AMSER Case of the Month September 2026

44 y/o male with recurrent skin lesions and fluctuant neck masses

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Patient Presentation

- HPI:
  - A 44-year-old male presents to the hospital with a skin lesion that formed on his right thigh
  - He has had a long history of recurrent lesions that mostly heal with proper wound care, but today presented with an ulcer that formed 1 week ago that has had increased wound drainage and poor healing
  - Has had significant pain over the wound area since it formed
  - Additionally has a new fluctuant lesion on his neck that has been drained in the past
  - Associated intermittent chills and fevers
  - He has chronic acne and episodes of hidradenitis suppurativa
  - The last flare 4 months ago required inpatient management

Pertinent Labs

- PMHx and Surgical History: HTN, Alcohol and Tobacco use Disorder, Major depressive disorder, chronic acne and wounds. Numerous mass I/D in the past
- Family History: No family history, has 4 children that are all healthy
- Physical Exam:
  - Large keloid mass on left anterior portion of neck with drainage
  - Bilateral hypopigmented scarring from previous lesions
  - Right thigh lesion covered by bandage. Pink granulation tissue present with no pus or purulence noted. Mild serosanguinous discharge from wound
- Vitals: BP: 172/82, Temperature: 97.8 °F
- Labs: WBC: 10.68, Hemoglobin: 9.7

What Imaging Should We Order?

Select the applicable ACR Appropriateness Criteria

Scenario	Scenario ID	Procedure	Adult RRL	Peds RRL	Appropriateness Category
Soft tissue infection suspected, neck, initial imaging	3195469	Radiography neck	0.1-1mSv ☢☢	0.03-0.3 mSv [ped] ☢☢	Usually appropriate
		US neck	0 mSv O	0 mSv [ped] O	Usually not appropriate
		MRI neck without and with IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
		MRI neck without IV contrast	0 mSv O	0 mSv [ped] O	Usually not appropriate
		3-phase bone scan neck	1-10 mSv ☢☢☢		Usually not appropriate
		CT neck with IV contrast	1-10 mSv ☢☢☢	0.3-3 mSv [ped] ☢☢☢	Usually not appropriate
		CT neck without and with IV contrast	1-10 mSv ☢☢☢	3-10 mSv [ped] ☢☢☢☢	Usually not appropriate
		CT neck without IV contrast	1-10 mSv ☢☢☢	0.3-3 mSv [ped] ☢☢☢	Usually not appropriate

Appropriate initial study was deferred by ED team

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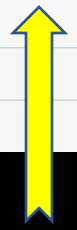
Select the applicable ACR Appropriateness Criteria

If radiography had been obtained and found to be unremarkable,

what should be ordered next?

Scenario	Scenario ID	Procedure	Adult RRL	Peds RRL	Appropriateness Category
Soft tissue infection suspected, neck, radiography normal, next imaging study	3195499	US neck	0 mSv O	0 mSv [ped] O	Usually appropriate
		Image-guided aspiration neck	Varies	Varies	Usually appropriate
		MRI neck without and with IV contrast	0 mSv O	0 mSv [ped] O	Usually appropriate
		MRI neck without IV contrast	0 mSv O	0 mSv [ped] O	Usually appropriate
		CT neck with IV contrast	1-10 mSv ☢☢☢	0.3-3 mSv [ped] ☢☢☢	Usually appropriate
		CT neck without IV contrast	1-10 mSv ☢☢☢	0.3-3 mSv [ped] ☢☢☢	May be appropriate
		3-phase bone scan neck	1-10 mSv ☢☢☢		Usually not appropriate
		CT neck without and with IV contrast	1-10 mSv ☢☢☢	3-10 mSv [ped] ☢☢☢☢	Usually not appropriate



