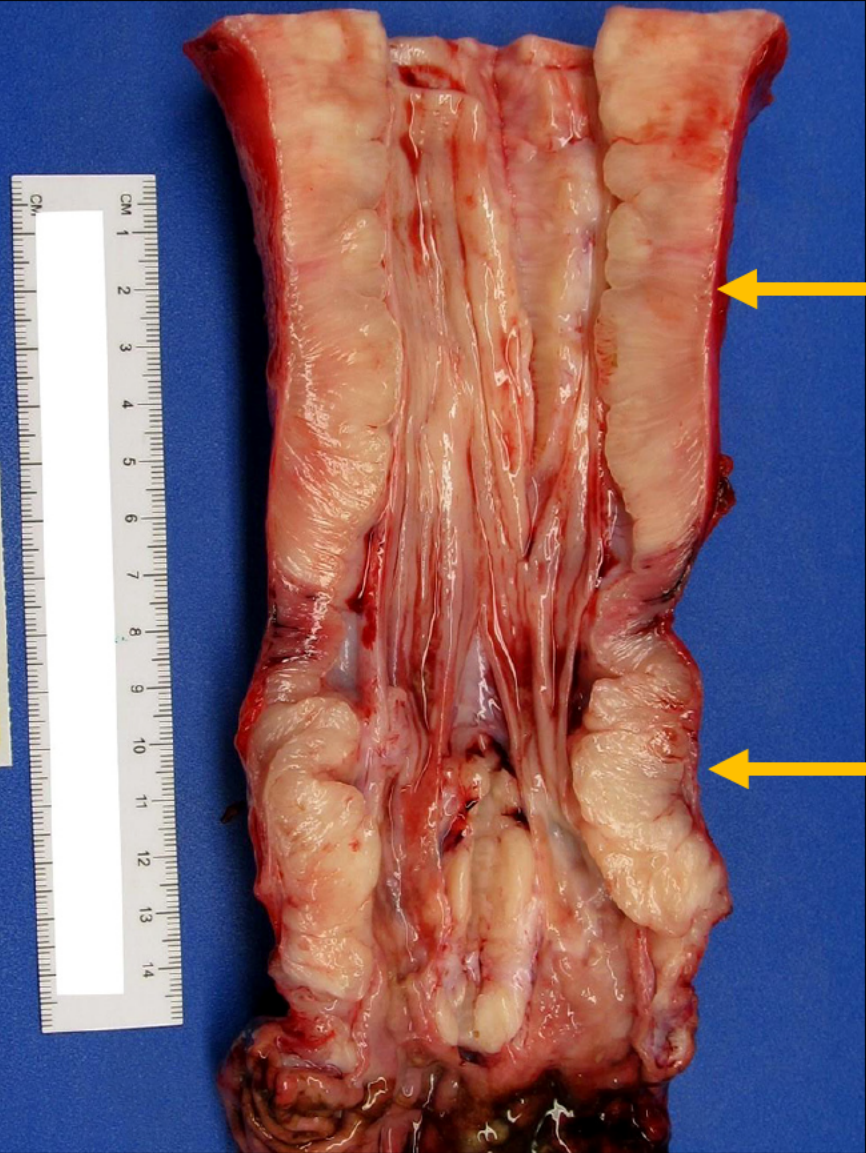
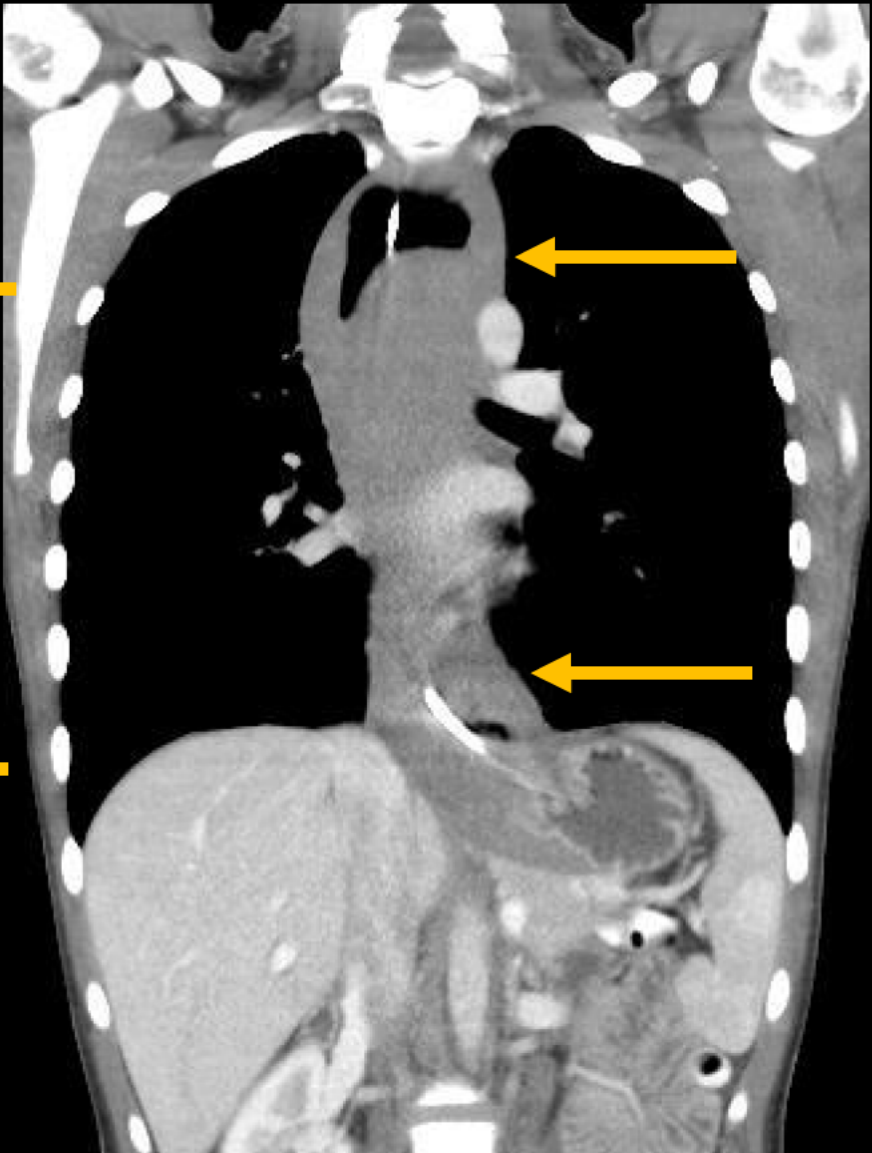
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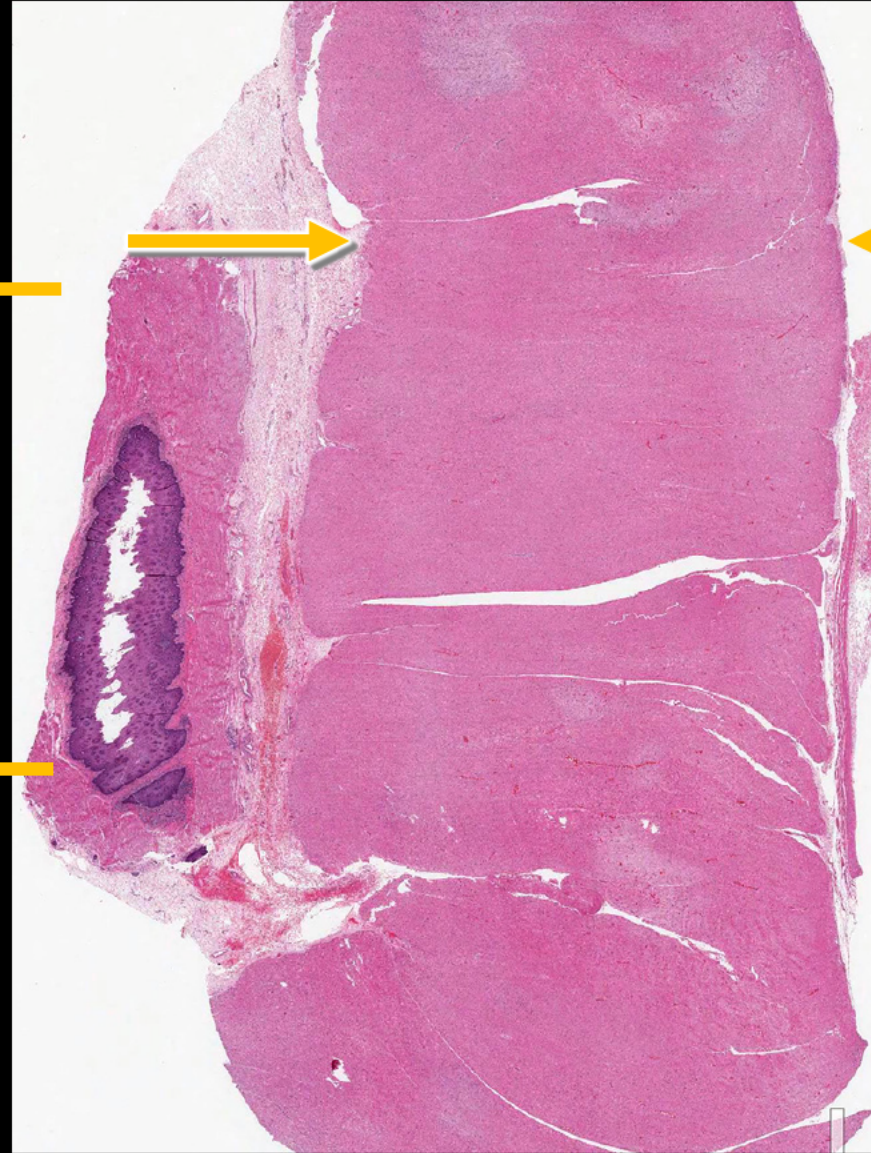
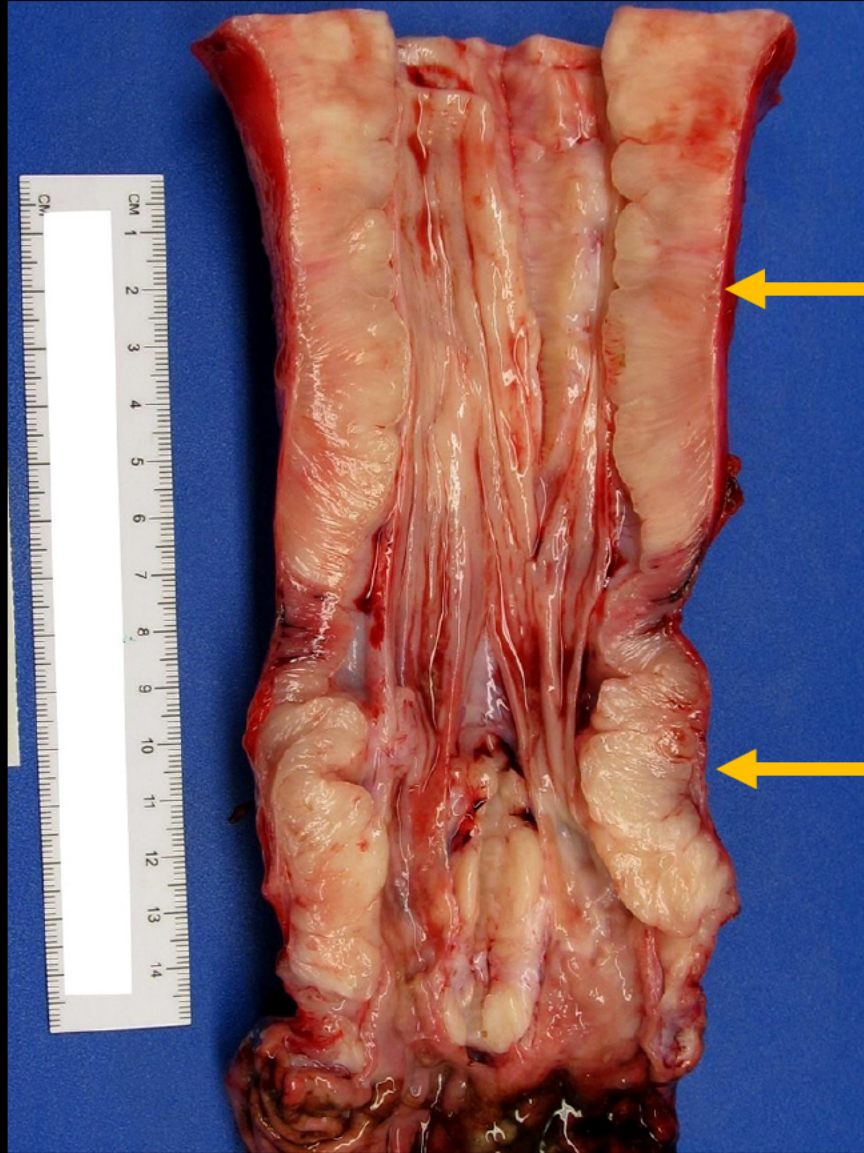
Clinical information

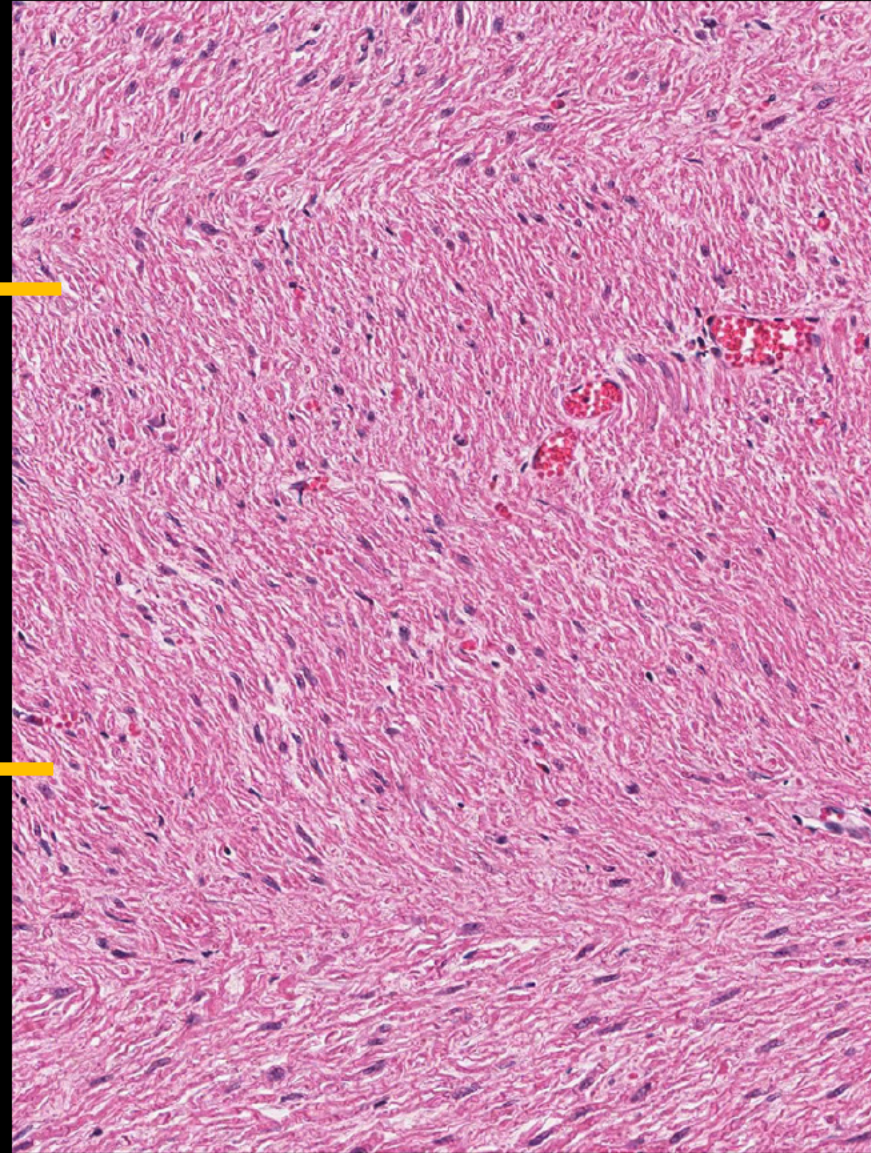
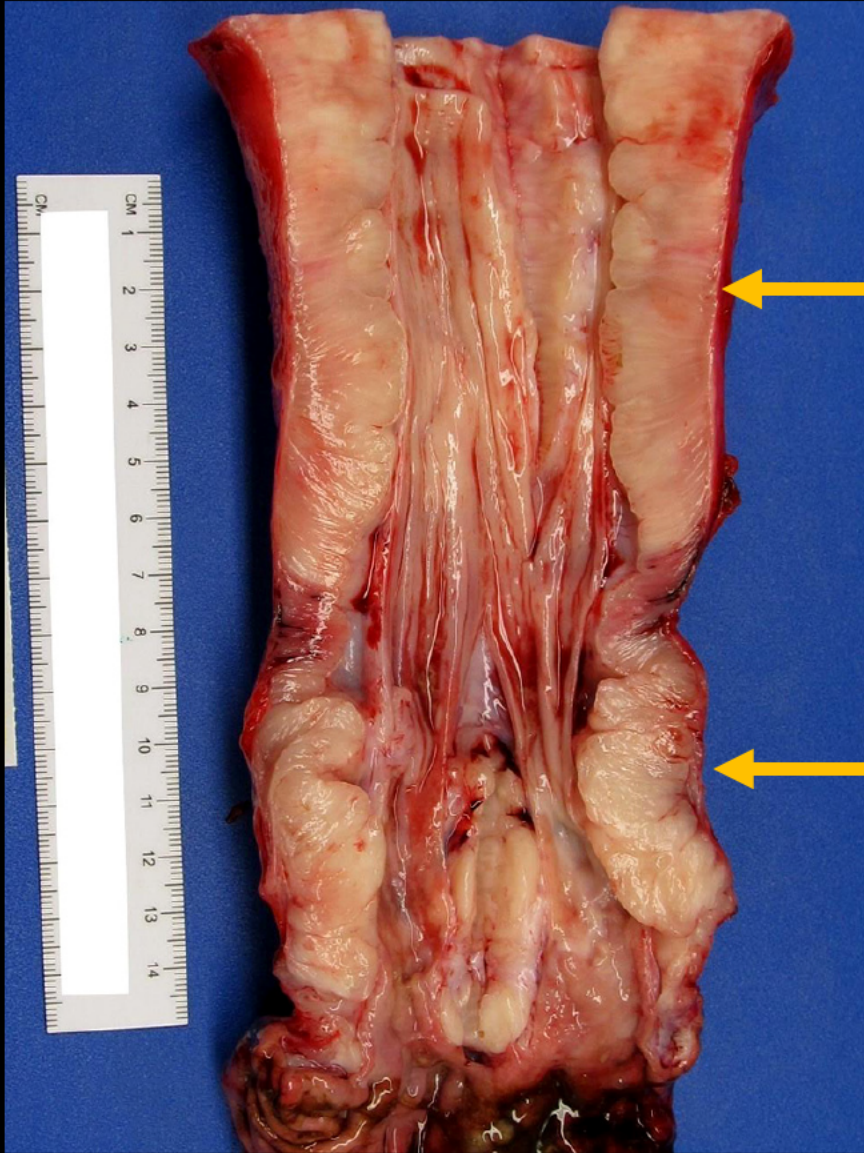
A 14-year-old female presents with progressive obstructive airway disease as well as a history of repeated choking episodes since 18 months old as well as poor weight gain.











