AMSER Radiology-Pathology Case of the Month:

Incidentally Found Right Thigh Mass

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Patient Presentation

- HPI: Patient is a 72-year-old male presenting as a referral from his PCP for evaluation of a right thigh soft tissue mass. One month prior to presentation, the patient had a CT scan for evaluation of nausea, vomiting, diarrhea, and abdominal pain. He was incidentally found to have a heterogeneous mass in his anterior medial right thigh. He reports having a dull ache in his right groin that has been present for a long time, however, denies any pain or limits in his daily life. He has not noticed the mass himself. Of note, he had a CT scan for evaluation of cholecystitis 2 years prior which showed evidence of this mass, but the patient states he was never informed. MRI was ordered by PCP and presented for evaluation.
- PMH: Coronary artery disease, cardiomyopathy, atrial fibrillation on warfarin, aortic stenosis, hypertension, obstructive sleep apnea, diabetes, AAA, hyperlipidemia, and peripheral neuropathy.
- PSH: Laparoscopic cholecystectomy, cardiac catheterization, biceps tendon repair, colonoscopy with polypectomy.
- Physical Exam: No palpable mass. Minimal tenderness to palpation. 5 out of 5 muscle strength in the lower extremities. Decrease sensation to light touch bilateral lower extremities which patient contributes to neuropathy.



Pertinent Labs

• No lab tests were ordered upon initial presentation



What Imaging Should We Order?



Select the applicable ACR Appropriateness Criteria

American College of Radiology ACR Appropriateness Criteria[®] Soft-Tissue Masses

Variant 1: Soft-tissue mass. Superficial or palpable. Initial imaging study.			
Procedure	Appropriateness Category	Relative Radiation Level	
X-ray area of interest	Usually Appropriate	Varies	
US area of interest	Usually Appropriate	0	
MRI area of interest without IV contrast	May Be Appropriate (Disagreement)	0	
CT area of interest with IV contrast	Usually Not Appropriate	Varies	
CT area of interest without and with IV contrast	Usually Not Appropriate	Varies	
CT area of interest without IV contrast	Usually Not Appropriate	Varies	
FDG-PET/CT area of interest	Usually Not Appropriate	ବବବବ	
MRI area of interest without and with IV contrast	Usually Not Appropriate	0	

<u>Variant 2:</u> Soft-tissue mass. Nonsuperficial (deep) or nonspecific clinical assessment or located in an area difficult to adequately evaluate with radiographs (flank, paraspinal region, groin, or deep soft tissues of the hands and feet). Initial imaging study.

Procedure	Appropriateness Category	Relative Radiation Level	
X-ray area of interest	Usually Appropriate	Varies	
CT area of interest without and with IV contrast	May Be Appropriate (Disagreement)	Varies	
CT area of interest without IV contrast	May Be Appropriate (Disagreement)	Varies	
MRI area of interest without and with IV contrast	May Be Appropriate (Disagreement)	0	
MRI area of interest without IV contrast	May Be Appropriate (Disagreement)	0	ſ
US area of interest	May Be Appropriate	0	
CT area of interest with IV contrast	Usually Not Appropriate	Varies	
FDG-PET/CT area of interest	Usually Not Appropriate	****	

Variant 3:

Soft-tissue mass. Nondiagnostic initial evaluation (ultrasound and/or radiograph). Next imaging study.

Procedure	Appropriateness Category	Relative Radiation Level
MRI area of interest without and with IV contrast	Usually Appropriate	0
MRI area of interest without IV contrast	Usually Appropriate	0
CT area of interest with IV contrast	May Be Appropriate (Disagreement)	Varies
CT area of interest without IV contrast	May Be Appropriate	Varies
CT area of interest without and with IV contrast	Usually Not Appropriate	Varies
FDG-PET/CT area of interest	Usually Not Appropriate	****

X-ray usually appropriate 1st step, however, patient presented with completed CT with and without contrast and MRI with and without contrast.



Findings (unlabeled)





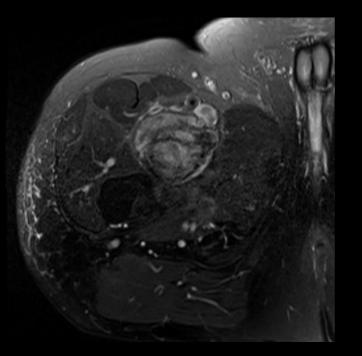


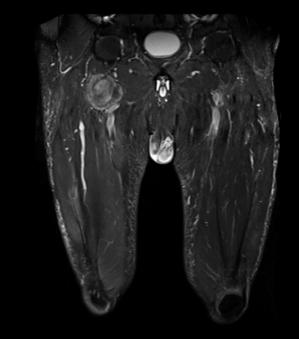
CT Scan-2 Years Prior to Presentation

CT Scan-1 Month Prior to Presentation



Findings (unlabeled)



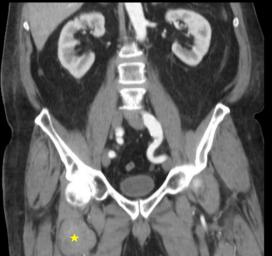


MRI with Axial T2 image (left) and Coronal STIR Image (right)





Findings (Labeled)



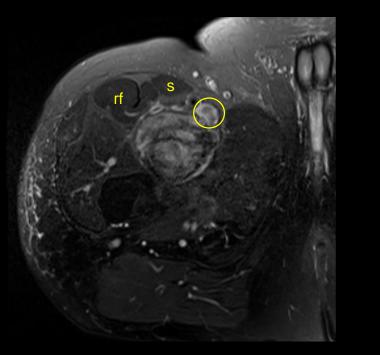
CT Scan-2 Years Prior to Presentation

CT Scan-1 Month Prior to Presentation 5.5 x 5.6 x 4.9 cm heterogeneous mass in anteromedial thigh (denoted by yellow star).