Ordered by the ENT team

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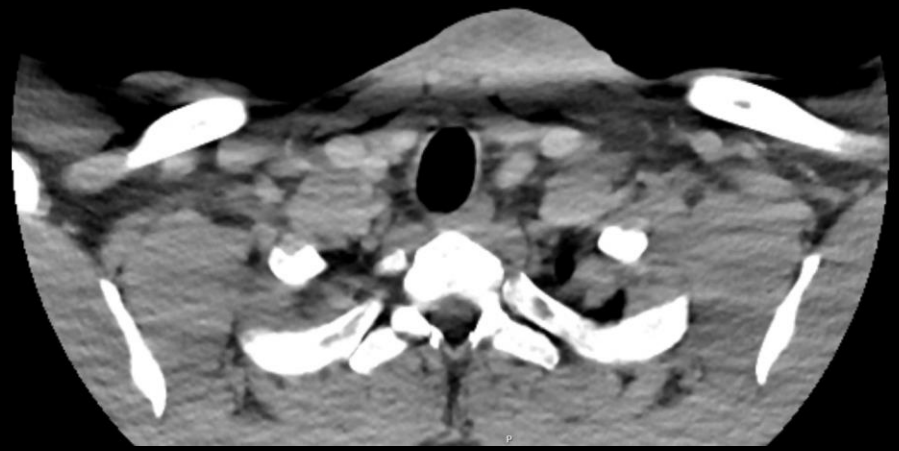
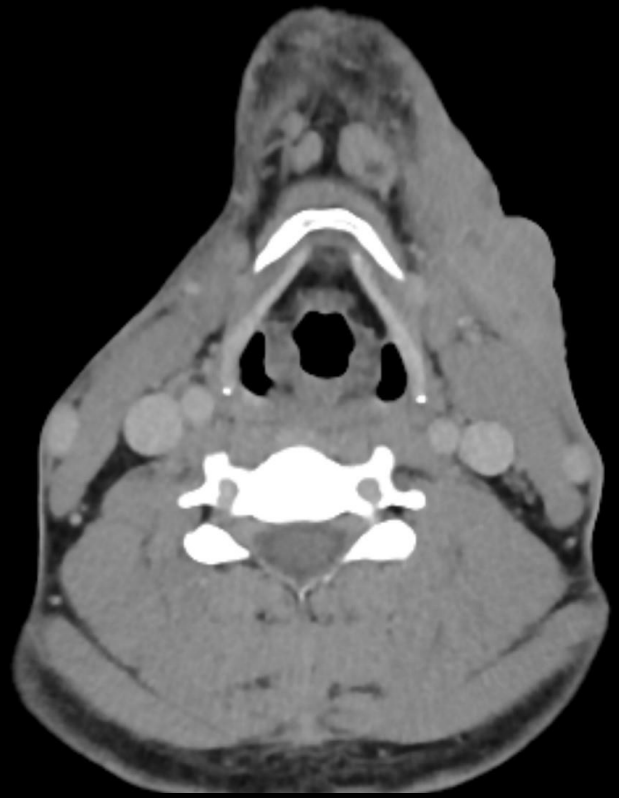
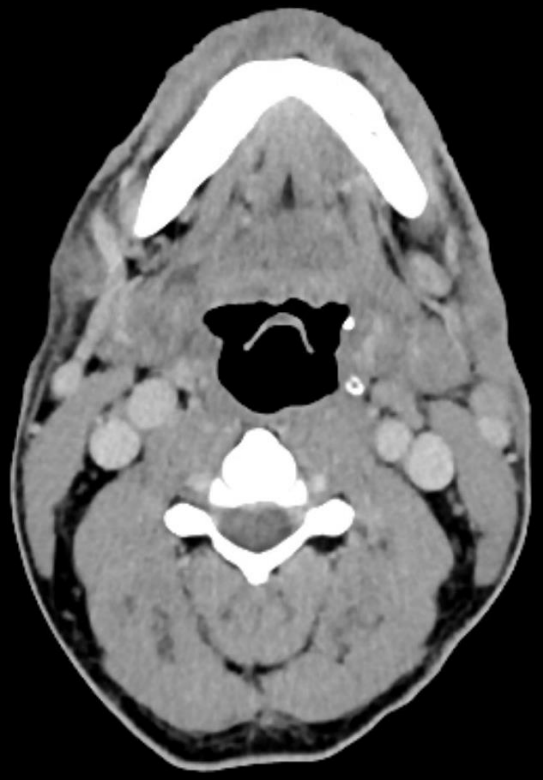
Findings (Unlabeled)