Esophageal Leiomyomatosis associated with Alport Syndrome

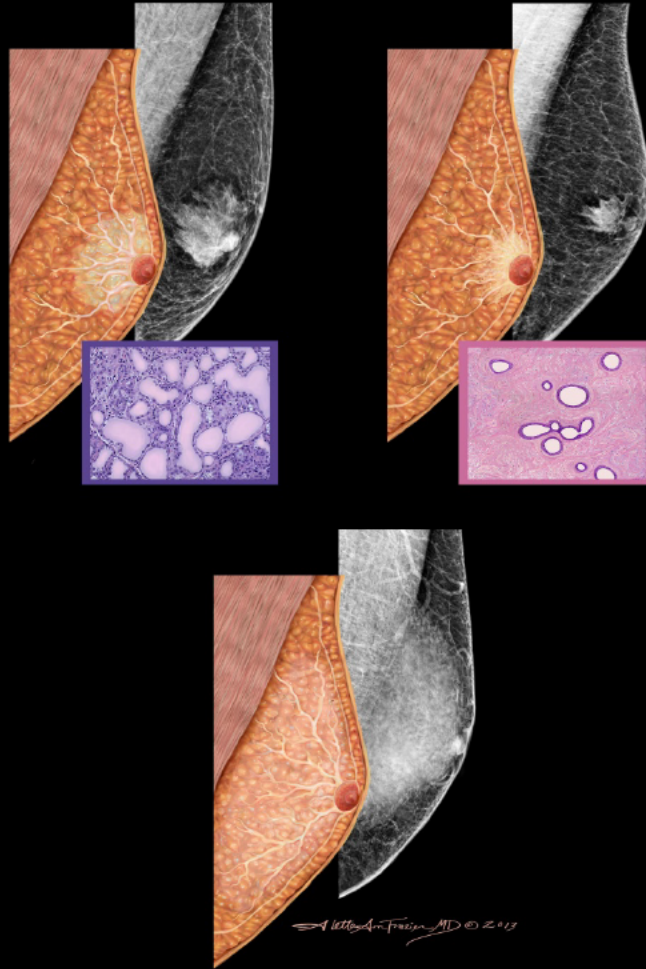
Brian Pogatchnik

**University of Minnesota Medical Center
Minneapolis, MN**



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Breast Best Case



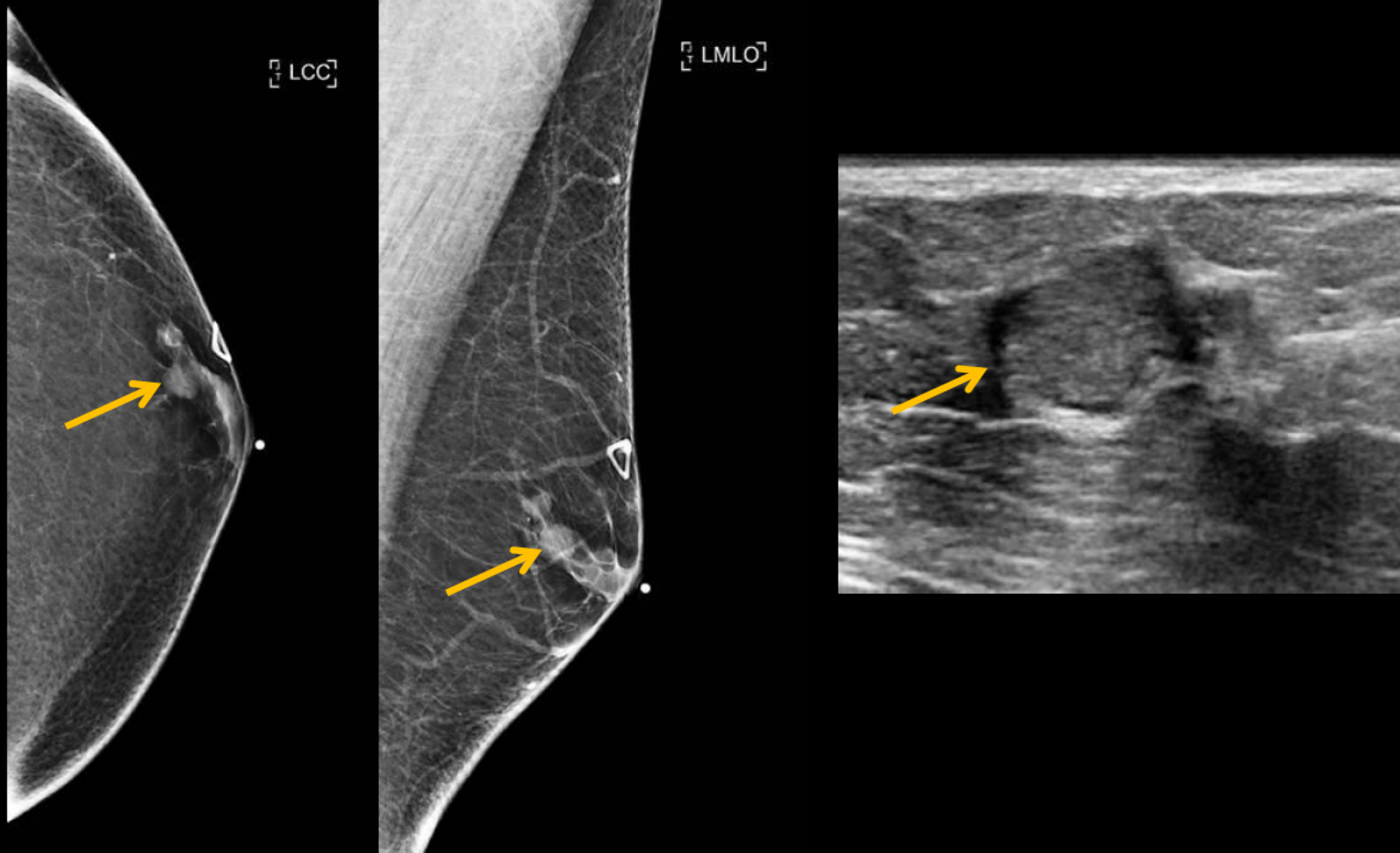
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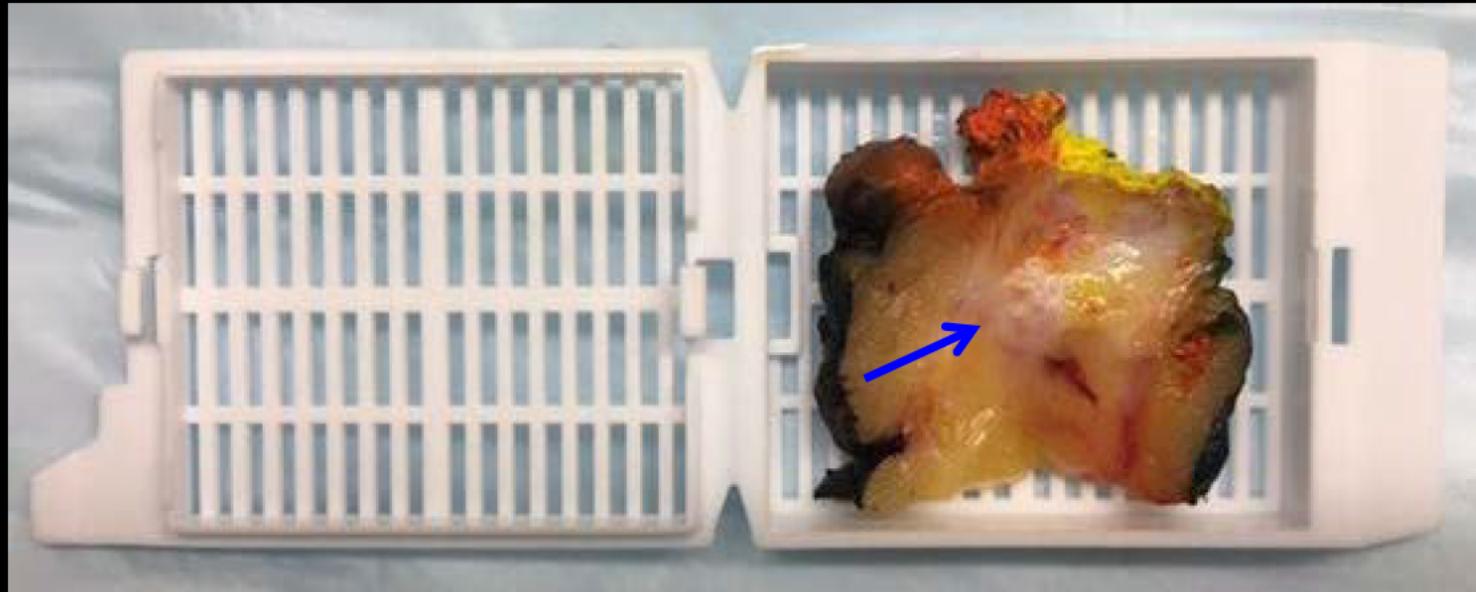
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32-year-old male with a palpable lump and serosanguinous discharge from his left nipple occurring 8 weeks prior to presentation.

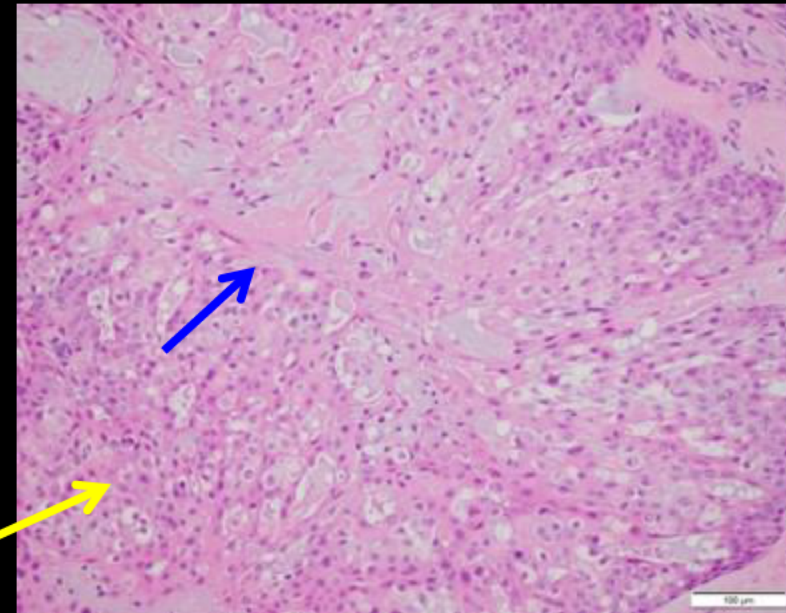
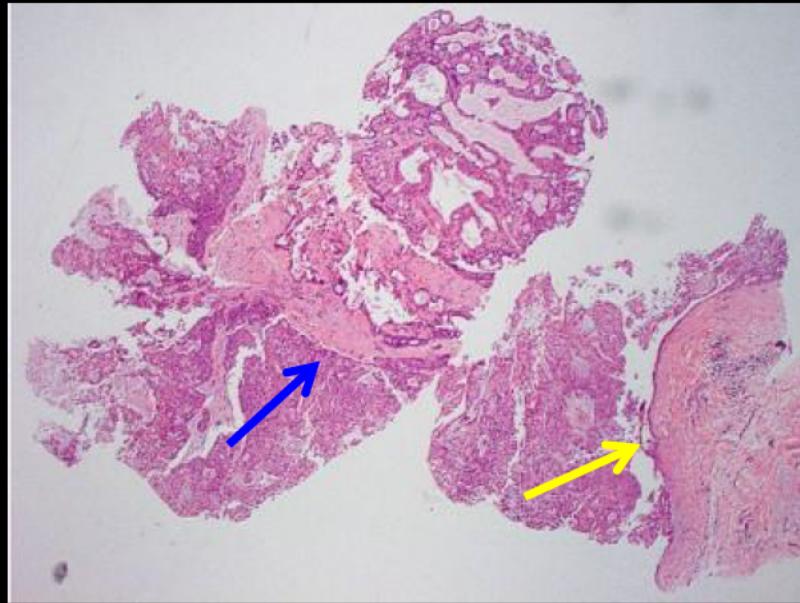
Mammogram and US



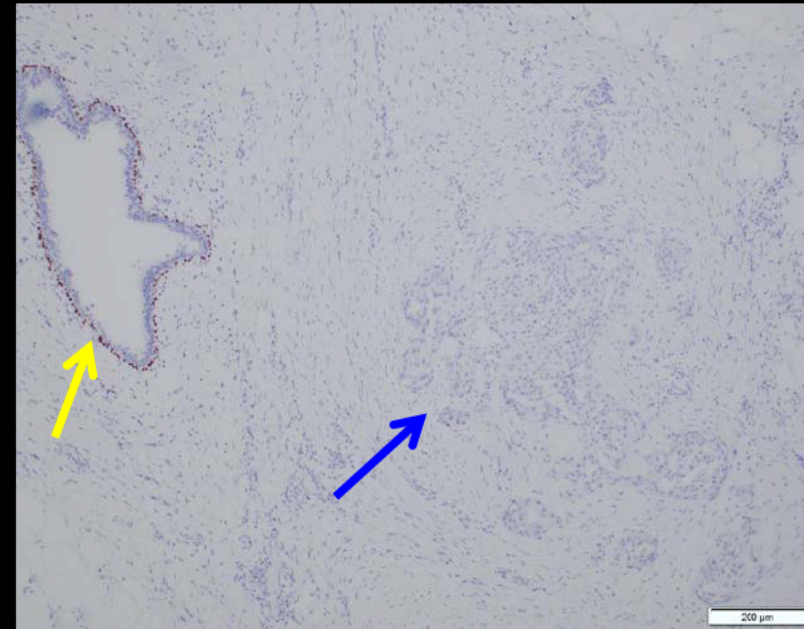
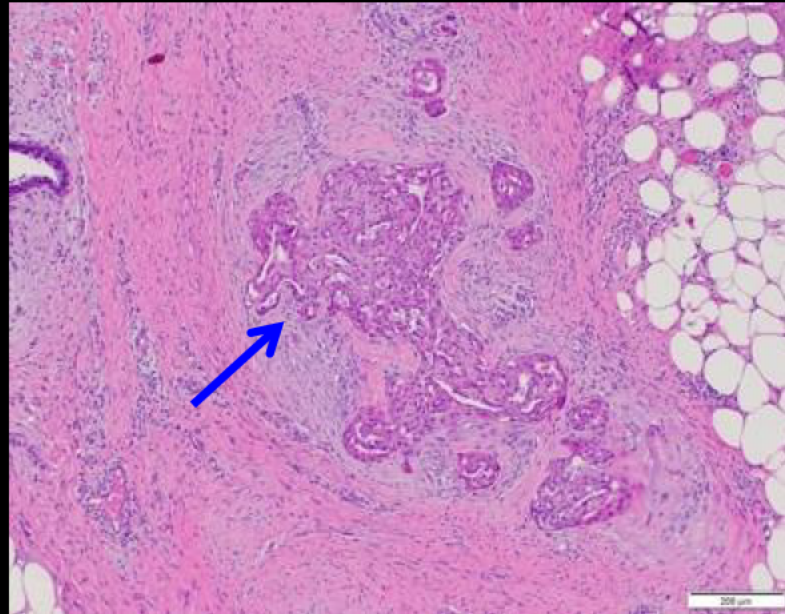
Gross



Histology



Histology



DCIS in a Papilloma with adjacent microinvasion in a Male

Karen Tran-Harding
University of Kentucky
Lexington, KY

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Nice educational article in RadioGraphics on male breast disease
Nguyen C. May 2013

Genitourinary Best Case



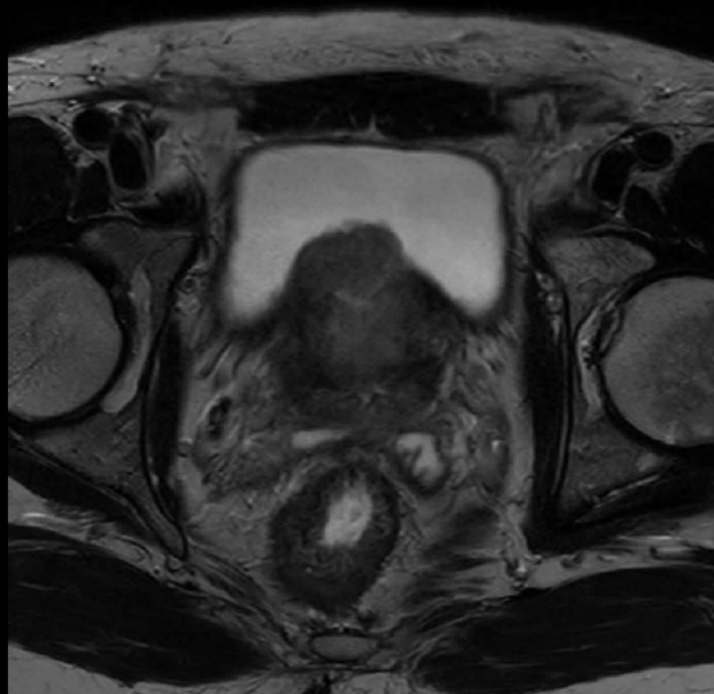
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Clinical Information

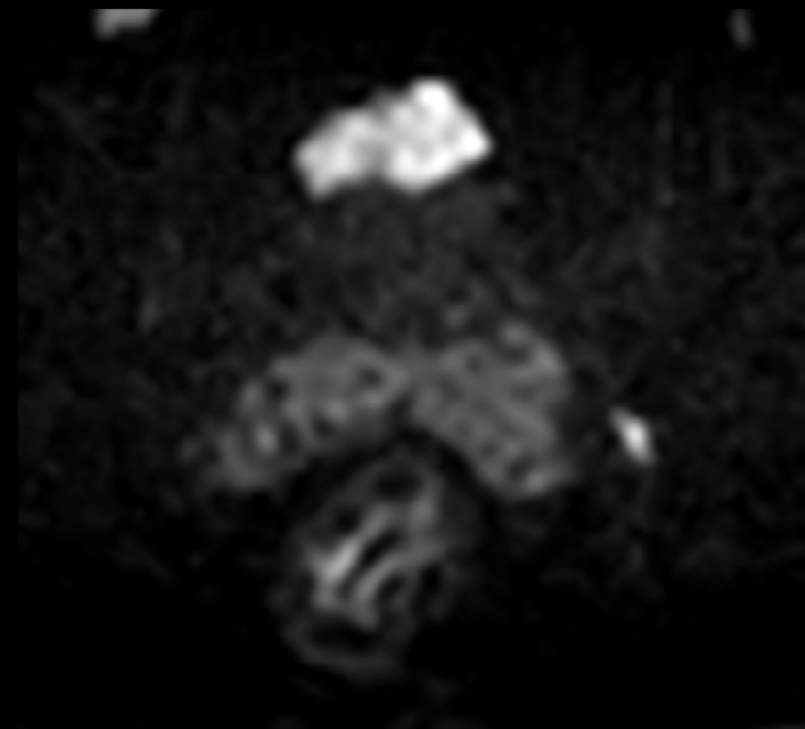
- 49 year old male with PSA 8.0. Prostate MRI performed.



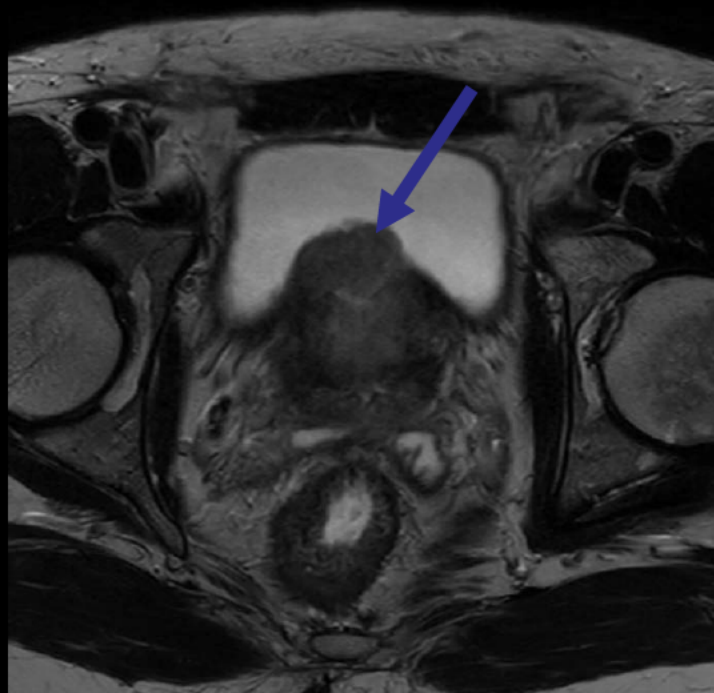
T2



ADC



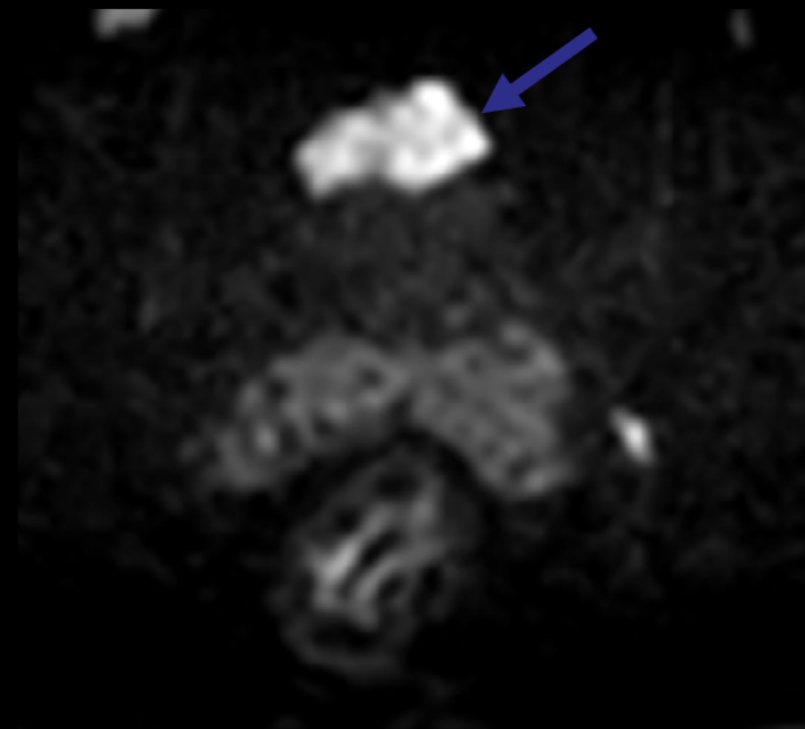
DWI



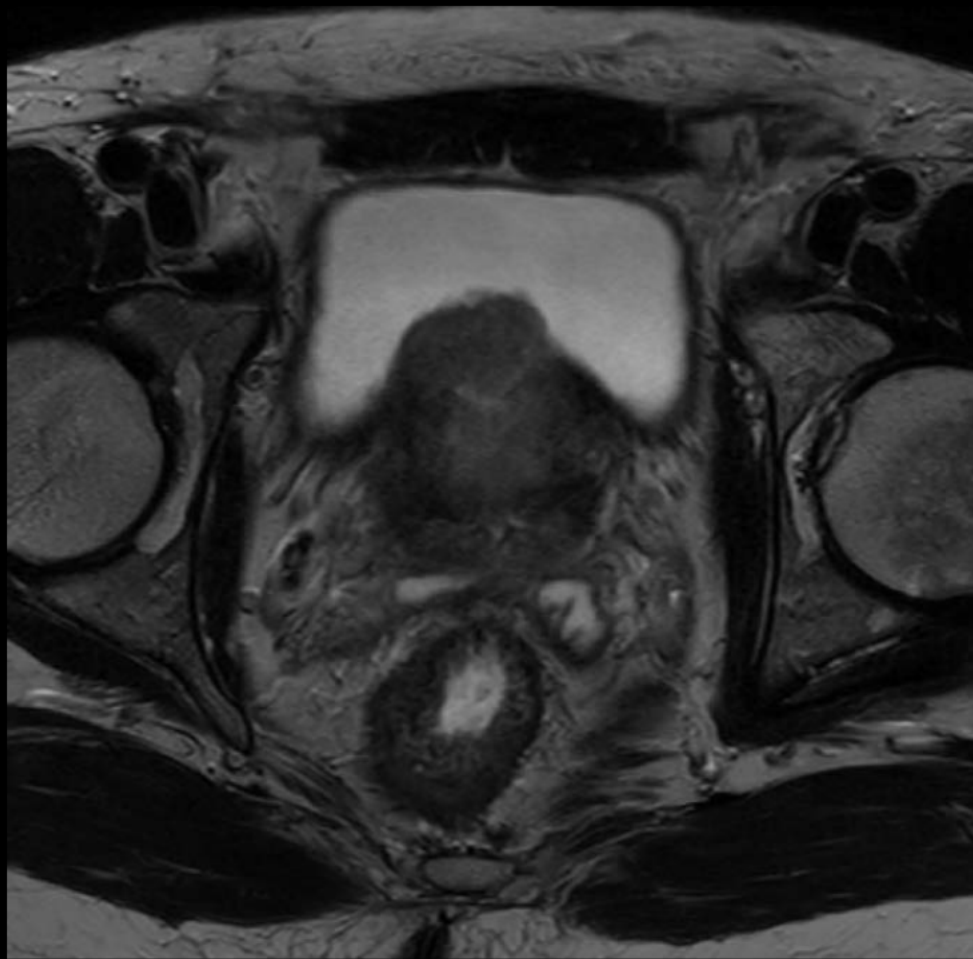
T2

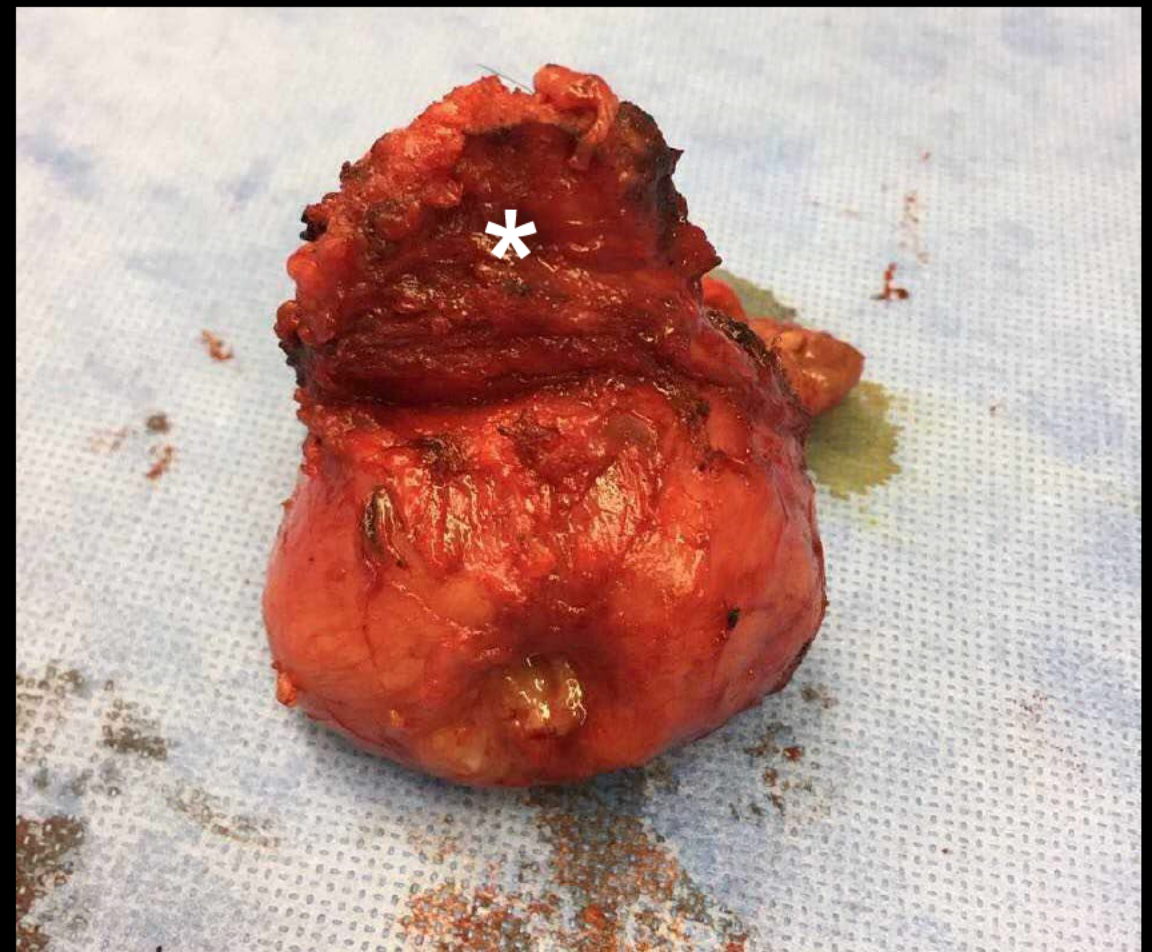


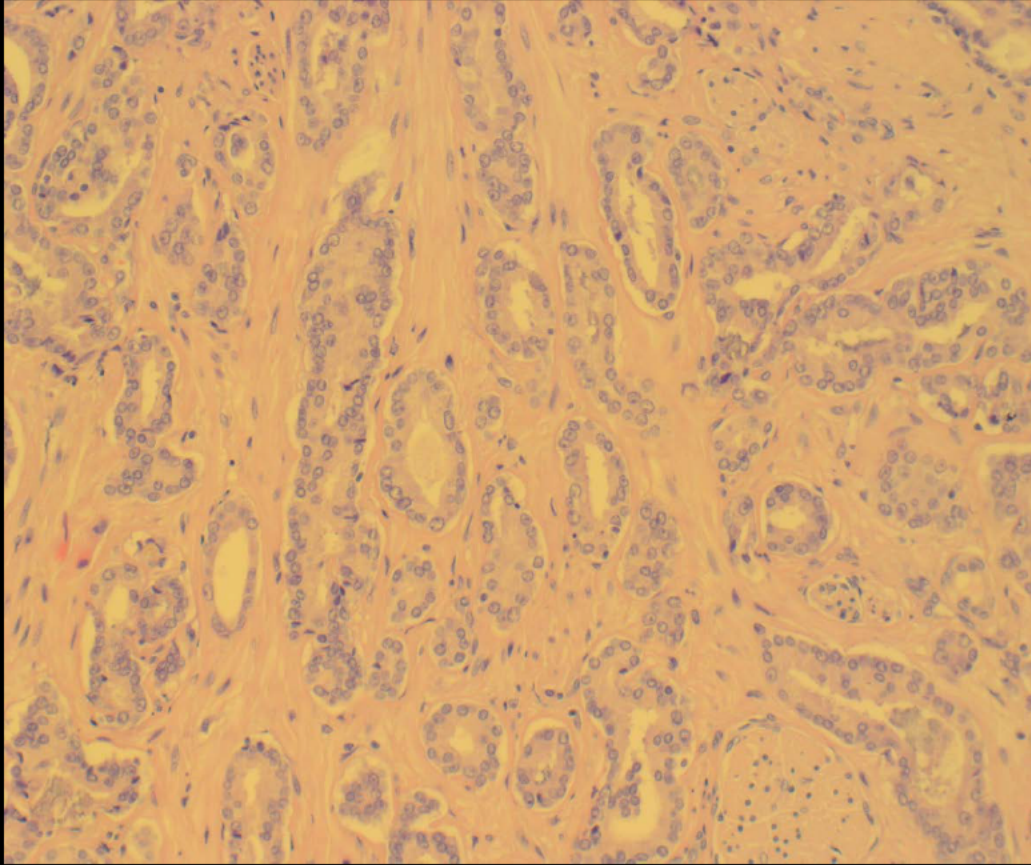
ADC



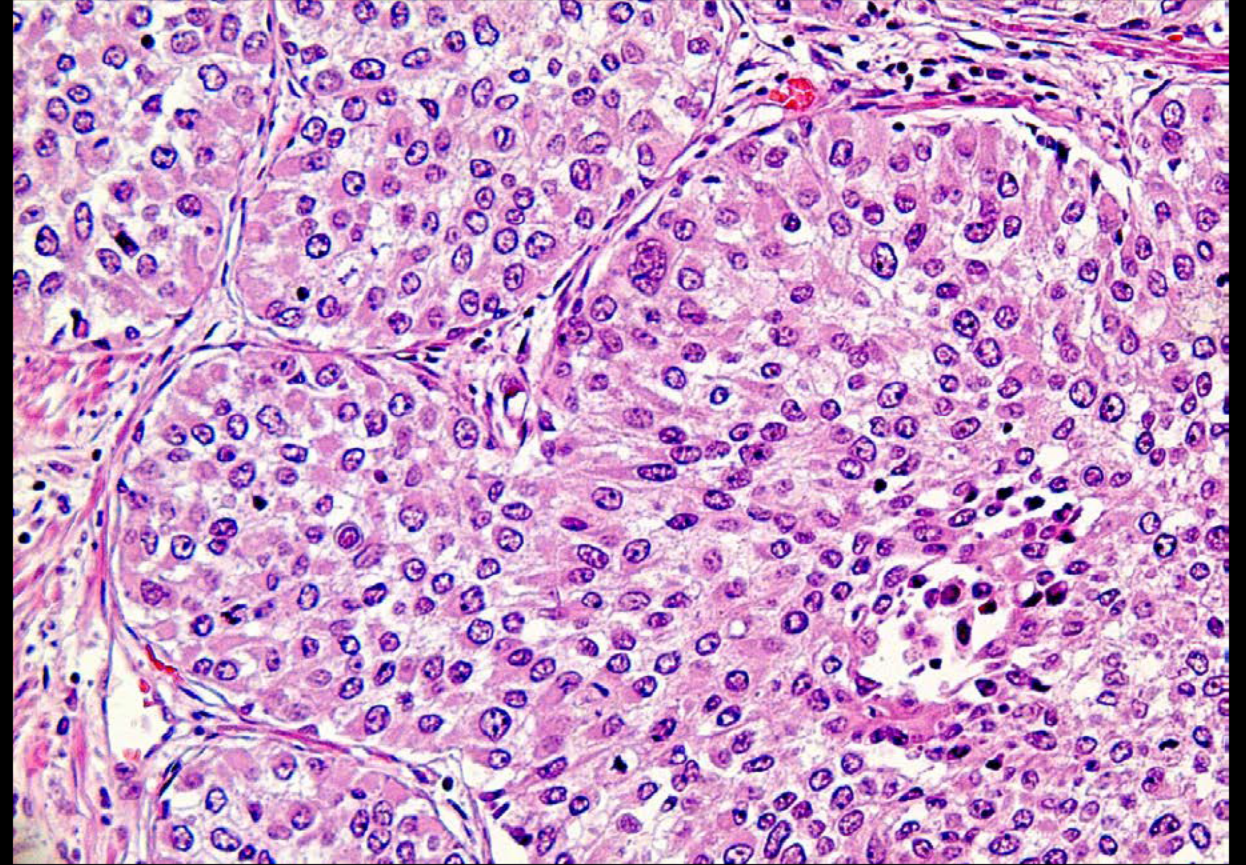
DWI







Prostate Adenocarcinoma



Urothelial Carcinoma

Prostatic Urethra Urothelial Carcinoma

Araji Abdallah

American University of Beirut

Beirut, Lebanon

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Neuroradiology Best Case

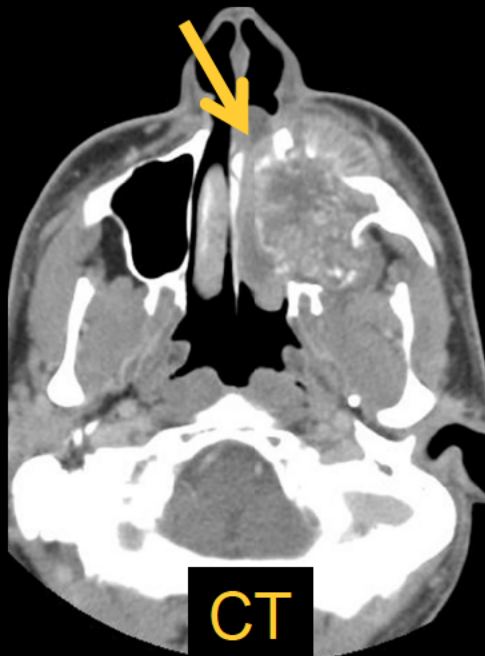


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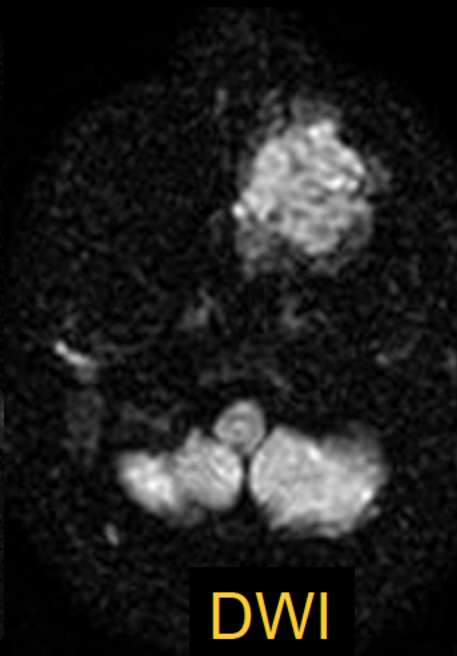
**25-year-old male with progressive left
nasal obstruction, enlarging mass of
hard palate, increasing left facial
swelling and numbness**



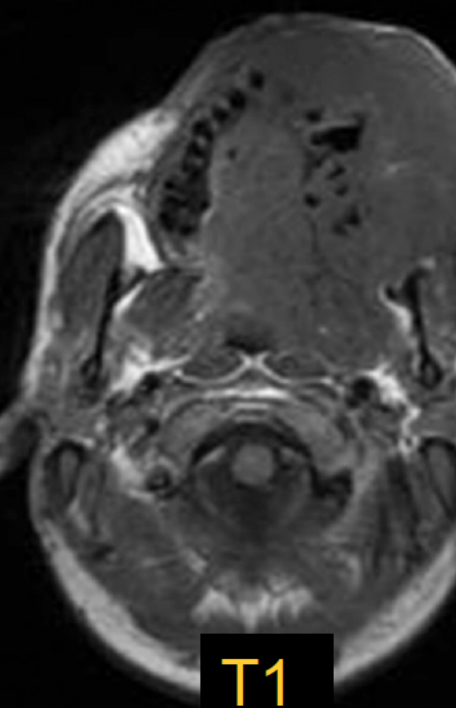
CT



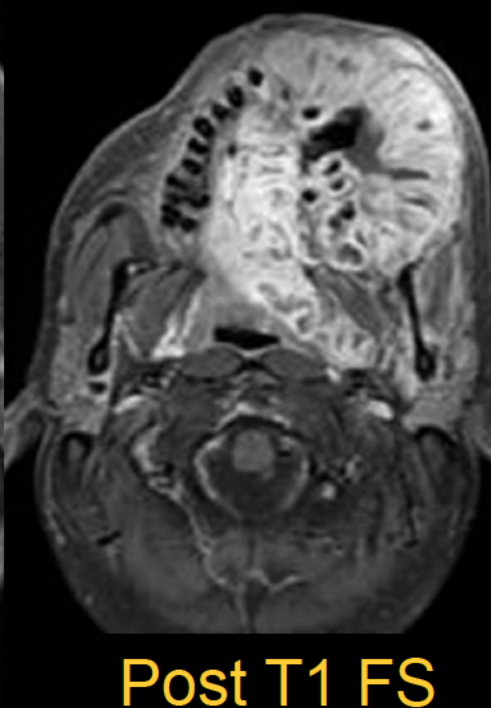
T2 FS



DWI

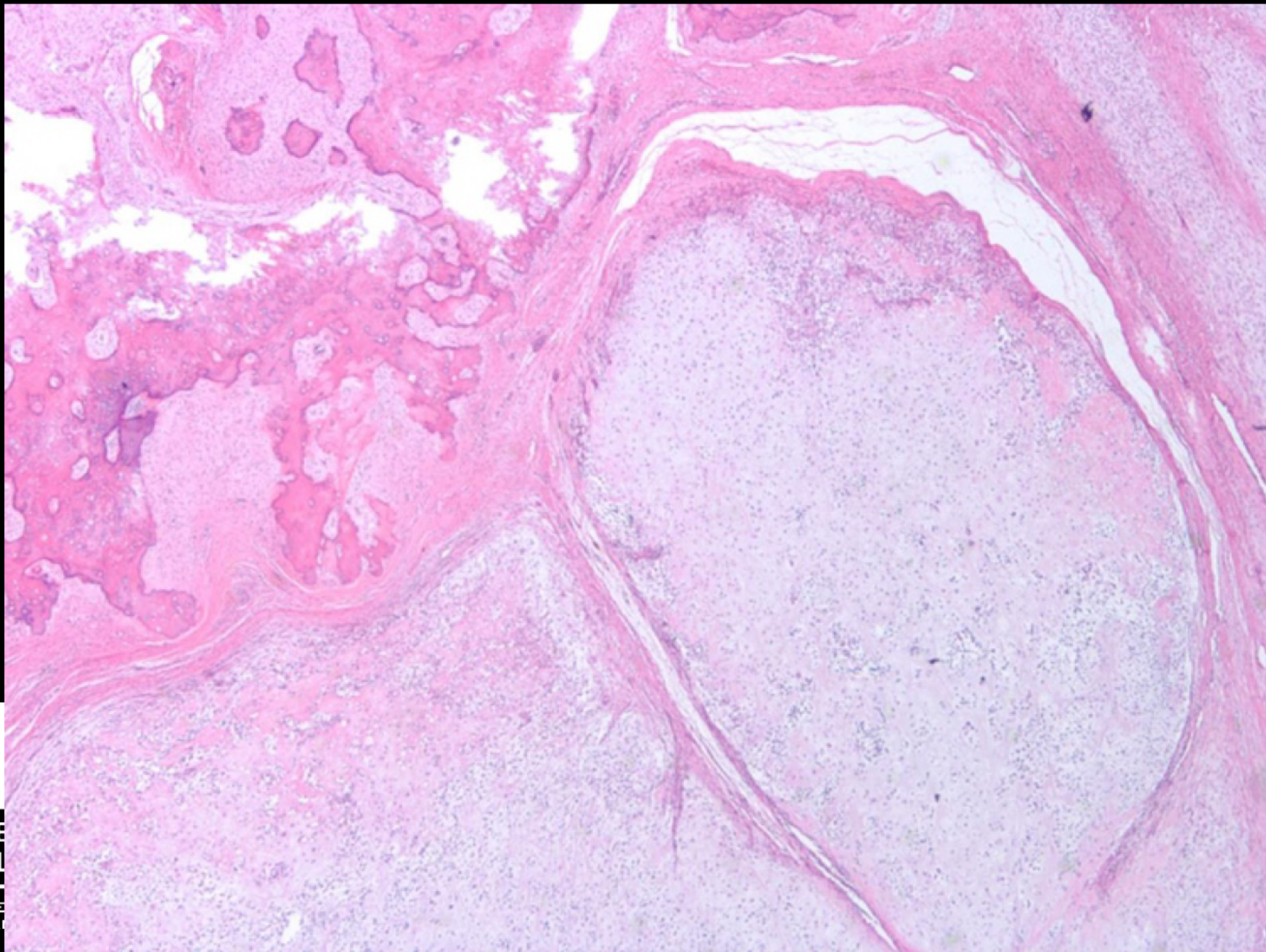


T1



Post T1 FS





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Chondroblastic Osteosarcoma

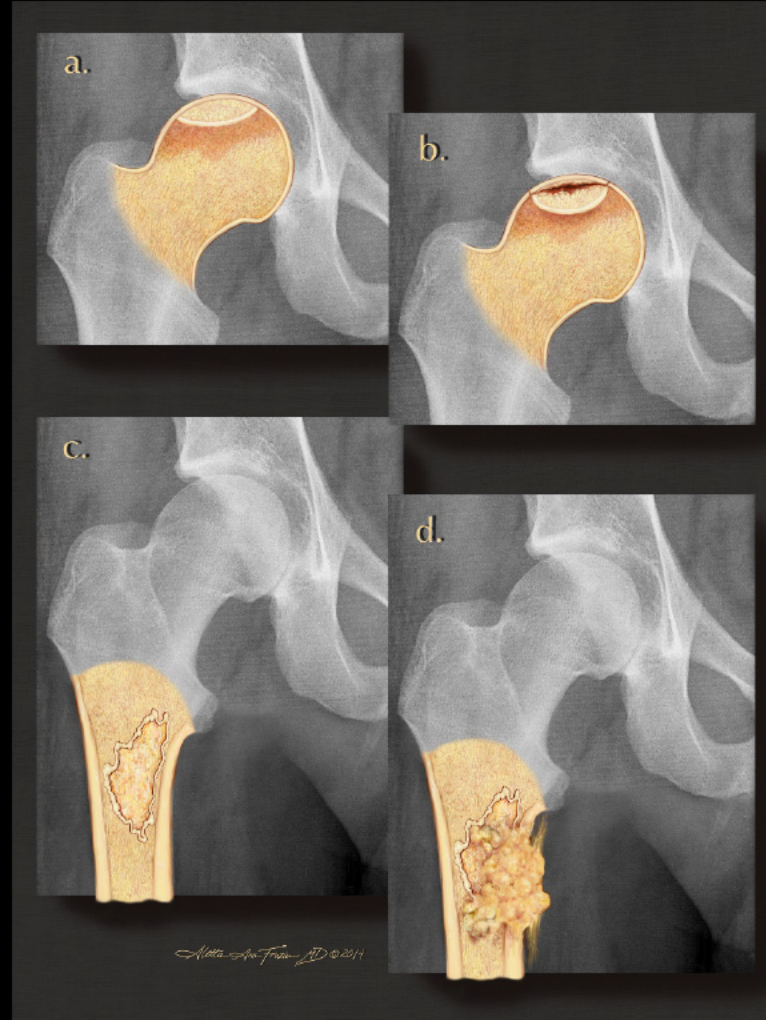
Louis Skidmore, M.D.
University of Pittsburgh
Pittsburgh, Pennsylvania

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Musculoskeletal Best Case



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**13 month old otherwise healthy
male presented initially with
slowly growing firm mass over 4
months along the ulnar aspect of
the right ring finger**