Axial contrast enhanced CT neck



Axial contrast enhanced CT neck



Axial contrast enhanced CT neck

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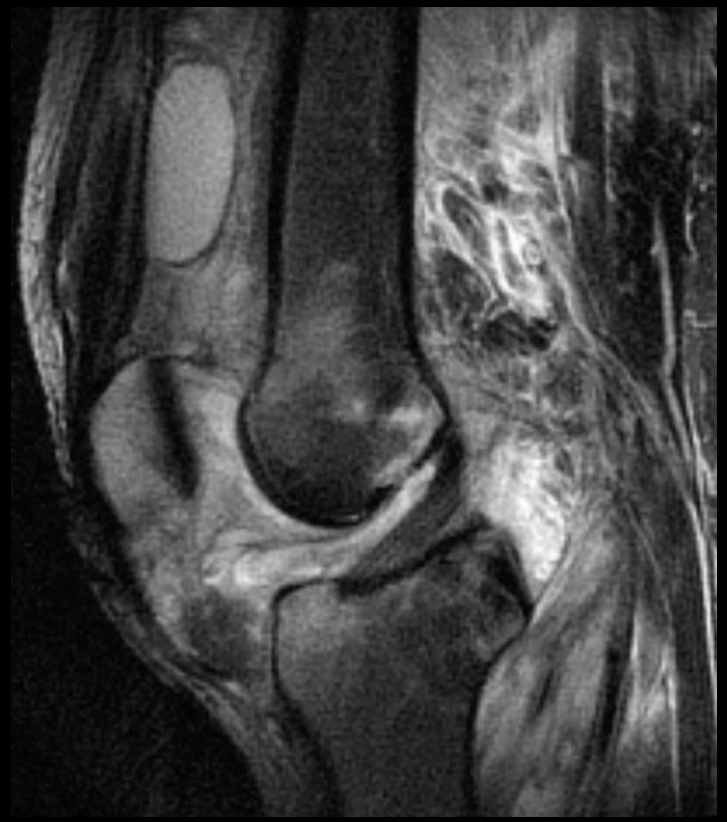
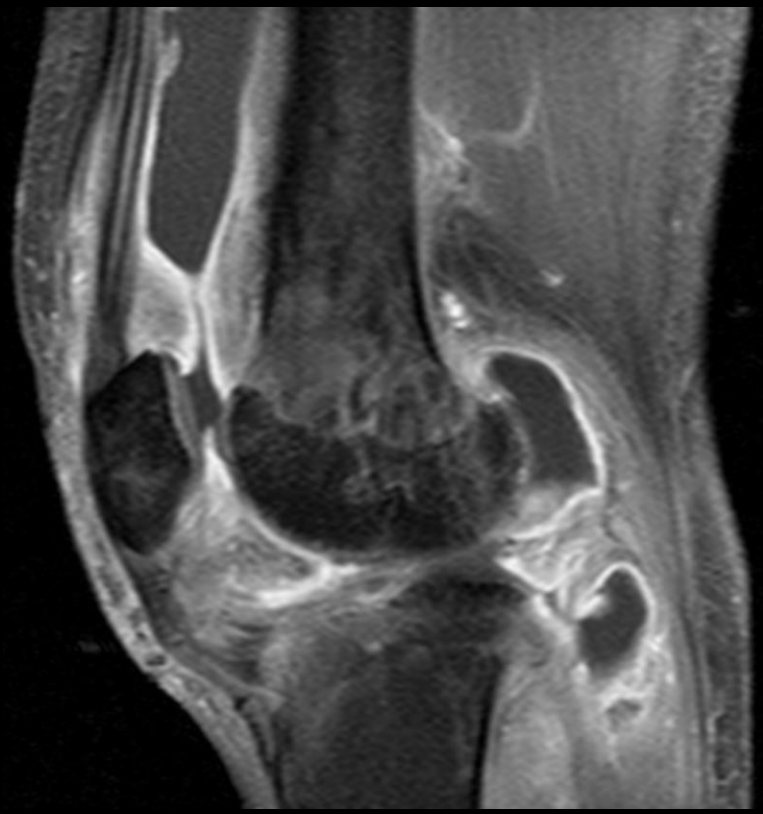
Relevant Prior Studies: Findings (Unlabeled)