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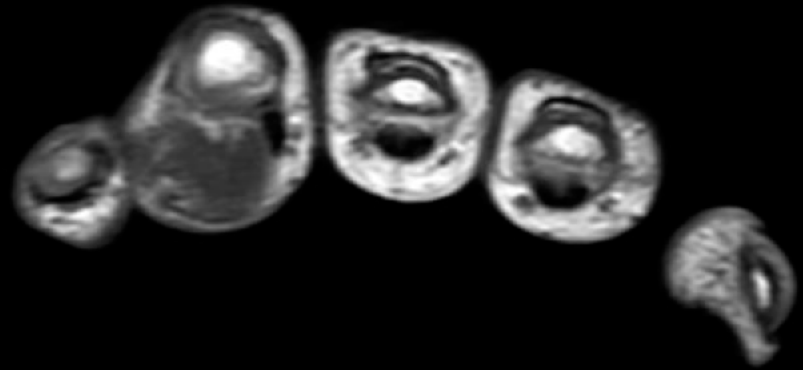
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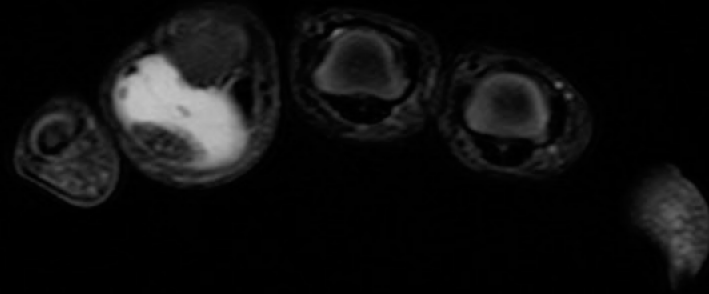
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6 months later





T1 AX



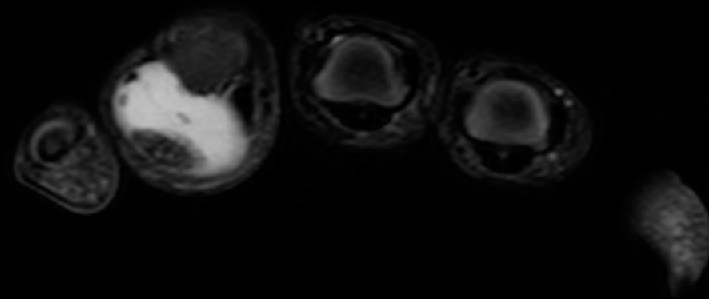
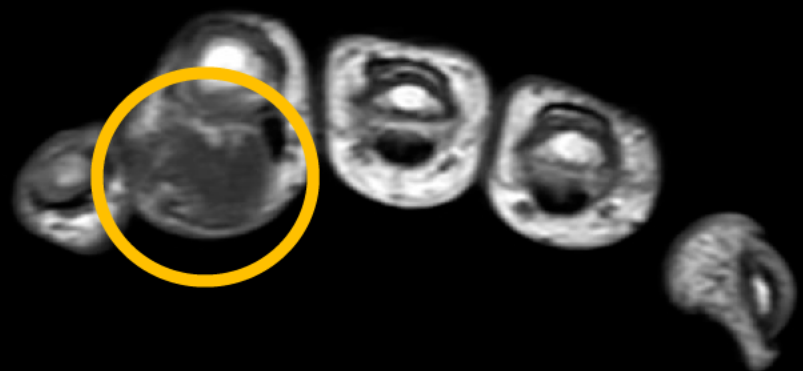
T2 FS



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T1 AX

T2 FS

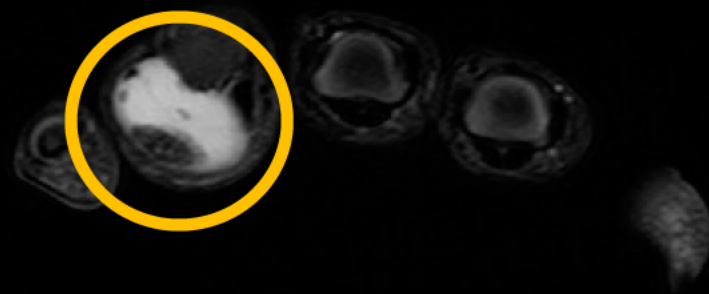


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T1 AX



T2 FS



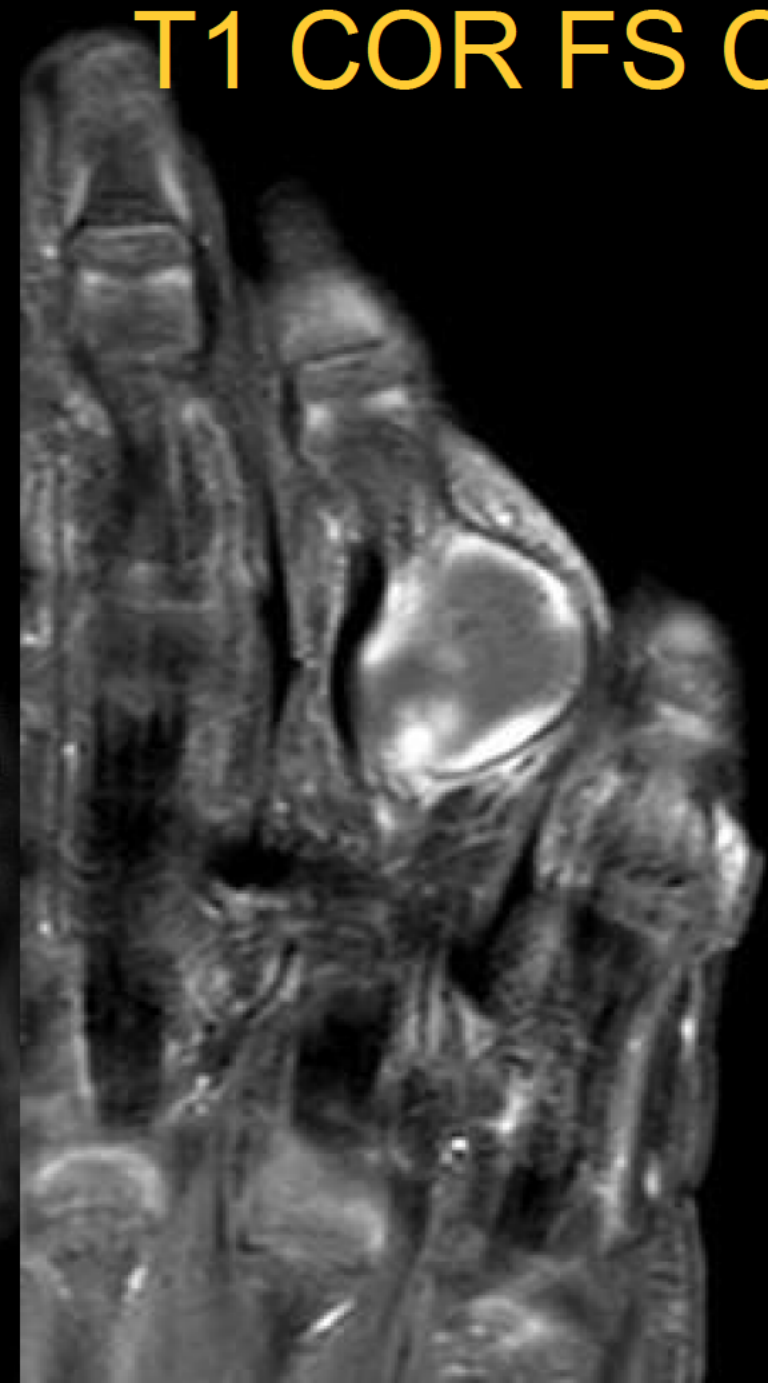
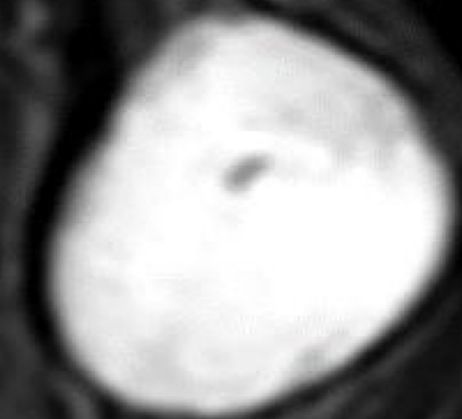
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T2 COR FS

T1 COR FS C+

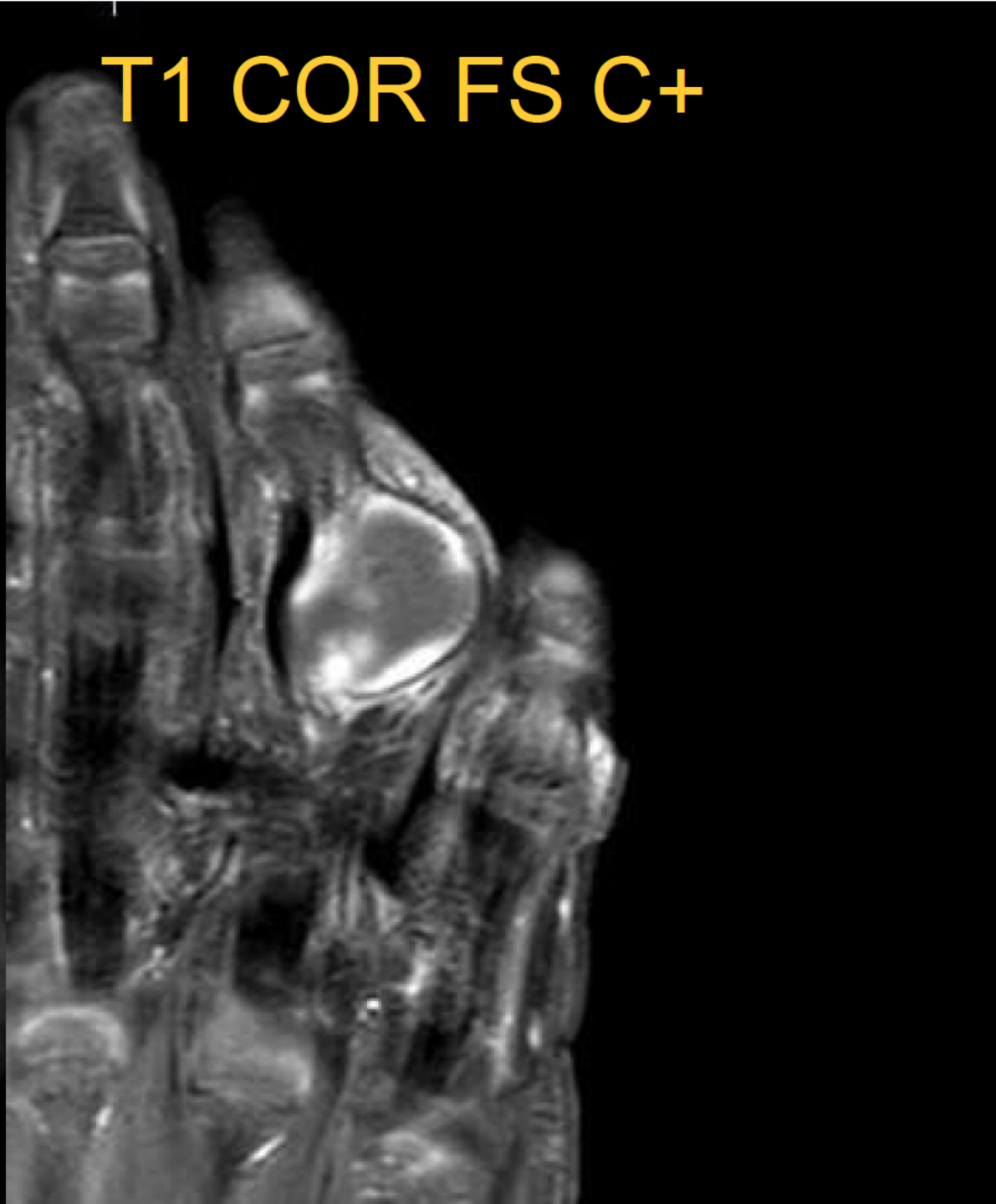


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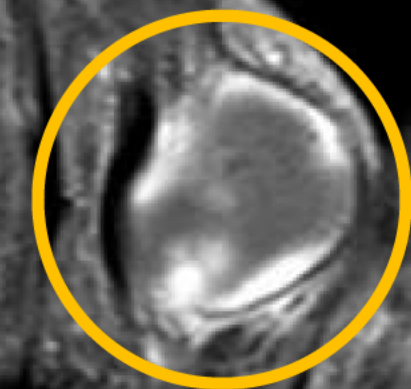
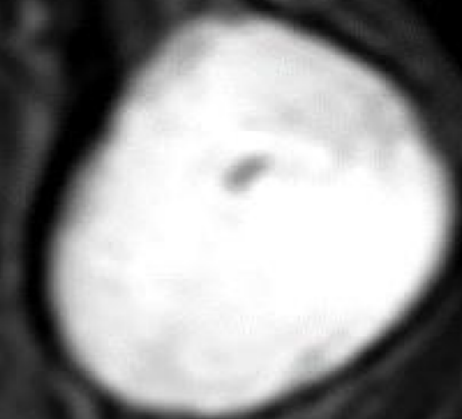
T2 COR FS

T1 COR FS C+

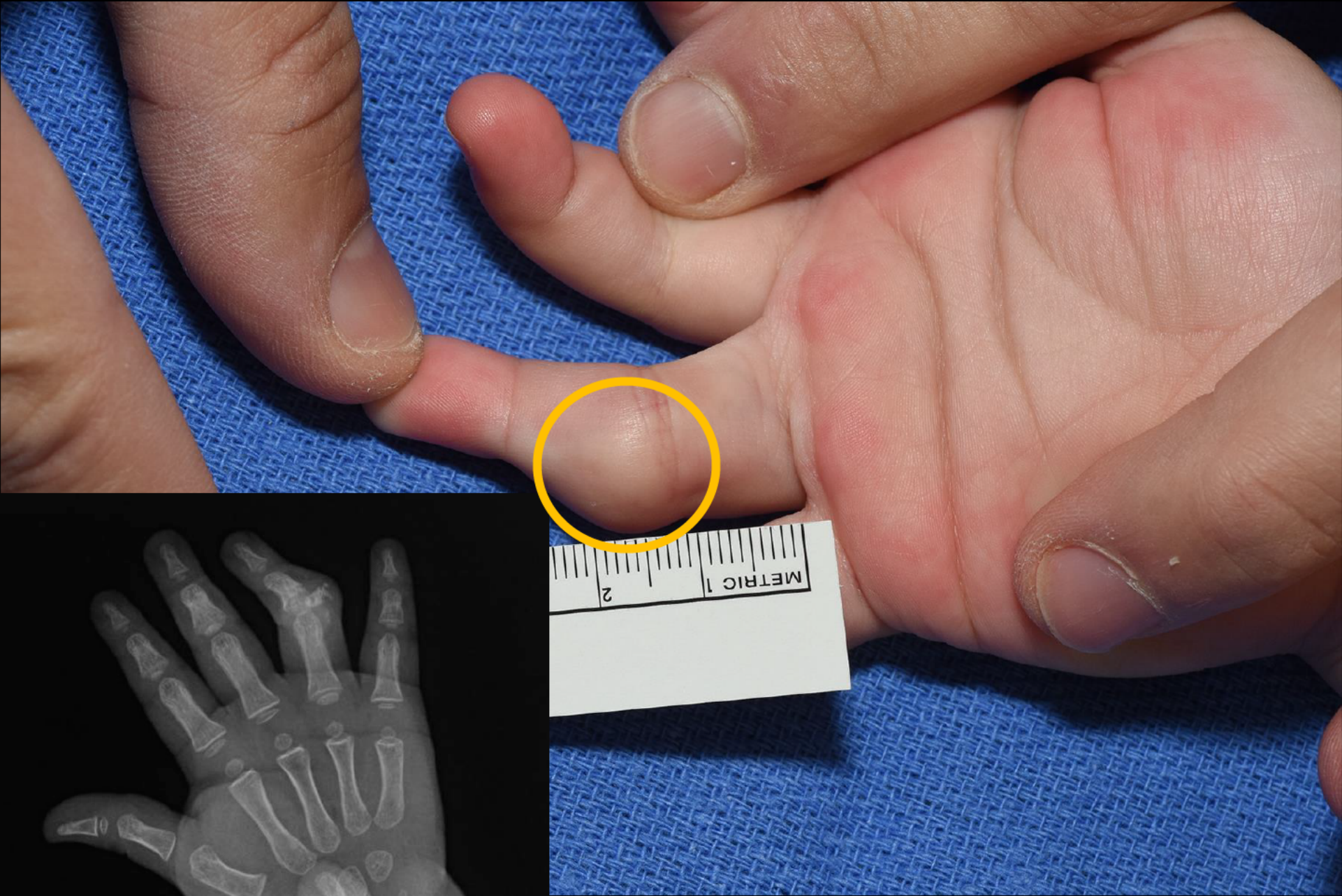


T2 COR FS

T1 COR FS C+





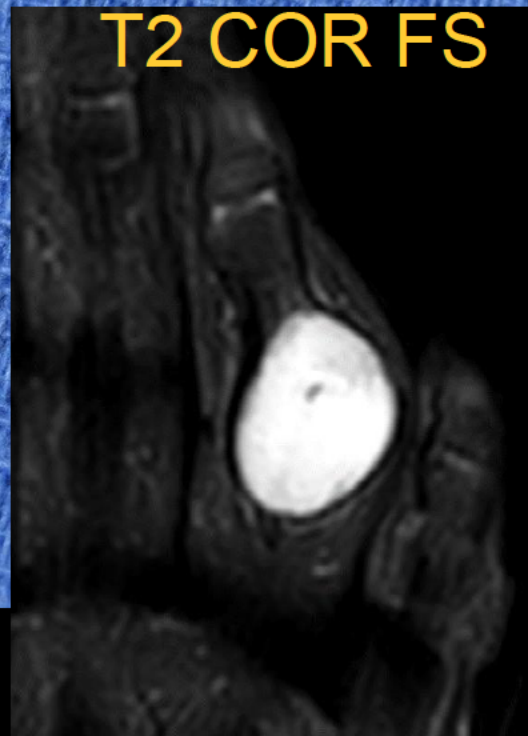
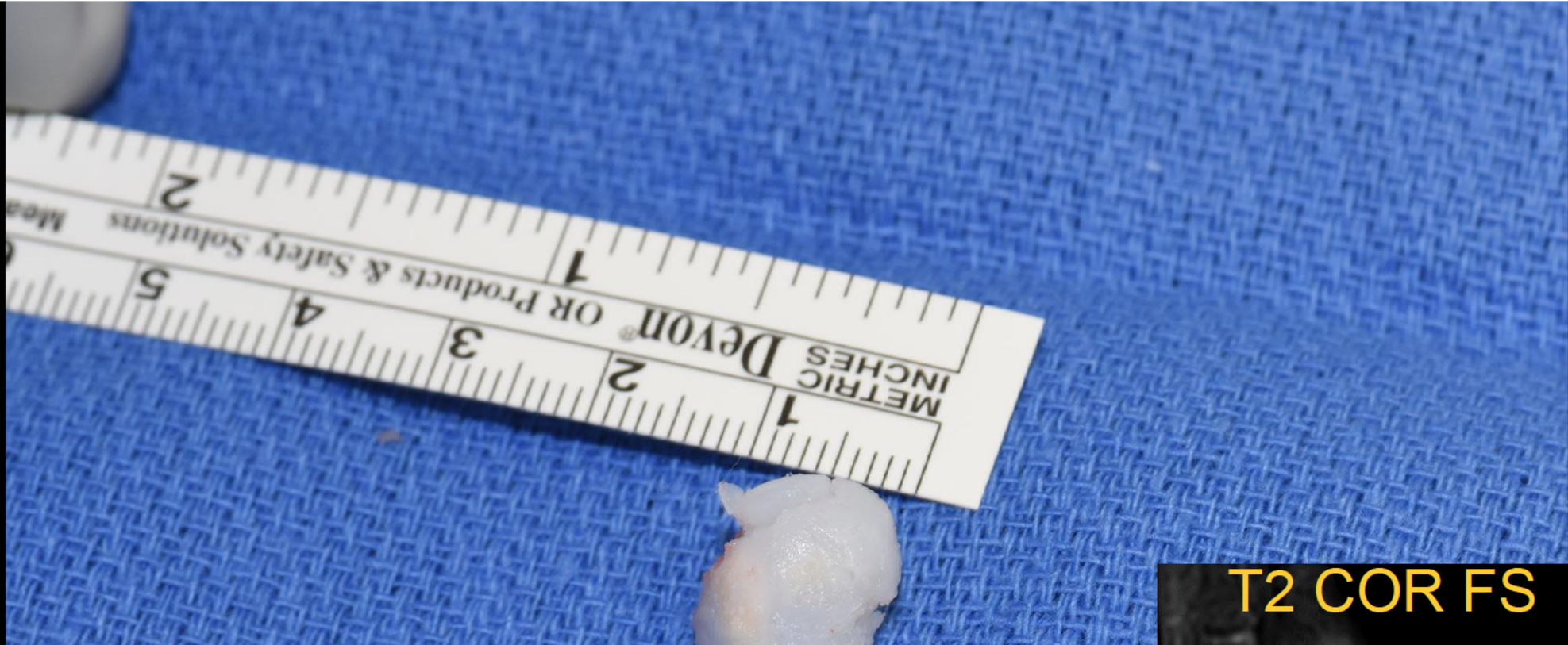


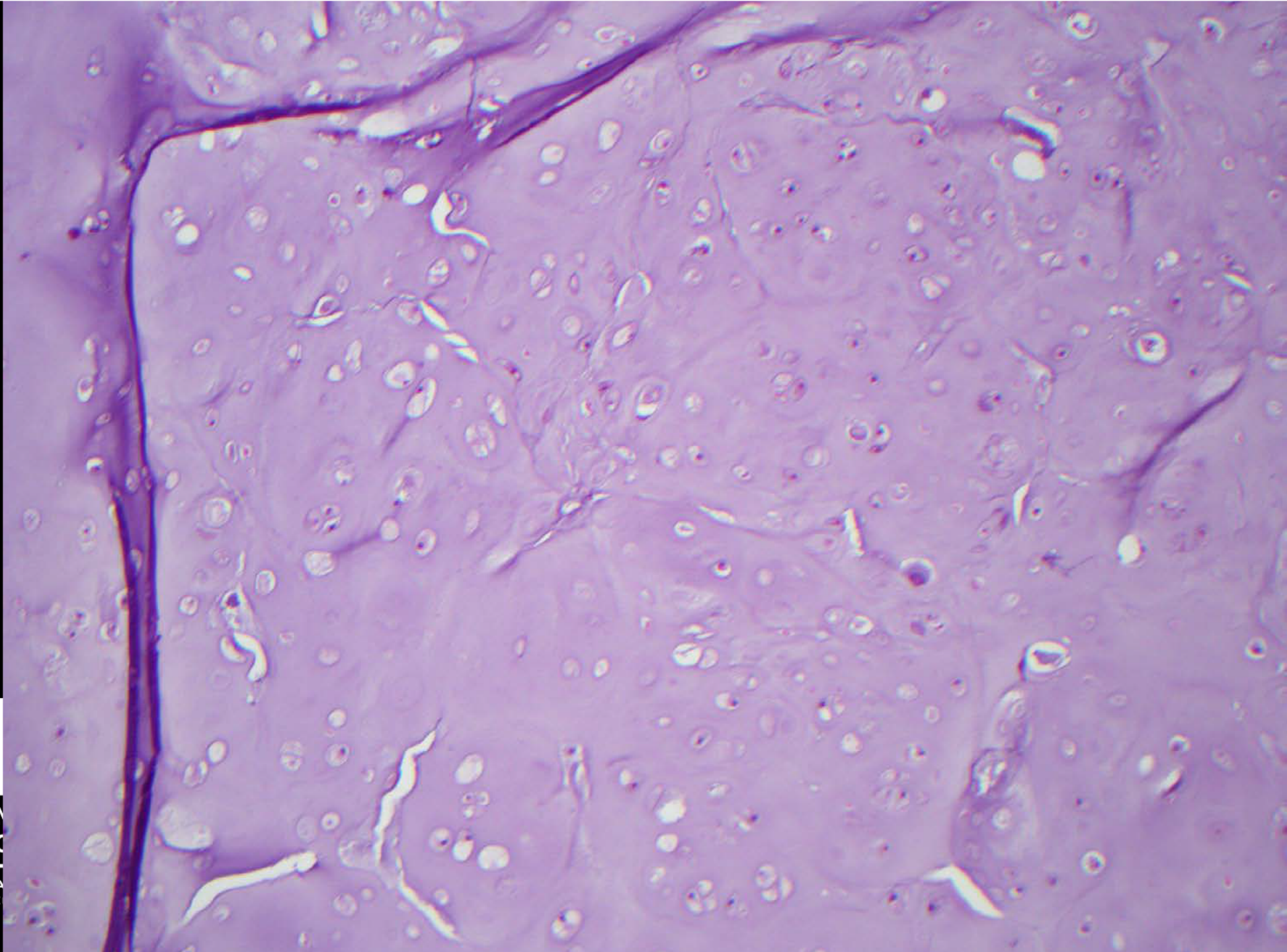
T2 COR FS



T2 COR FS



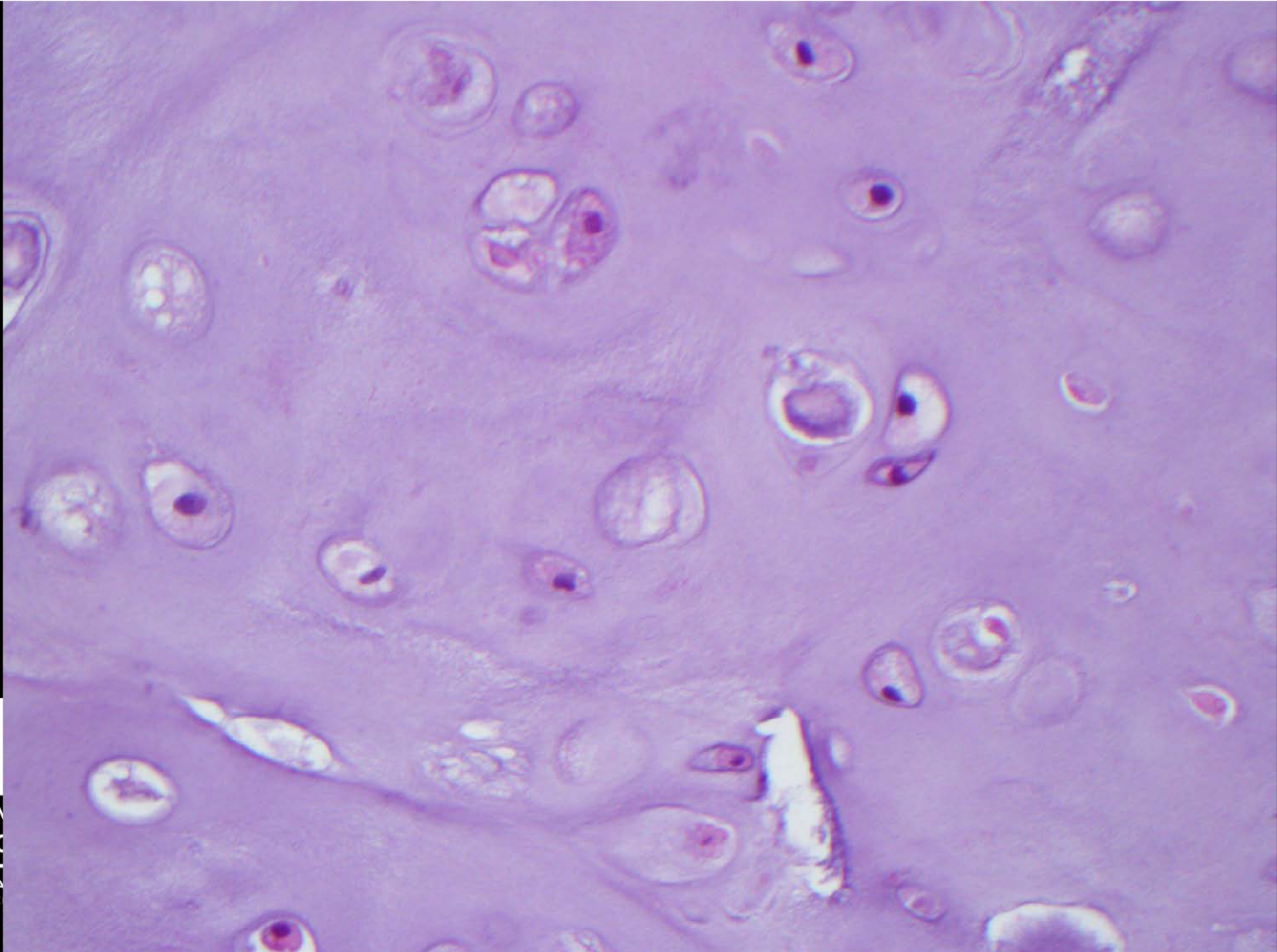




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SOFT TISSUE CHONDROMA

Dr. Matthew Burgess
Naval Medical Center
San Diego, CA

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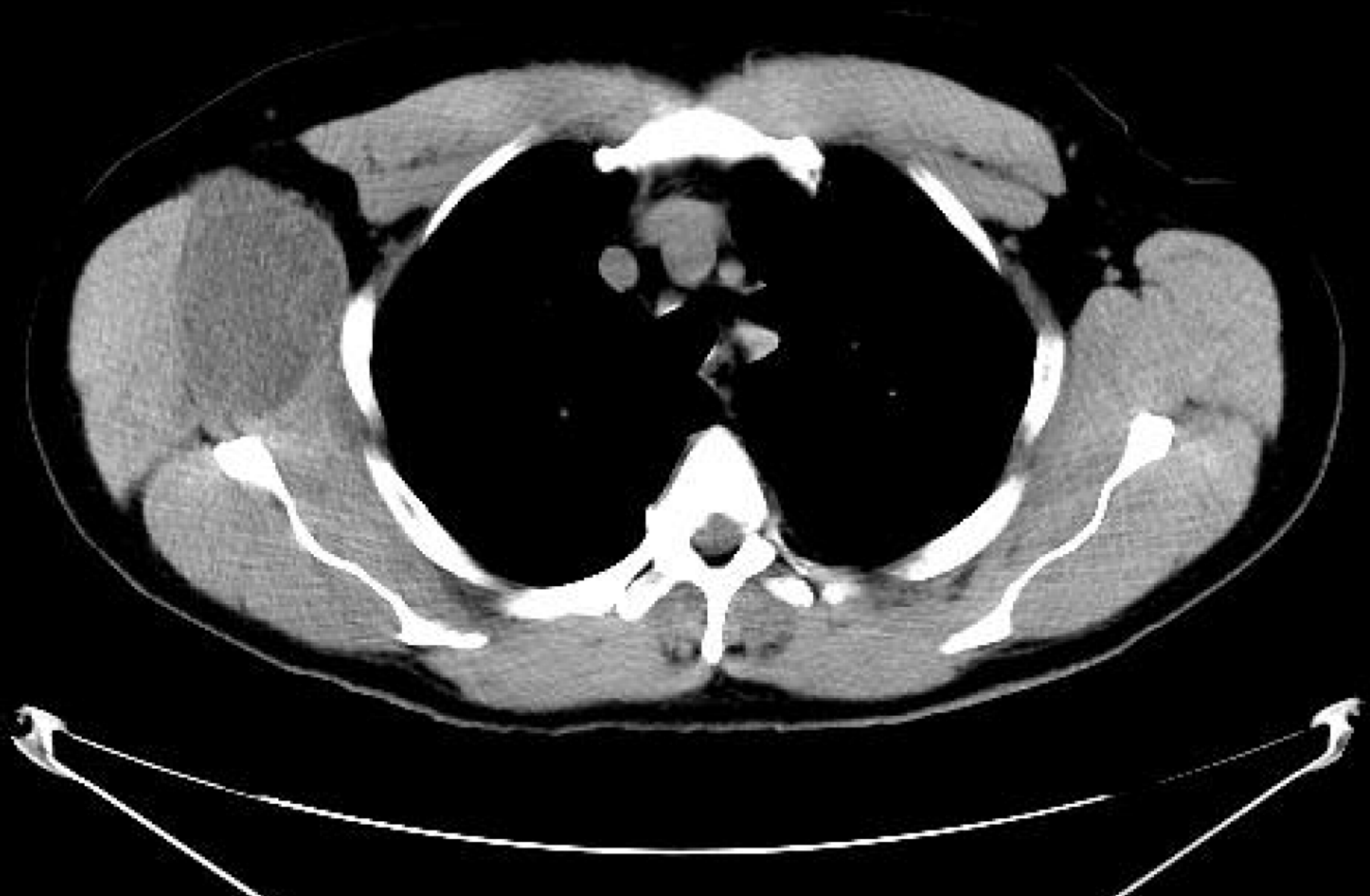
Honorable mention



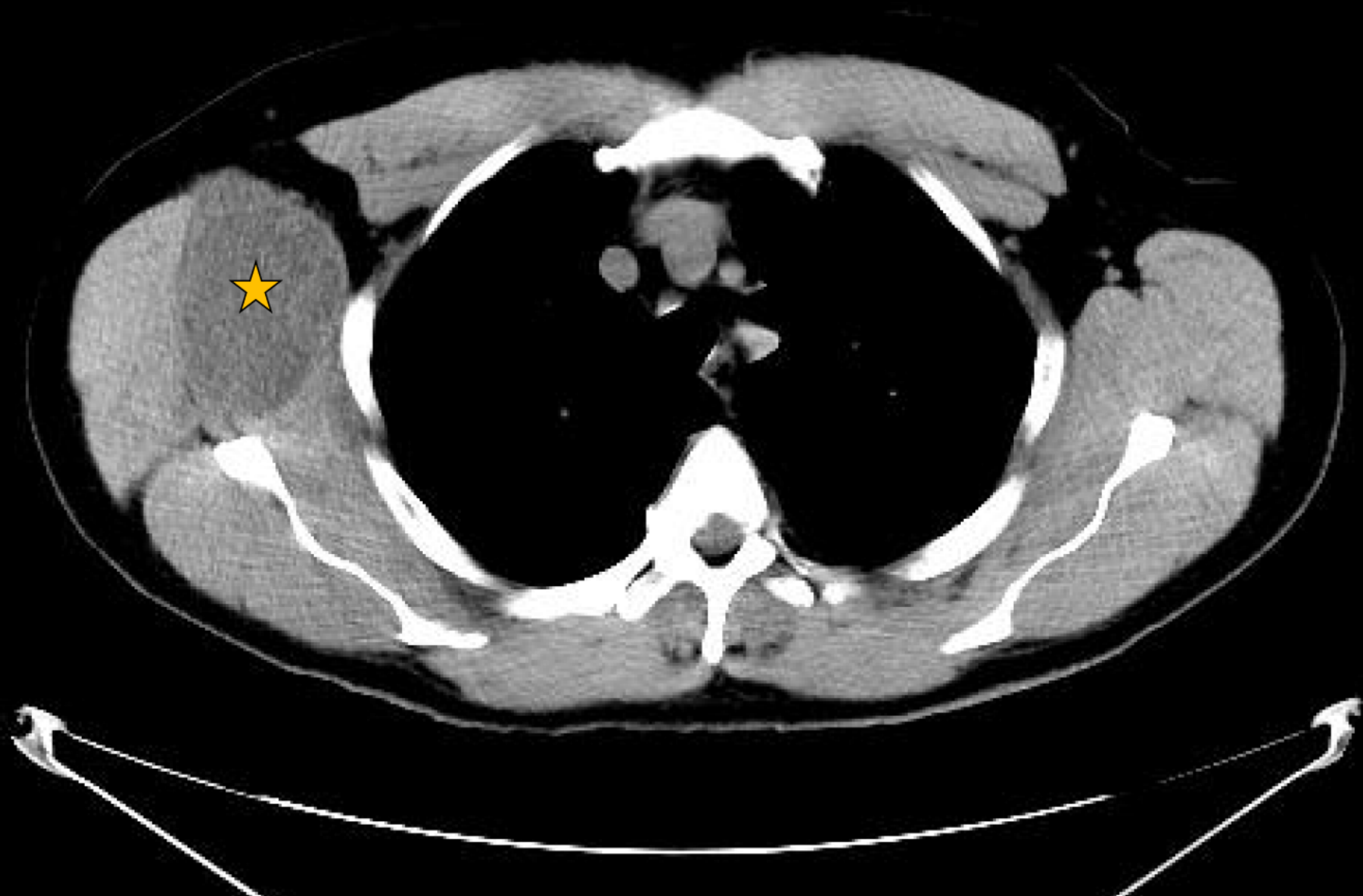
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**62 year old African American
male mass in his right axilla
causing decreased shoulder
range of motion without
associated pain**

Axial CT



Axial CT



CT SAG



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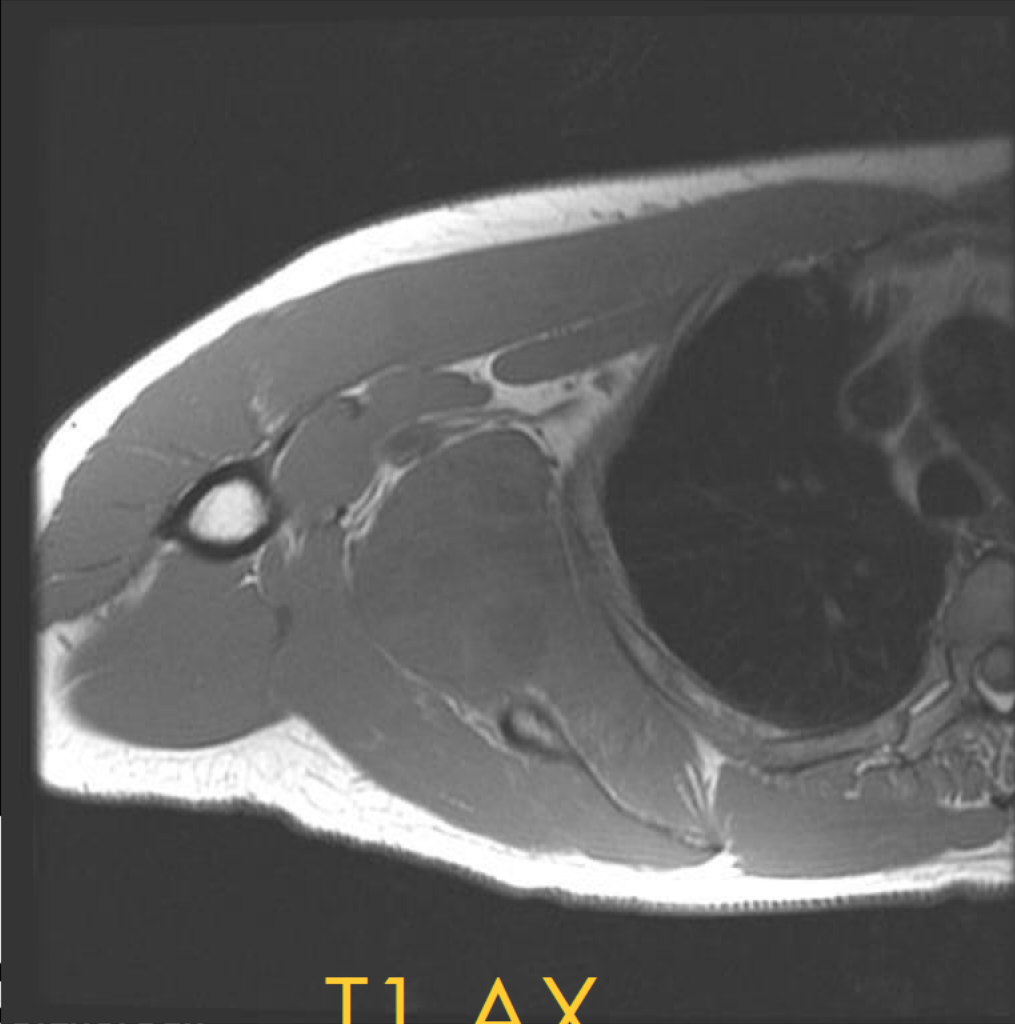
CT SAG



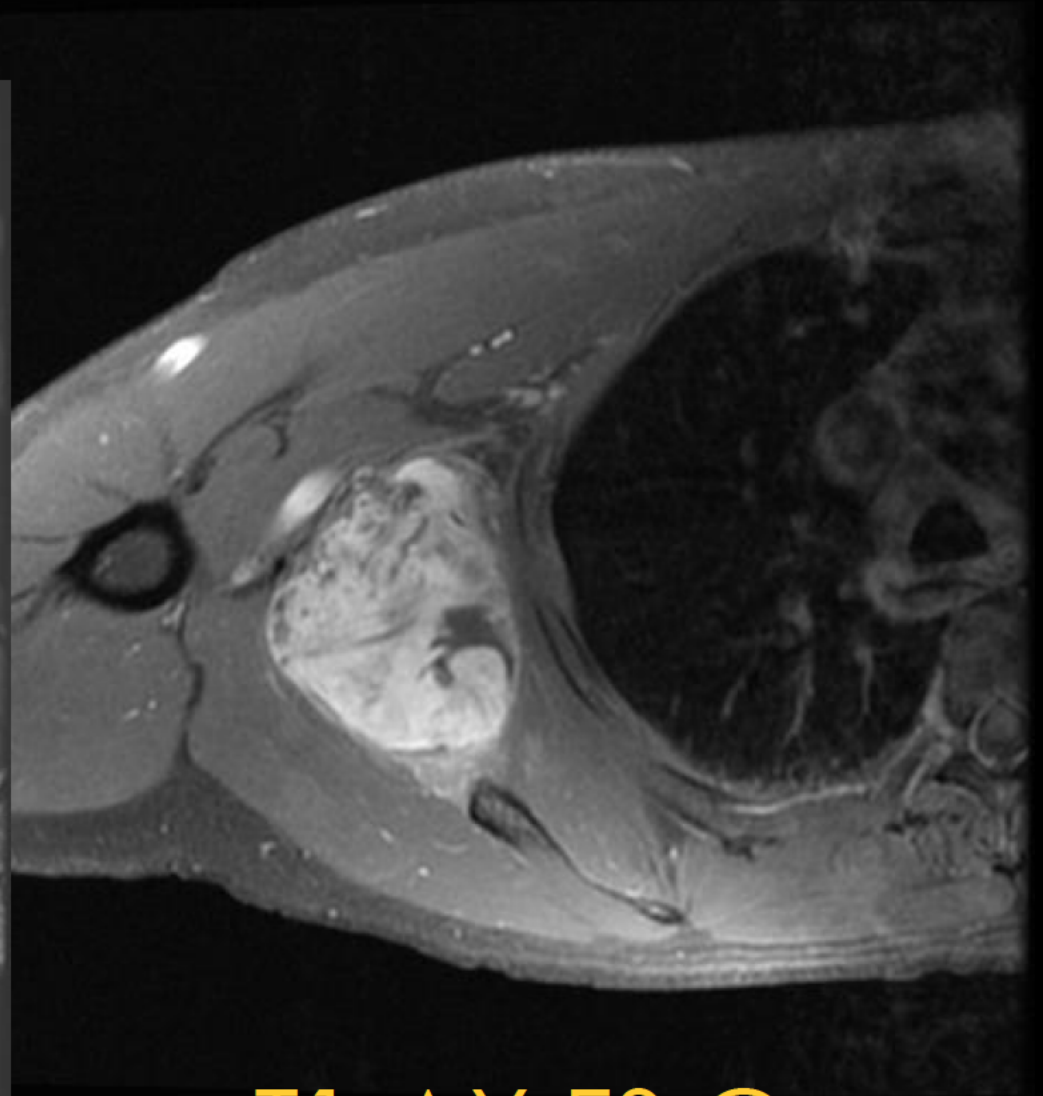
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T1 AX