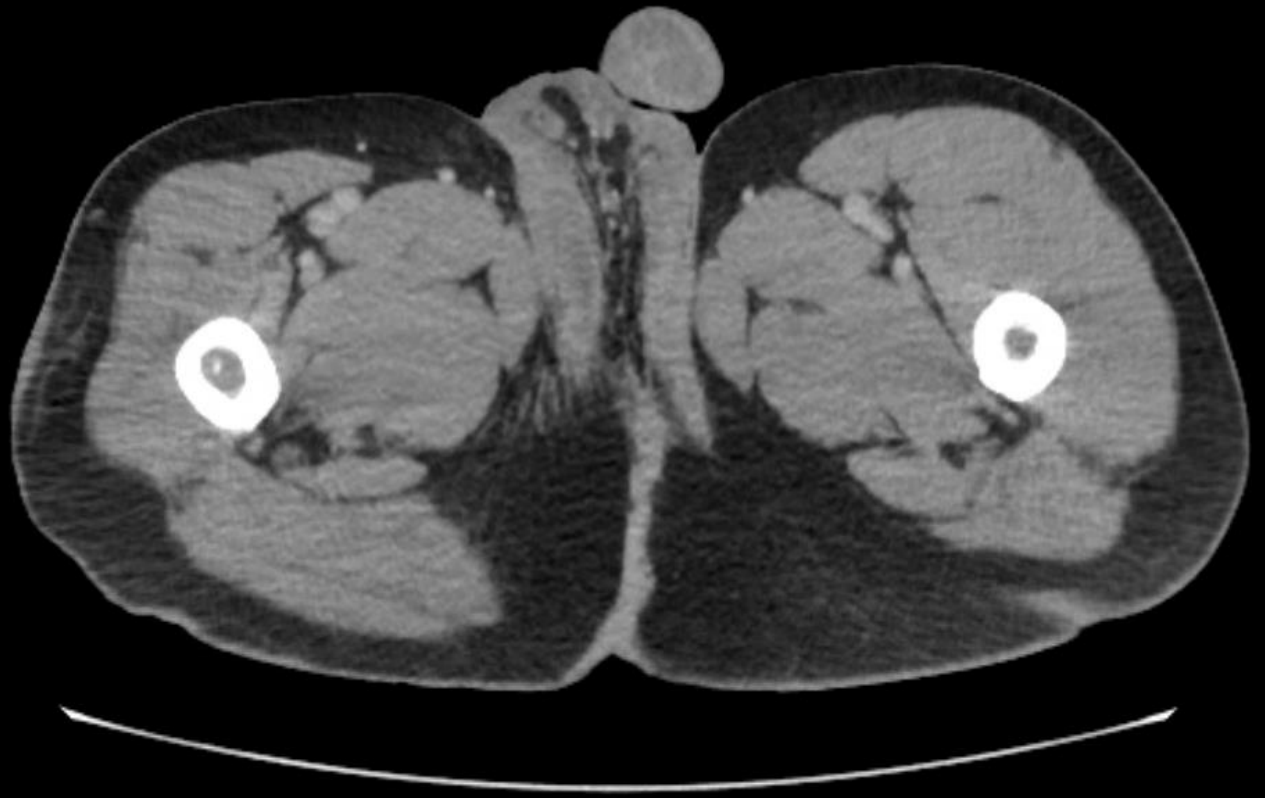


Sagittal MRI T2 fat-saturated right knee



Sagittal MRI T1 fat-saturated post contrast right knee

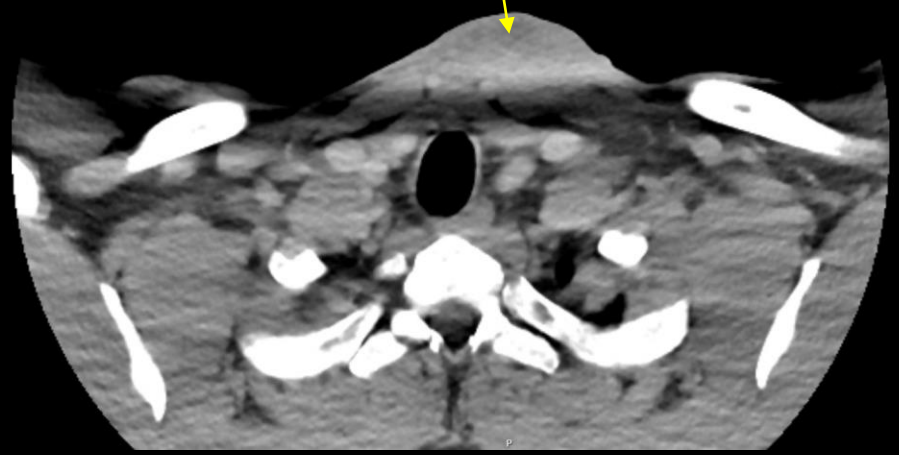
Relevant Prior Studies: Findings (Unlabeled)