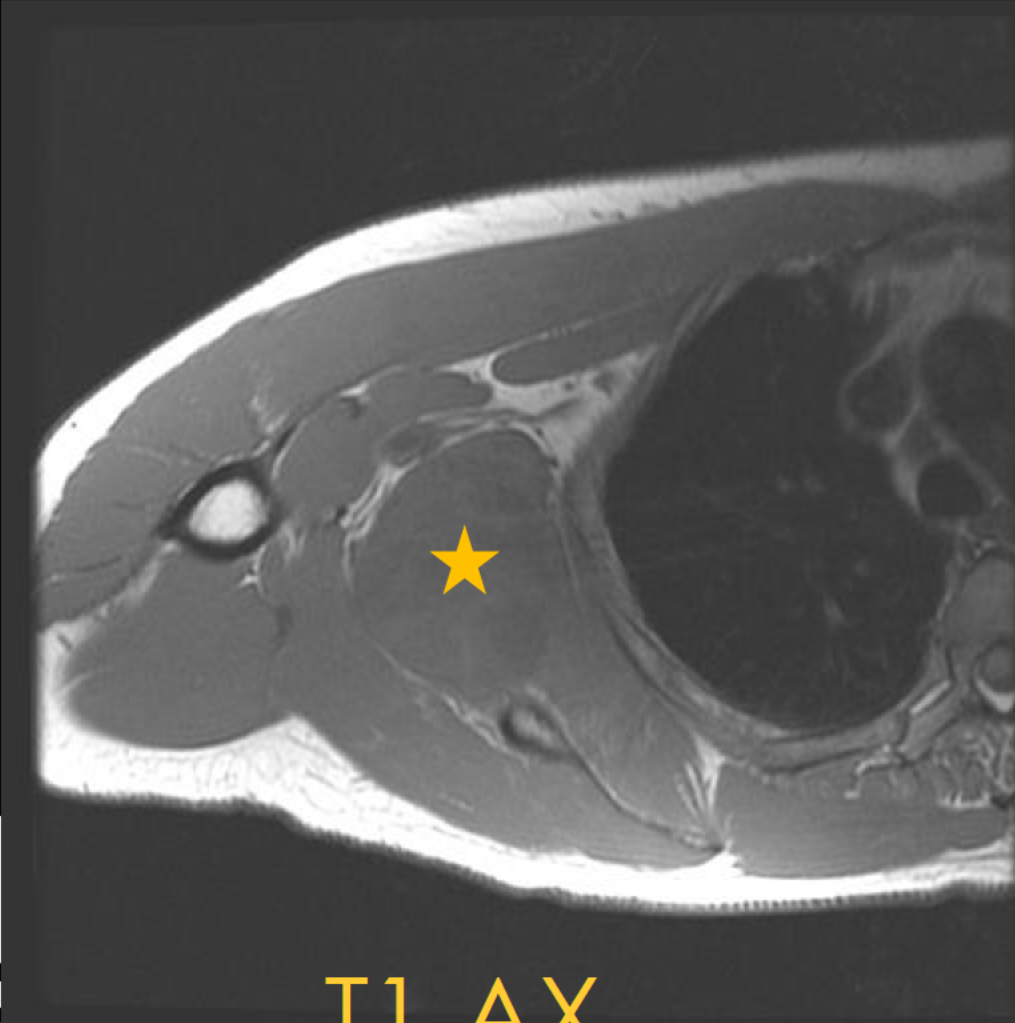


T1 AX FS C+

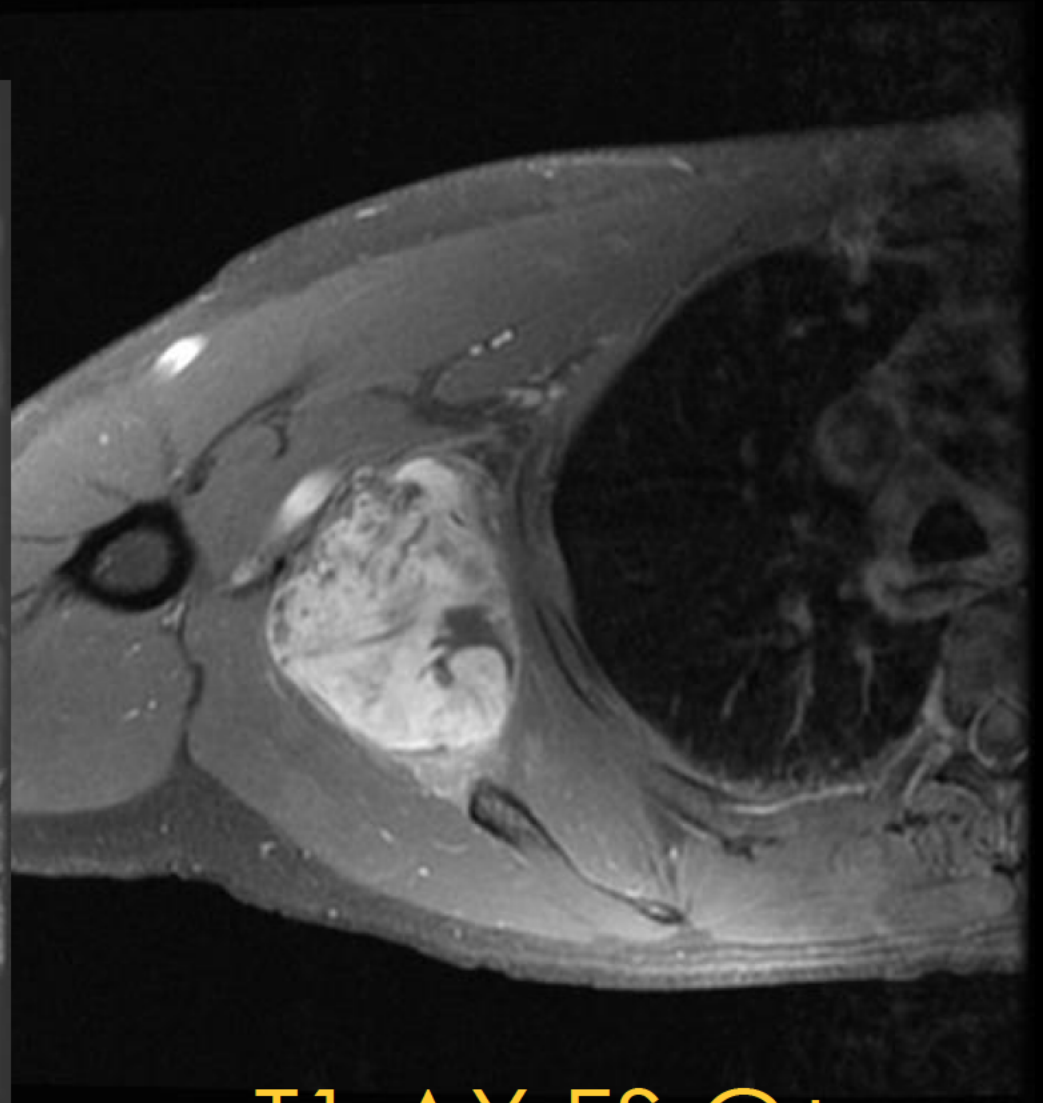
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T1 AX

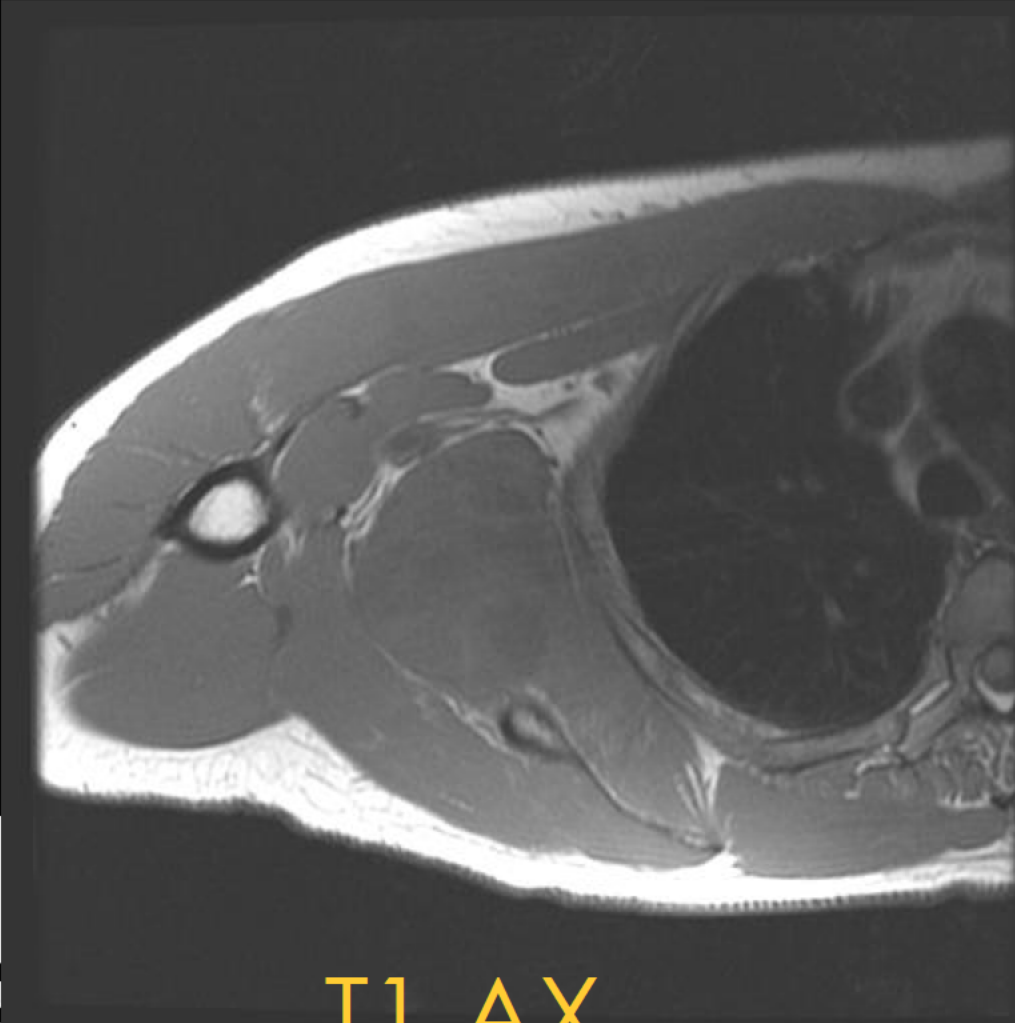


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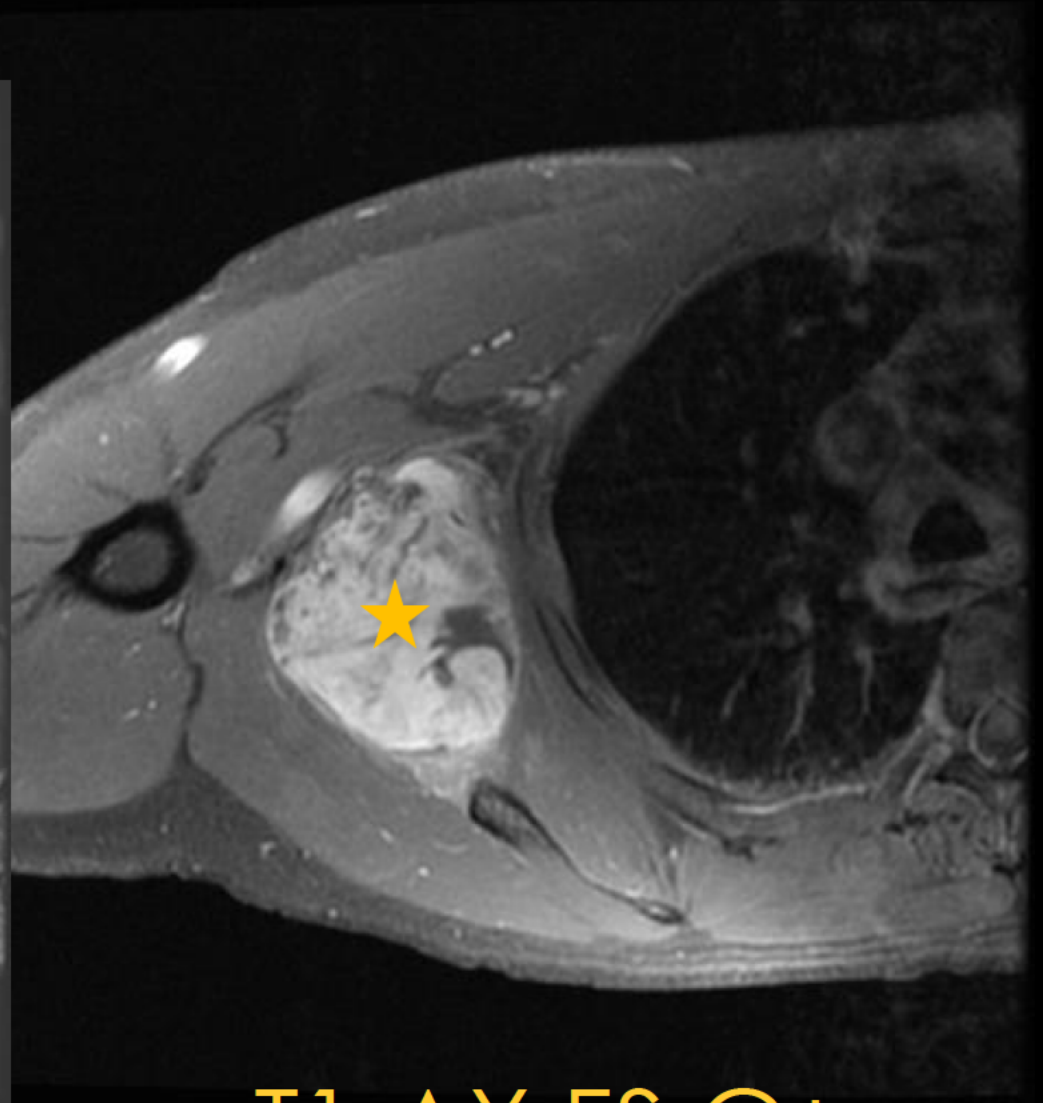
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T1 AX

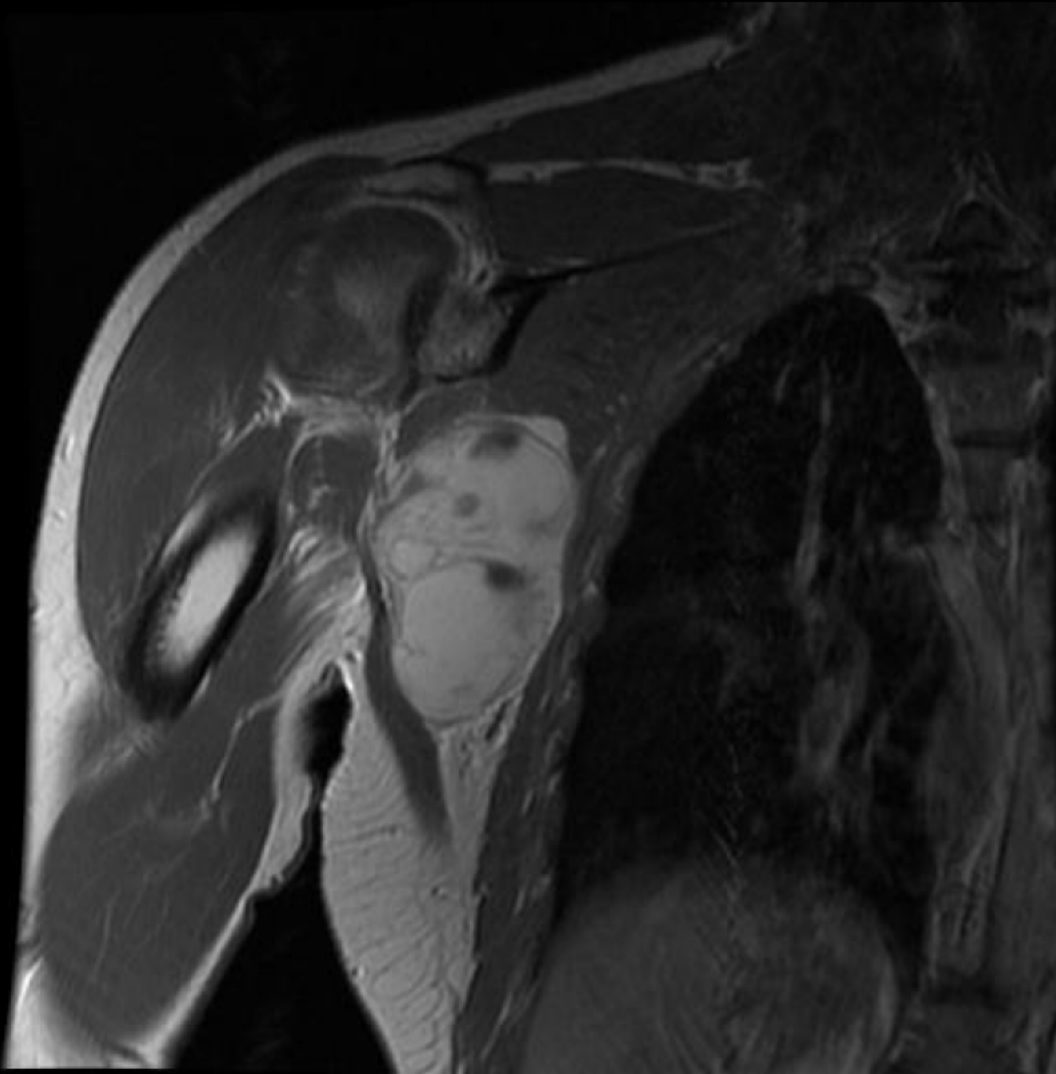


T1 AX FS C+

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T1 COR C+

T2 COR



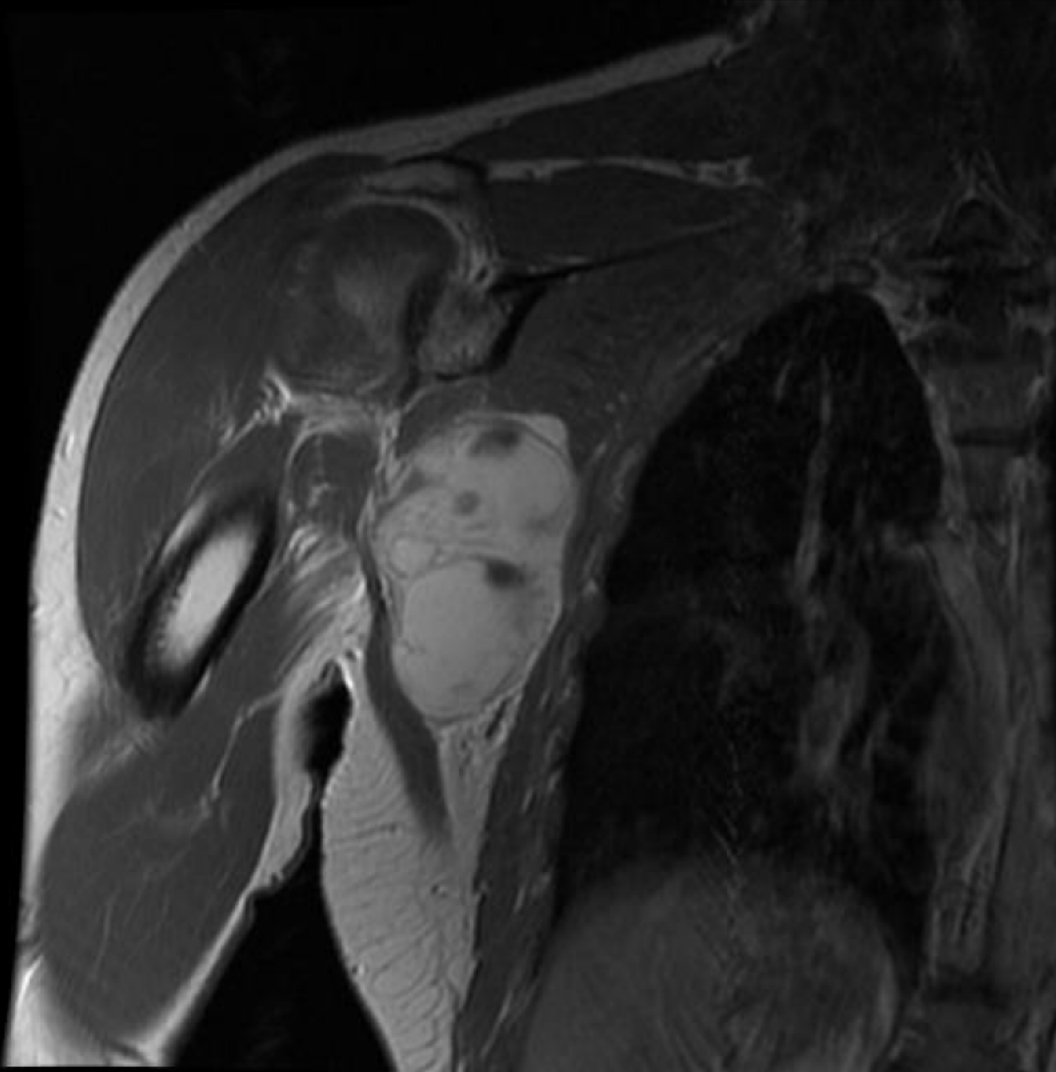
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T1 COR C+

T2 COR



T1 COR C+



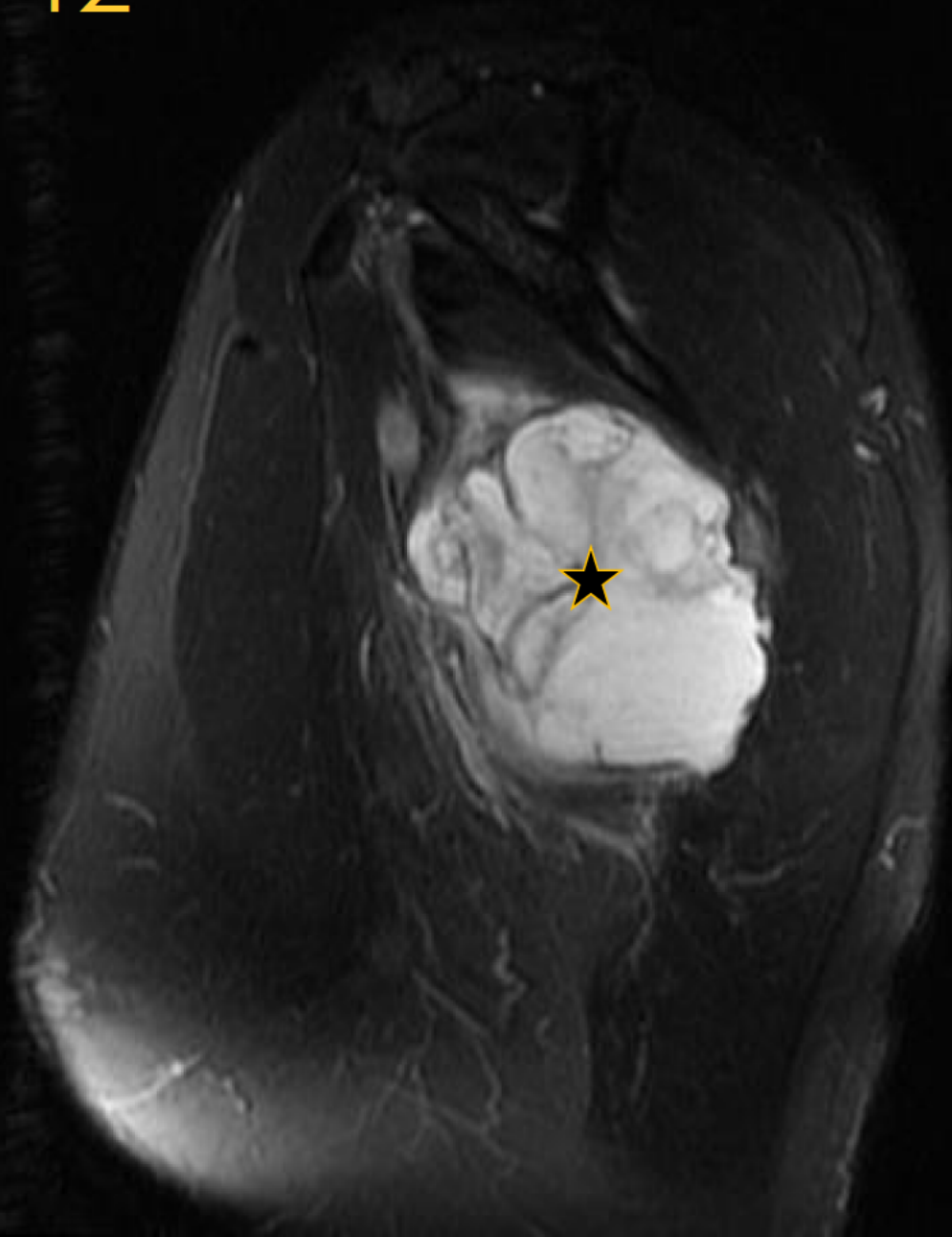
T2 COR

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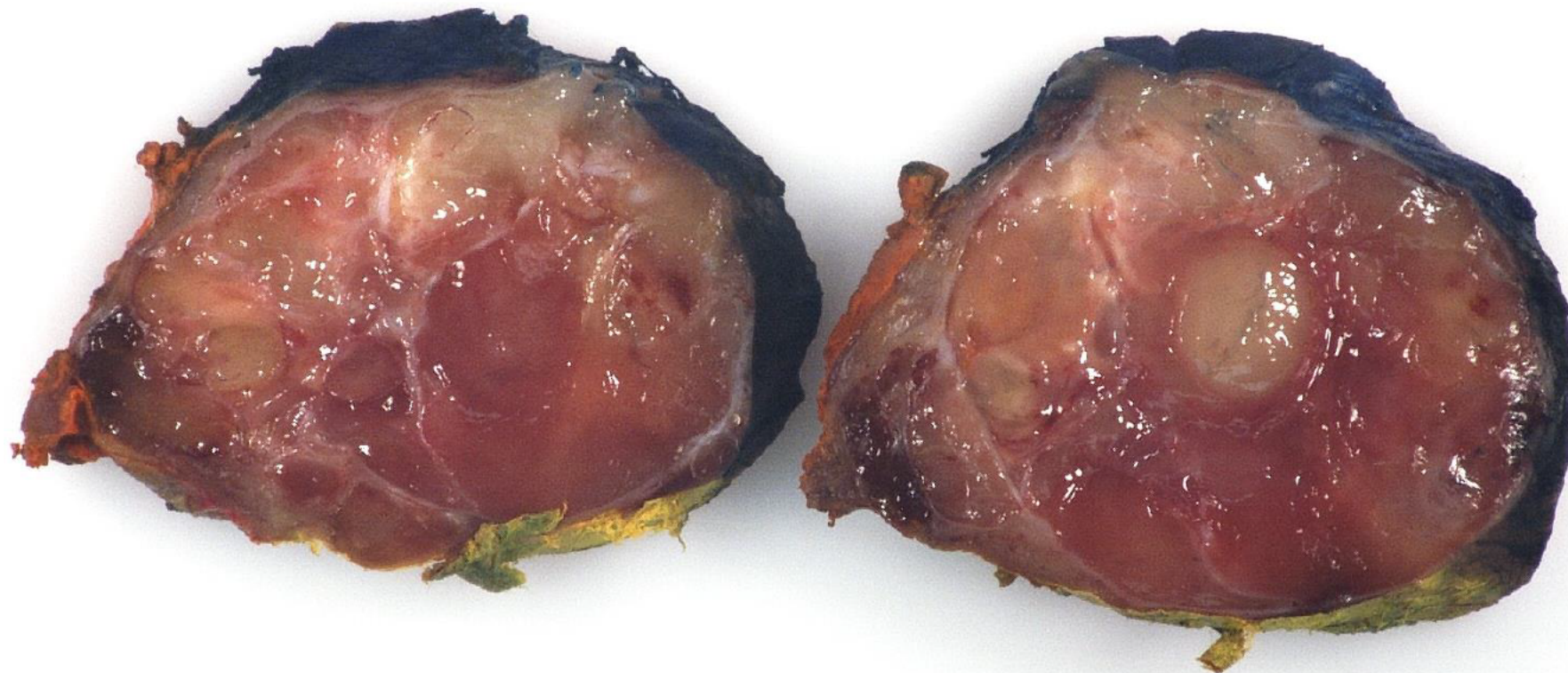
SAG T2



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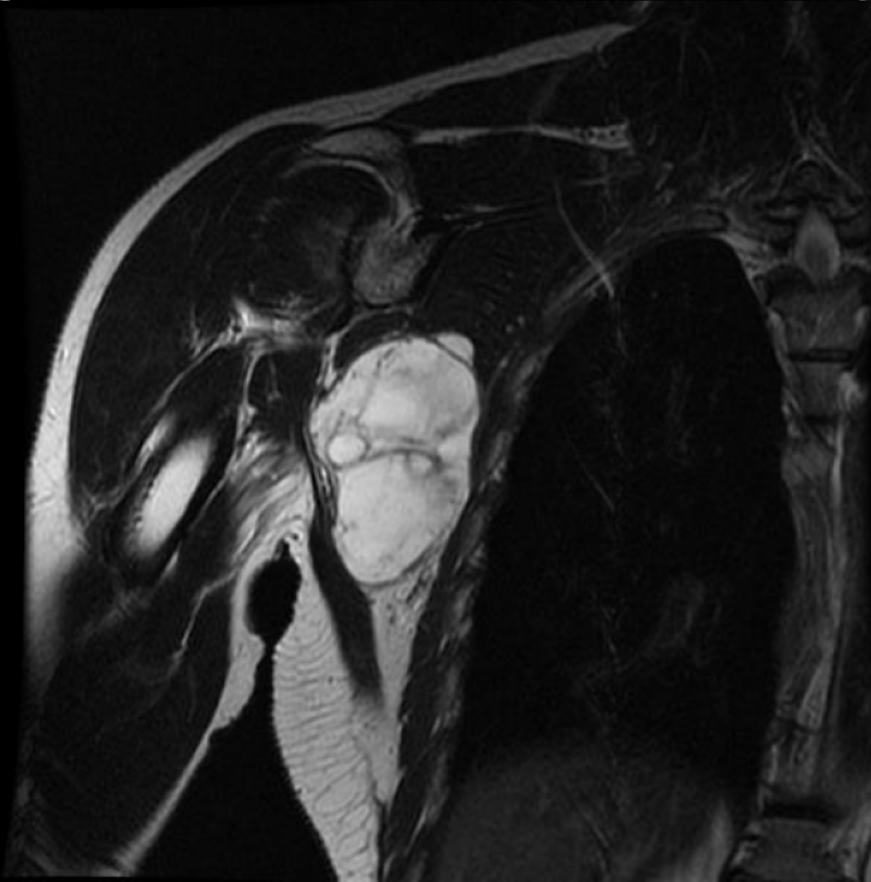
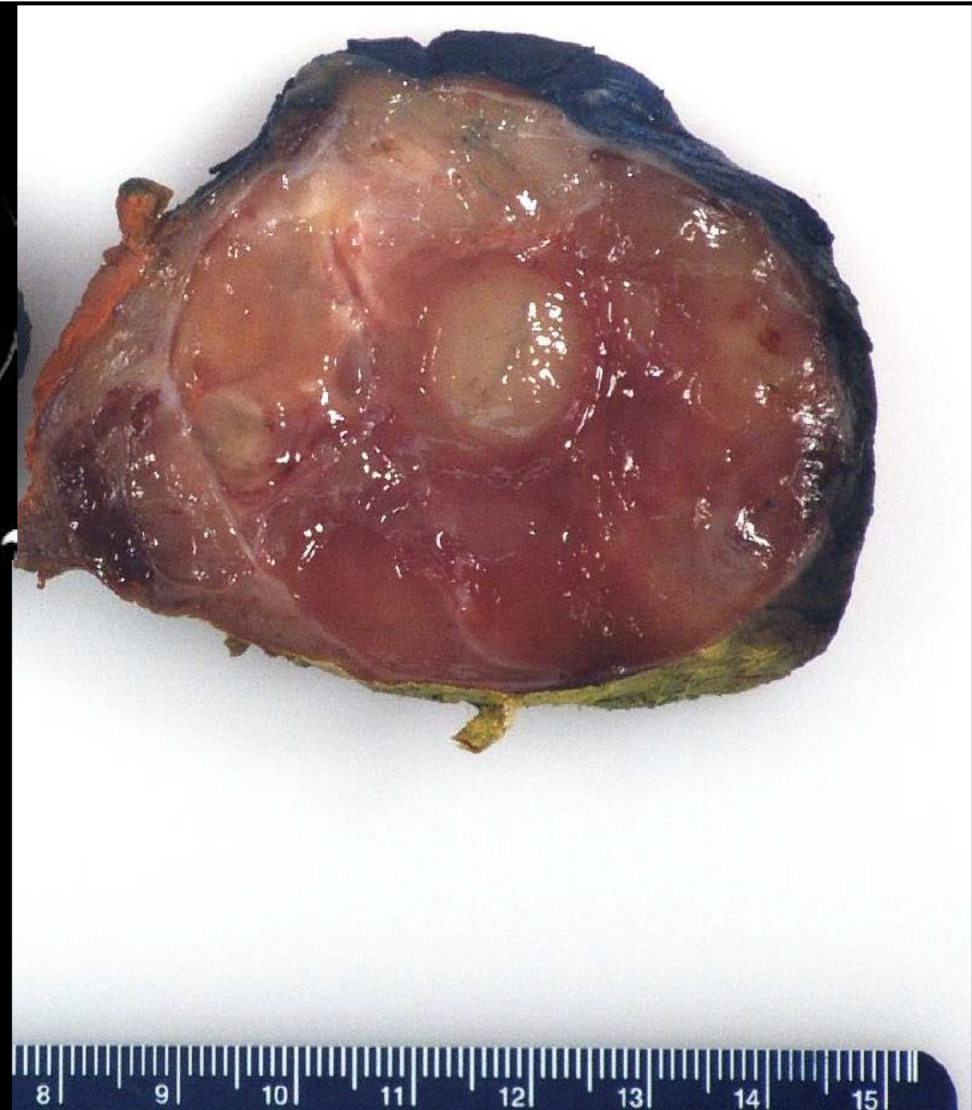
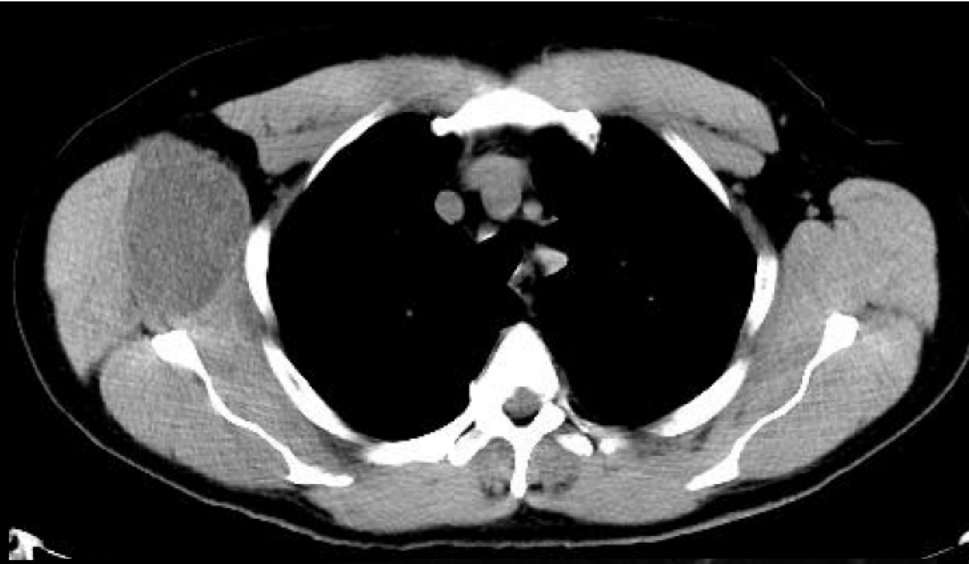
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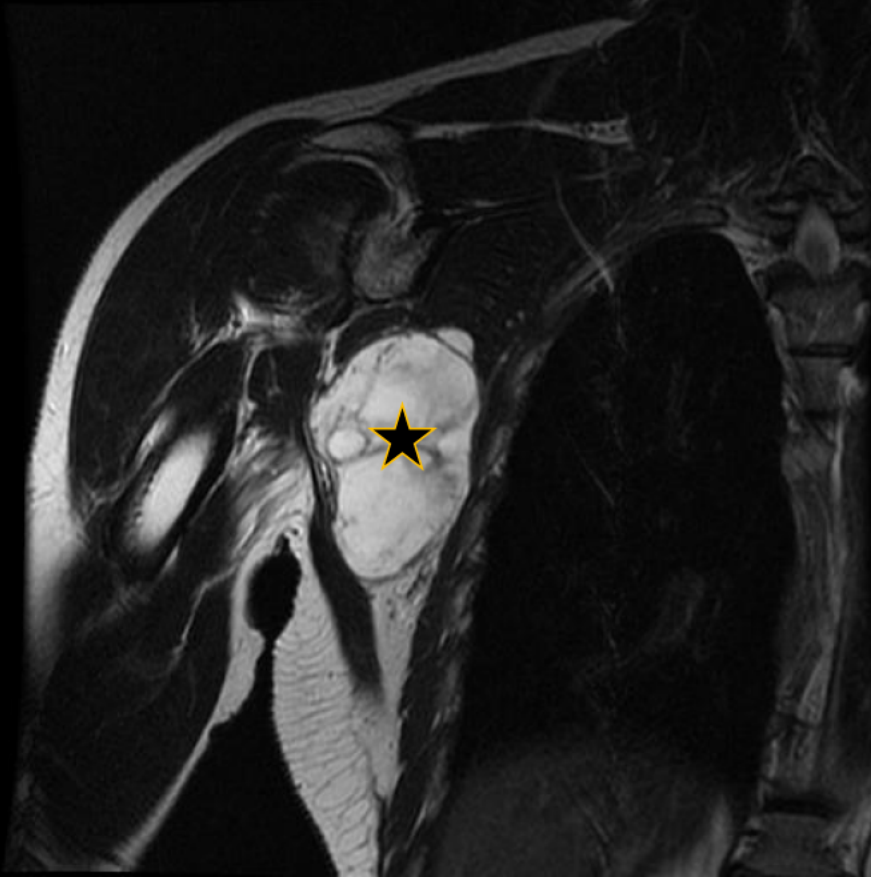
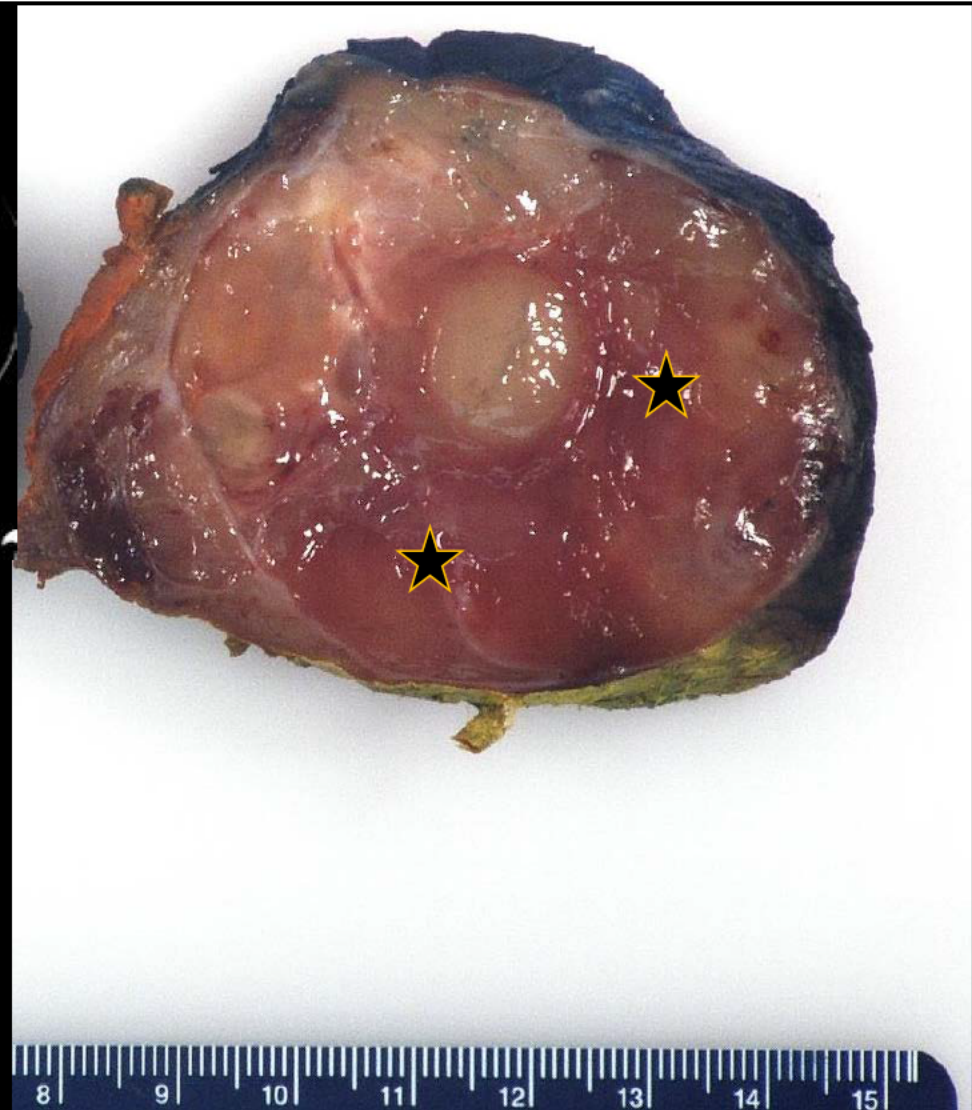