Axial contrast enhanced CT pelvis and upper thighs

Findings (Labeled)

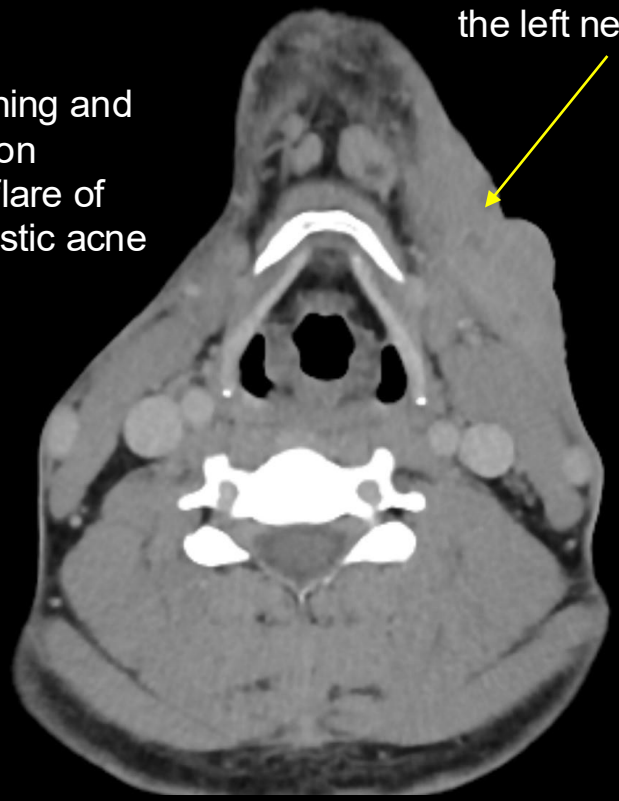
Ill-defined region with low attenuating center along the anterior neck with surrounding soft tissue thickening suggestive of phlegmon



Axial contrast enhanced CT neck

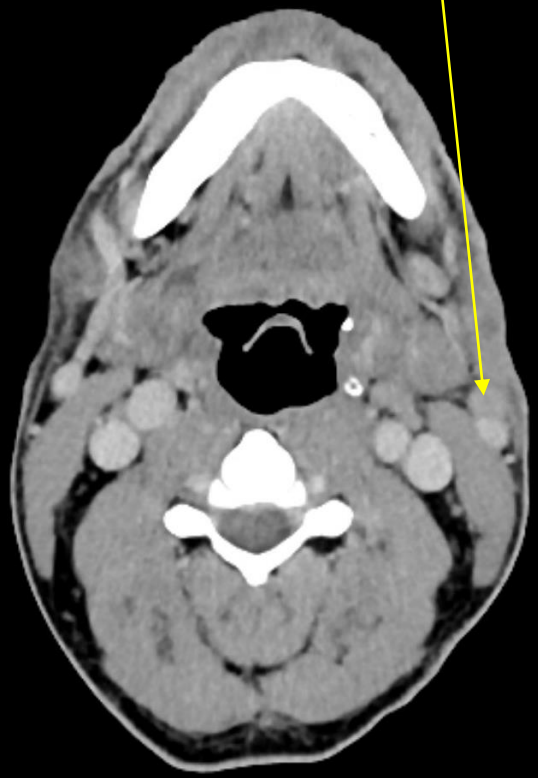
Both skin thickening and areas of phlegmon correlate with a flare of severe nodulocystic acne

Subcutaneous soft tissue thickening along the left neck



Axial contrast enhanced CT neck

11 mm reactive/persistent lymph node draining overlying soft tissue thickening and inflammation