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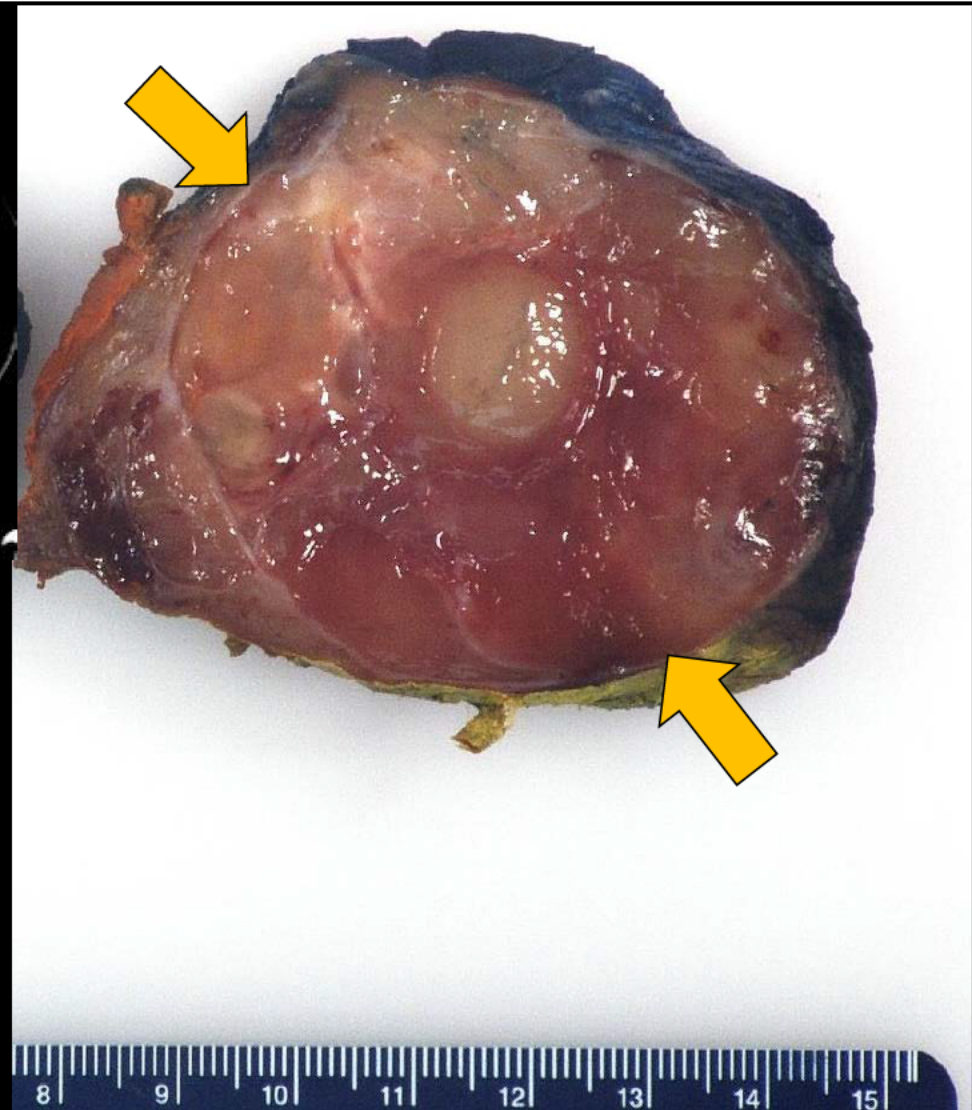
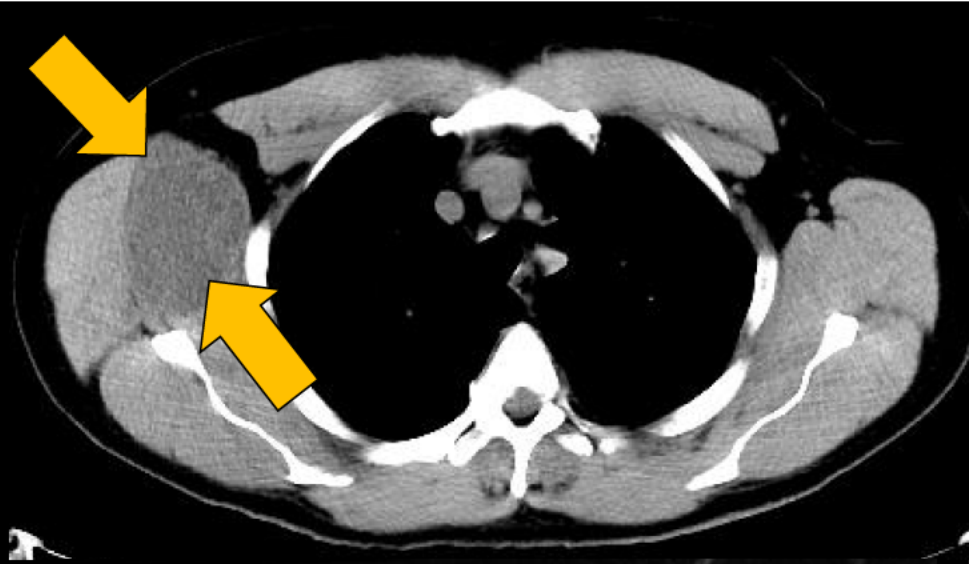
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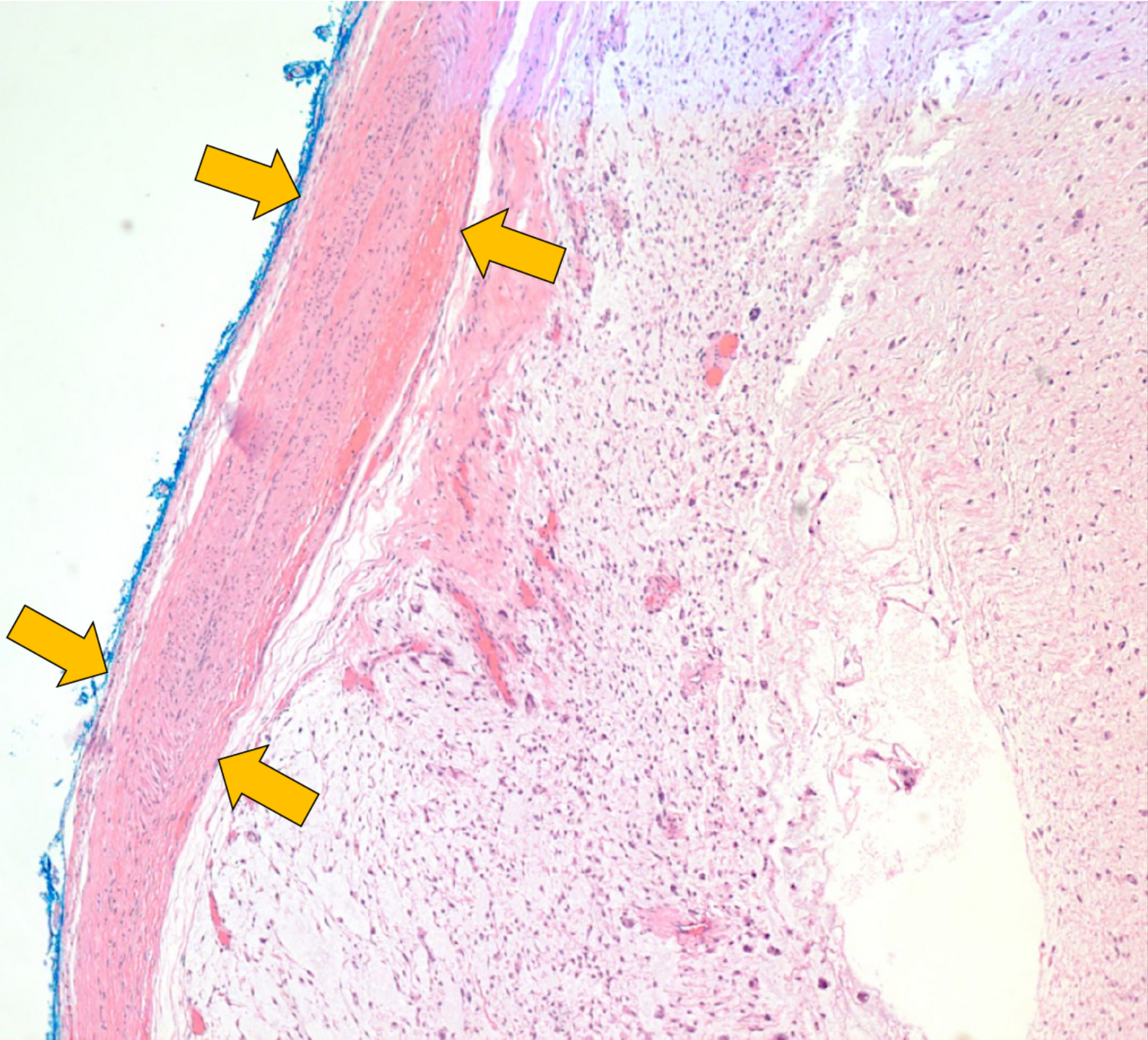
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T2 COR



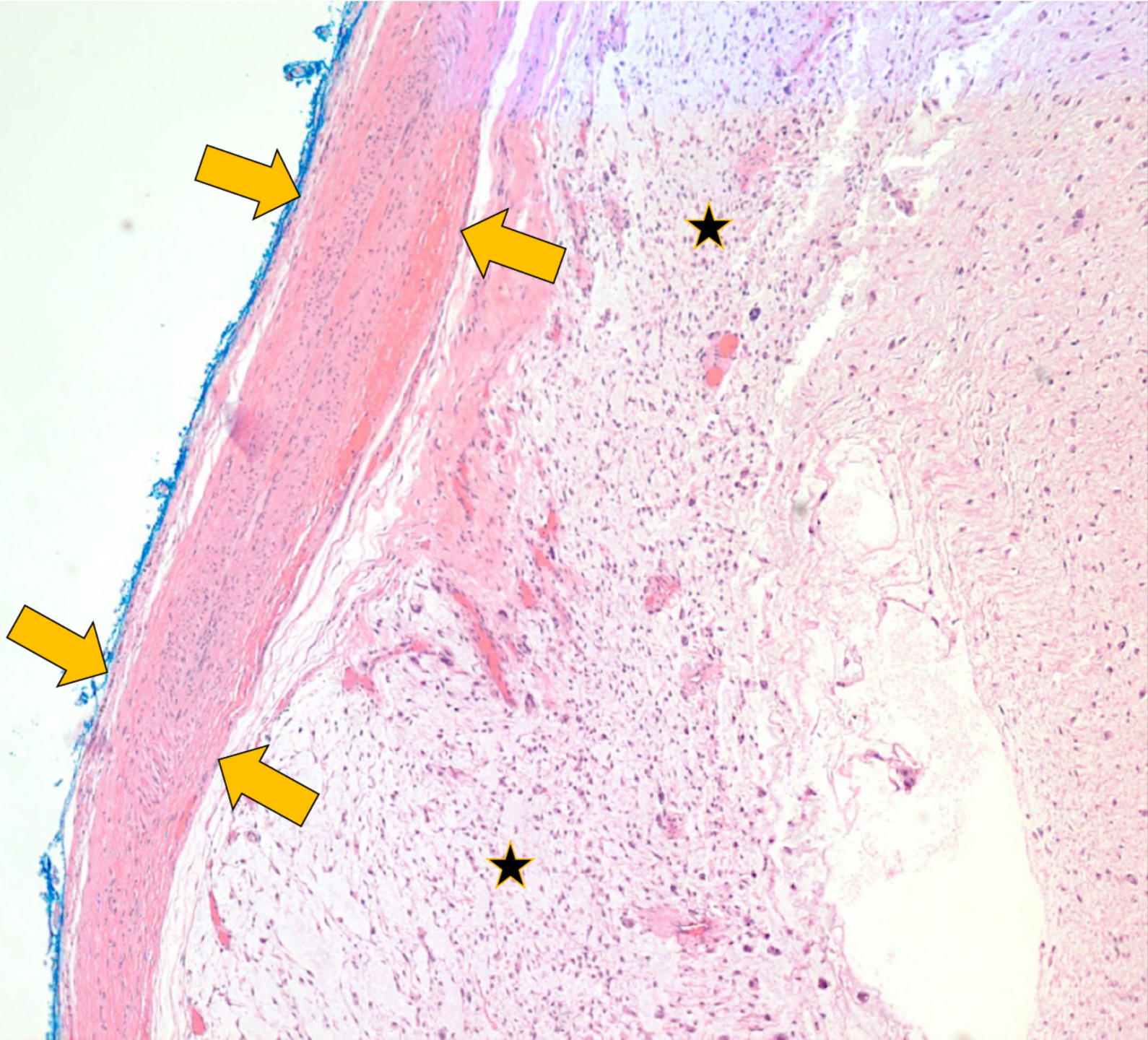
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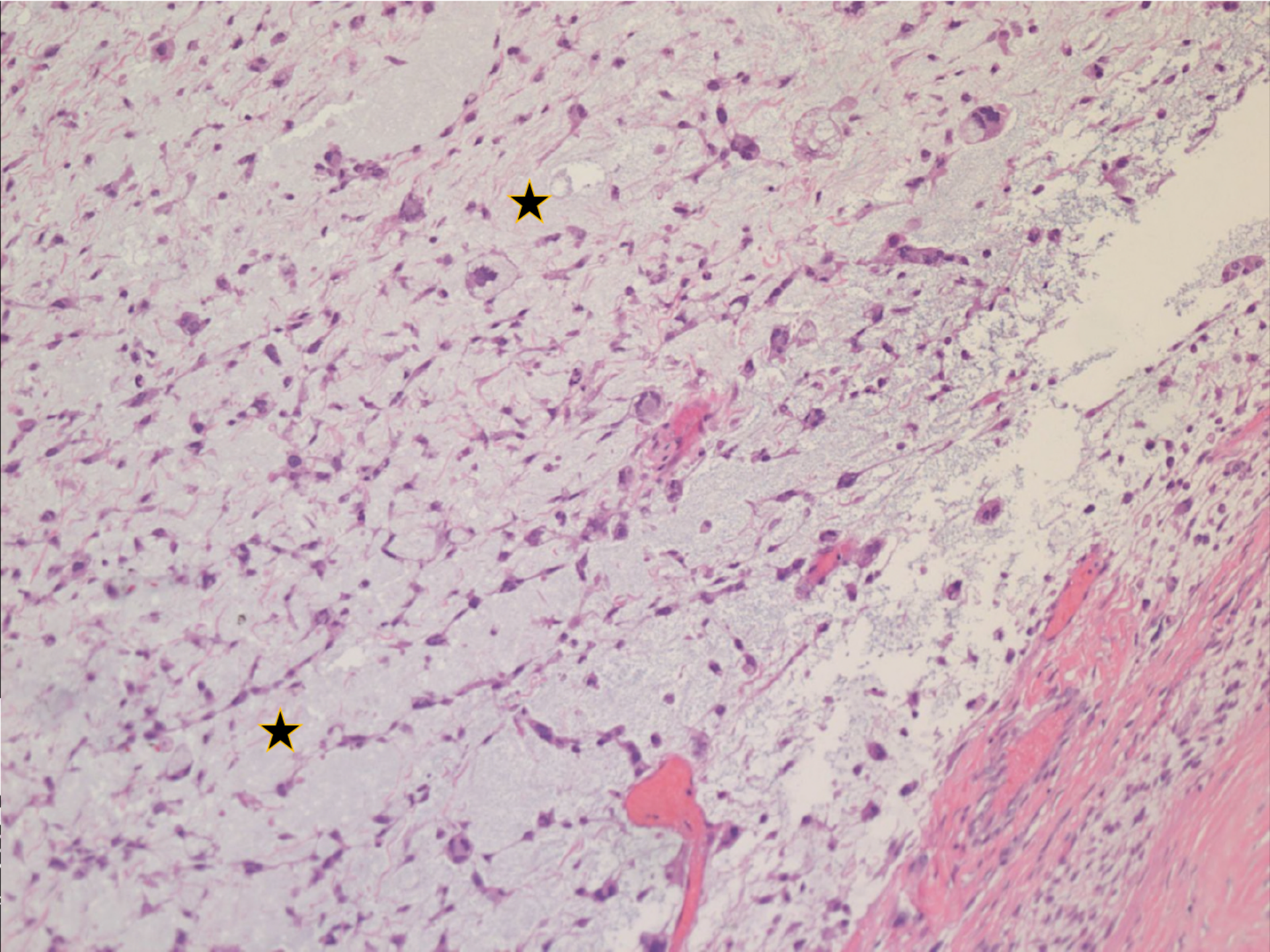


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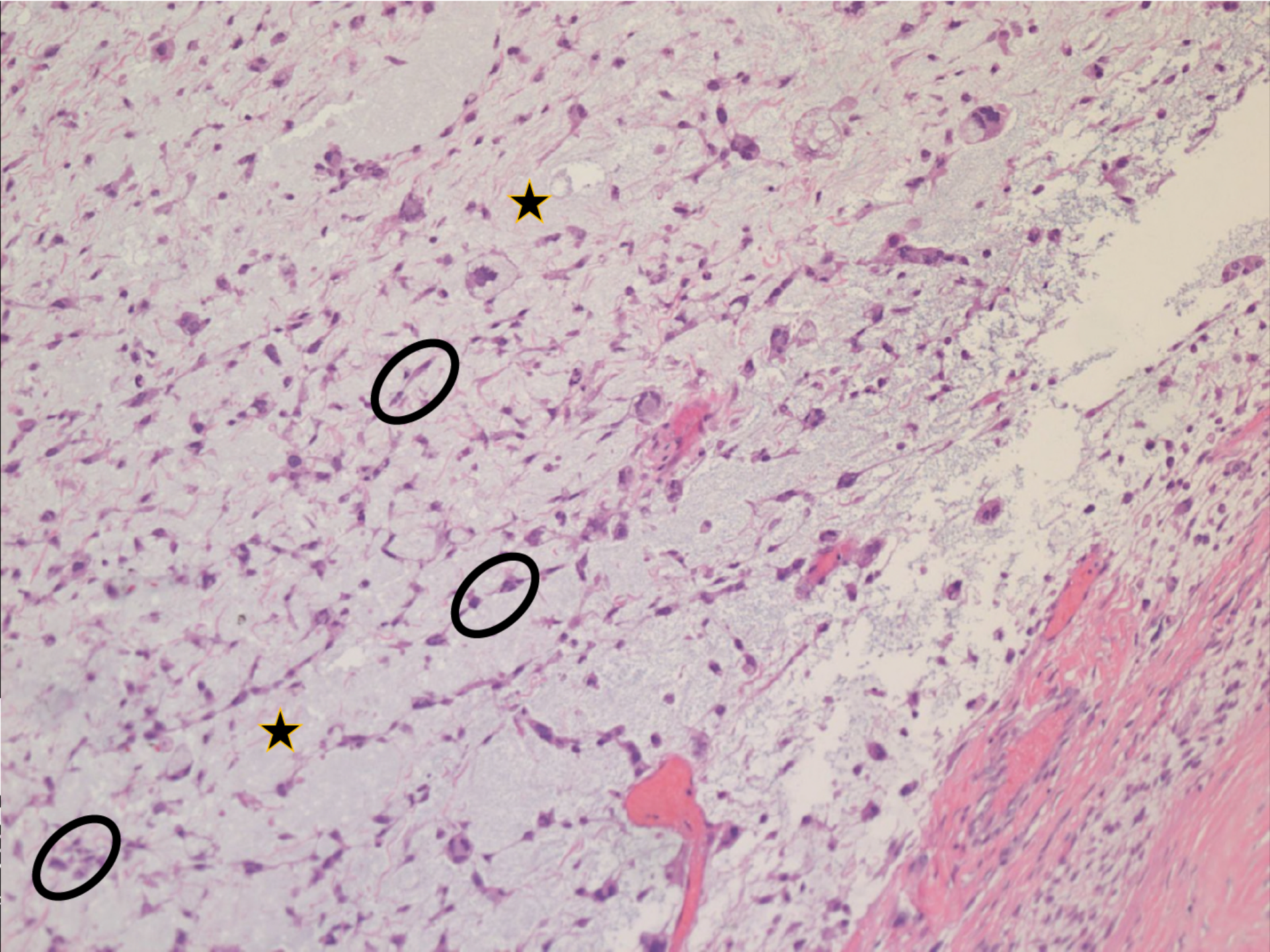


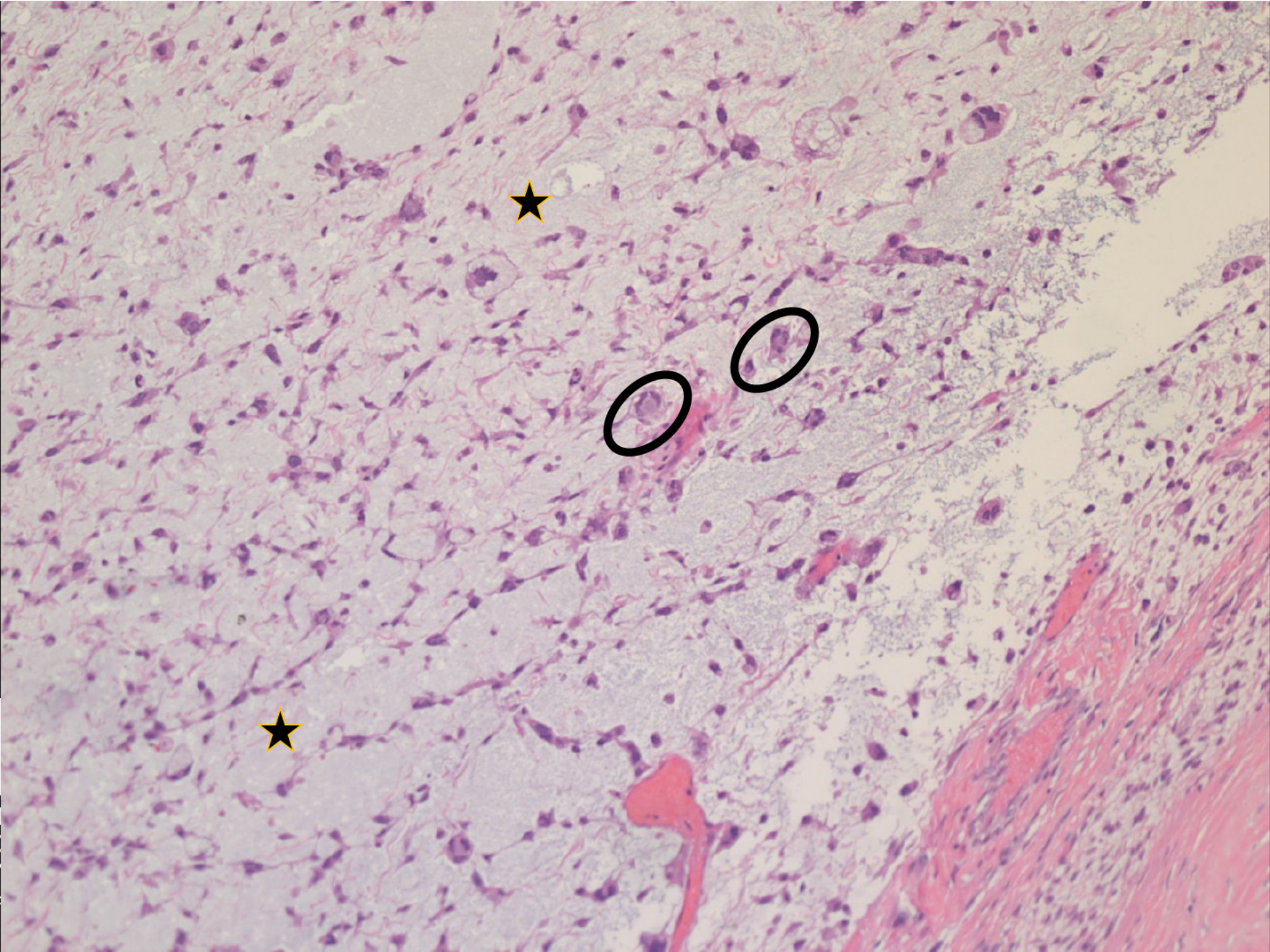


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Dr. Trilochan Hiremath
University of Pittsburgh Medical Center
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