Axial contrast enhanced CT neck

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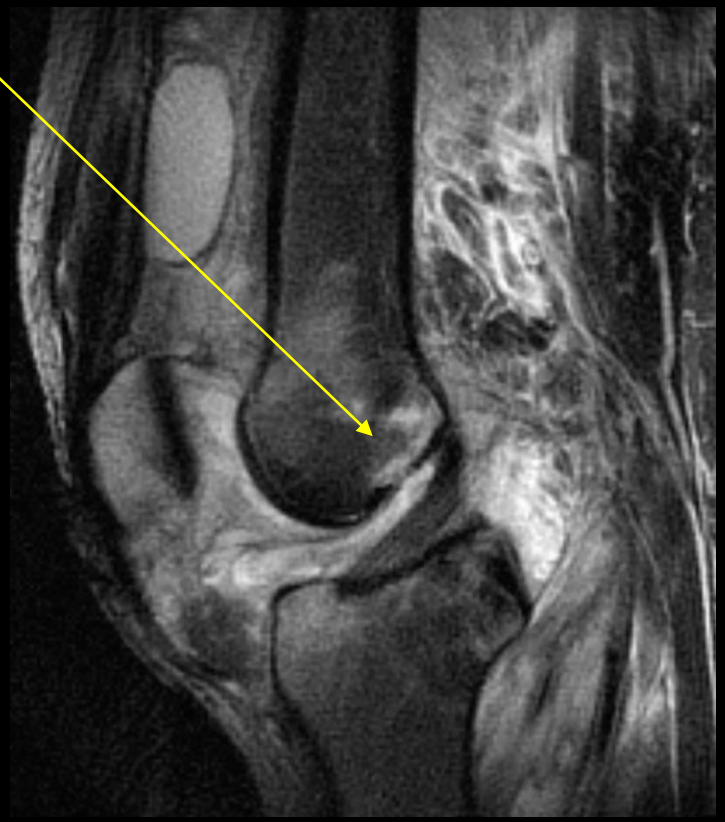
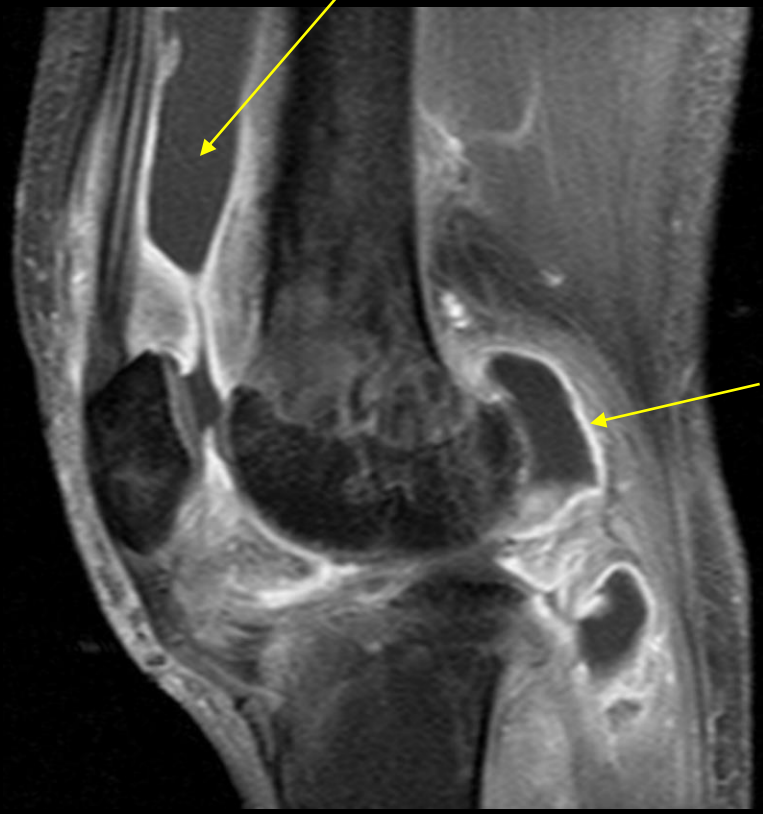
Relevant Prior Studies: Findings (Labeled)



Sagittal MRI T2 fat-saturated right knee

The combination of bone marrow edema, complex joint effusion, and synovial enhancement supports diagnosis of pyogenic arthritis. Frank pus was encountered at arthrotomy (however cultures remained sterile).



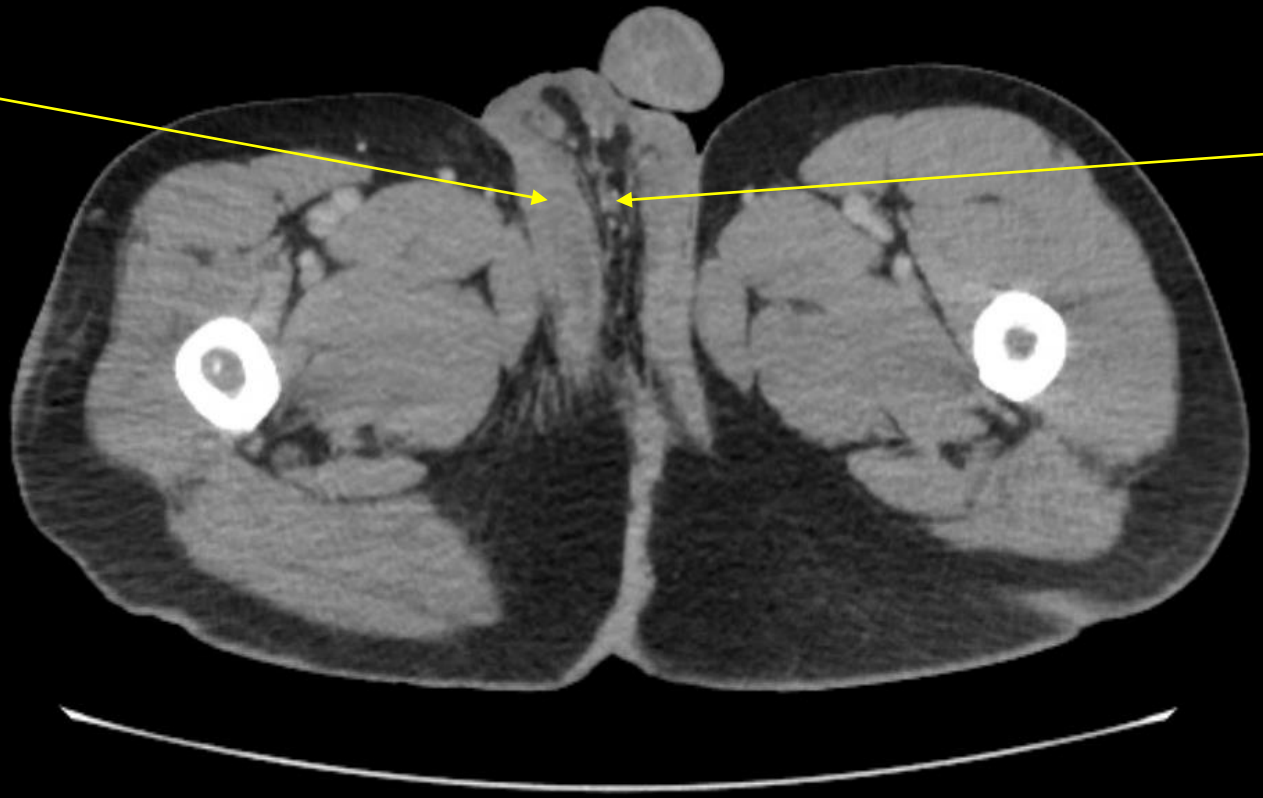
Sagittal MRI T1 fat-saturated post contrast right knee

Relevant Prior Studies: Findings (Labeled)

Coupled with clinical findings, skin thickening and subcutaneous fat stranding support diagnosis of chronic suppurative hidradenitis

Skin thickening along the medial thigh/perineal region

Prominent perineal subcutaneous fat stranding from chronic inflammation



Axial contrast enhanced CT pelvis and upper thighs

Final Dx:

PAPASH Syndrome

characterized by phlegmon, skin thickening, lymphadenopathy,
fat stranding, and complex joint effusion

Case Discussion

- Background:
  - PAPA (pyogenic arthritis, pyoderma gangrenosum, and acne), PASH (pyoderma gangrenosum, acne, and suppurative hidradenitis), and PAPASH (pyogenic arthritis, pyoderma gangrenosum, acne and hidradenitis suppurativa) syndrome are all autoimmune inflammatory diseases[1]
  - It was first described in 2013[2]
  - It is a predominantly neutrophilic disfunction disease that causes autoimmune attacks against the joints and skin of the body
- Epidemiology:
  - It is an exceedingly rare disease in literature with only 14 cases reported upon review as of 2025[1]
  - Various gene mutations are theorized to cause the disease including PSTPIP1, MEFV, NCSTN, NLRC4[1]
  - It is inherited in an autosomal dominant fashion[4]
  - PAPASH syndrome is a subtype of both PAP and PASH syndromes[3]
  - The average diagnostic age of onset of PAPASH syndrome is between 16 and 44 years old with acne occurring around 15 years old[3]

Case Discussion

- Disease Mechanism:
  - Mutated *PSTPIP1* gene regulates the actin cytoskeleton and interacts with pyrin which is encoded by the MEFV gene[3]
  - This leads to assembly of the pyrin inflammasome which leads to uncontrolled release of IL-1β[3]
  - Other inflammatory markers such as TNF-α, IL-17, IL-8, MMP-2, and MMP-9 are all significantly upregulated compared to healthy skin[3]
  - These inflammatory cytokines signal neutrophils to activate and attack the skin causing skin lesions and manifestations of pyoderma gangrenosum
  - In hidradenitis suppurativa, immune cell infiltration and hyperkeratosis and hyperplasia of the infundibular epithelium causes eventual follicular rupture[3]
    - The loss of the hair follicle causes a nodule/abscess to form leading to eventual bacterial infiltration[3]
    - The eventual chronic infection and healing leads to a larger sinus tract formation with persistent pus and drainage[3]
  - PAPASH syndrome is primarily a clinical and phenotypic diagnosis. While mutations such as *PSTPIP1* have been described in isolated reports, routine genetic testing lacks diagnostic specificity and is not required to establish the diagnosis[3]

Case Discussion

- Risk Factors[3]:
  - Up to 50% of individuals with hidradenitis suppurativa have metabolic syndrome (mainly obesity), IBD, spondyloarthritis, and smoking
  - Pyoderma gangrenosum is also associated with IBD, inflammatory arthritis, and solid organ malignant neoplasms
- Clinical Presentation:
  - Pyoderma gangrenosum is mostly found on the legs with other sites such as the head, face, ears, finger, and oral mucosa[3]
  - Hidradenitis suppurativa most commonly occurs in the axilla, inguinal , gluteal, perianal, and perineal regions of hair growth[3]
  - In our patient, chronic inflammation and ulcerations can cause areas where fluid can accumulate such in the lateral neck presenting as chronic nodulocystic acne
  - This area presented with phlegmon, thickened skin, and enlarged bilateral enlarged lymph nodes that required incision and drainage

Case Discussion

- Treatment:
  - There is currently not a targeted therapy to treat PAPASH syndrome
  - Current medications are focused on treating the individual pathologies that make up the syndrome and target individual cytokines
  - First line treatments included antibiotics, systemic glucocorticoids, and immunosuppressive therapy[1]
  - Disease Modifying Antirheumatic Drugs (DMARDs) are becoming more common with increasing clinical data[1]
  - Common DMARDs include TNF-alpha inhibitors such as adalimumab and infliximab[1]
  - While hidradenitis suppurativa does not have an FDA approved treatment, Secukinumab is an IL-17 inhibitor that is has be shown to improve the symptoms of moderate to severe hidradenitis suppurativa[1]
  - Current management targets symptom recurrence as there is no curative options to date
- Radiologic Findings:
  - For pyoderma gangrenosum, the inflammation is localized to the skin and is mainly a clinical diagnosis so radiological findings will be rare. However, severe cases can present with sterile osteomyelitis beneath the skin ulcers[5]
    - Our patient's pyoderma gangrenosum was dermatologically confirmed with no evidence of deep osseous extension on diagnostic scans
  - For hidradenitis suppurativa, ultrasound can be used to assess the lesions themselves along with assessing the skin thickness and hair follicle structure[6]
    - STIR sequencing on MRI can be used to diagnose the full extent of skin thickening, subcutaneous abscess, and sinus/fistulas that might have formed[6]
  - Soft tissue masses and accumulations part of pyogenic arthritis, such as those seen in our patient, can be explored with CT scan and MRI for depth and size before eventual incision and drainage

References:

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3. Satoh TK. Genetic mutations in pyoderma gangrenosum, hidradenitis suppurativa, and associated autoinflammatory syndromes: Insights into pathogenic mechanisms and shared pathways. *J Dermatol*. 2024;51(2):160-171. doi:10.1111/1346-8138.17028
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5. Smith H, Sharma R, Tao W, et al. Pyoderma gangrenosum. Reference article, Radiopaedia.org (Accessed on 11 May 2026) https://doi.org/10.53347/rID-67054
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