CT scan performed 2 years later demonstrates the same mass which has grown to 7.3 x 4.7 x 6.0 cm.



Findings (Labeled)





MRI with Axial T2 image (left) and Coronal STIR Image (right)

- 7.3 x 4.7 x 6.0 cm mass with extensive contrast enhancement located directly posterior to femoral vessels (yellow circle), sartorius (denoted by s), and rectus femoris (denoted by rf) muscles.
- No evidence of direct invasion of adjacent structures
- Most likely evidence of malignant lesion favoring soft tissue sarcoma
- Biopsy/Excision recommended to establish diagnosis



Differential Diagnosis

- Undifferentiated pleomorphic sarcoma (malignant fibrous histiocytoma)
- Pleomorphic liposarcoma
- Dedifferentiated liposarcoma

- Dermatofibrosarcoma protuberans
- Fibrous Histiocytoma
- Neurofibroma
- Perineurioma



Additional Imaging & Tests

- Ultrasound guided soft tissue biopsy was subsequently performed.
- Final Pathology demonstrated spindle cell neoplasm consistent with soft tissue perineurioma.



Final Cytopathology Diagnosis: MASS, RIGHT THIGH, CORE BIOPSY WITH TOUCH PREPARATION CYTOLOGY: A. SPINDLE CELL NEOPLASM CONSISTENT WITH SOFT TISSUE PERINEURIOMA

Tumor immunohistochemical profile:

- Positive: EMA, CD34
- Negative: CAM5.2, S-100, desmin, MSA, SOX-10, STAT-6, MUC4, Pan-melanotic marker



Next Steps

- Given the benign nature of this neoplasm, the patient was informed of his option to have the mass removed.
- He was educated that excision is not required but is certainly an option for definitive diagnosis, symptom management, and to prevent further increase in size.
- He decided to ultimately have the mass surgically excised.



Surgical Excision & Gross Pathology

- A ~10cm incision was made Over the medial thigh just lateral to the palpable femoral vessels.
- An interval was developed between the sartorius muscle and the rectus femoris muscle while keeping the femoral vessels retracted medially within their sheath.
- This permitted blunt dissection of the mass which was circumferentially released and removed from the wound bed.







Micropathology

- The mass was taken frozen section which was consistent with the biopsy results of a soft tissue perineurioma.
- Final pathology was also consistent with this diagnosis.
- Histologic Features
 - Bland, elongated cells in parallel bundles with storiform growth pattern
 - No atypia, minimal or no mitotic figures

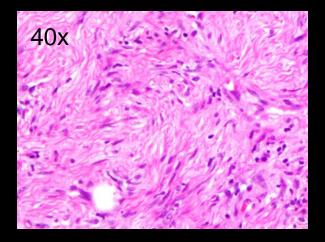
Final Cytopathology Diagnosis:

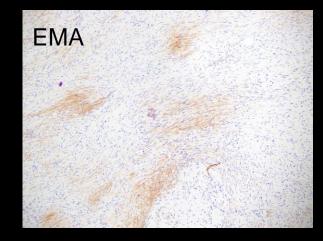
MASS, RIGHT THIGH, CORE BIOPSY WITH TOUCH PREPARATION CYTOLOGY: A. SPINDLE CELL NEOPLASM CONSISTENT WITH SOFT TISSUE PERINEURIOMA

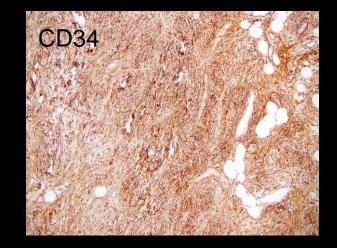
Tumor immunohistochemical profile:

Positive: EMA, CD34

 Negative: CAM5.2, S-100, desmin, MSA, SOX-10, STAT-6, MUC4, Pan-melanotic marker 10x









Final Dx:

Extraneural/Soft Tissue Perineurioma of the Right Thigh



Case Discussion

- Perineurioma
 - Definition: Tumor composed of neoplastic perineurial cells
- Several Forms
 - <u>Intraneural</u>-proliferates throughout endoneurium
 - Extraneural/Soft-Tissue
 - <u>Sclerosing</u>-variant of extraneural with perivascular arrangement
 - <u>Reticular</u>-variant of extraneural which have degenerative myxoid changes
 - <u>Malignant (Perineurial MPNST)</u>-arise exclusively from extraneural perineuriomas



Case Discussion-Extraneural Perineurioma

- Epidemiology
 - Primarily affects adults and involve superficial soft tissues of extremities
 - 30% develop in deep soft tissue (as in this case)
- Clinical Pathology
 - Typically not grossly associated with a nerve
 - Symptoms arise due to non-specific mass effect or are incidentally found
 - Well-circumscribed but no capsule
- Radiographic Features
 - CT: lesions tend to be hypodense compared to skeletal muscle
 - MRI Findings: nonspecific, variable intensity in both T1/T2 sequences relative to skeletal muscle



Case Discussion-Extraneural Perineurioma

- Histologic Features
 - Most common is a spindle-cell lesion with long/slender fibroblast-like cells
 - Varying growth patterns with storiform being most common
 - Other patterns included whorled, Pacinian, or short fascicular
- Immunohistochemistry Features
 - Definition: Benign perineuriomas are EMA-positive and S-100 negative
 - Normal immunophenotype of perineurial cells
 - A majority express claudin-1 and GLUT1
 - 20%-60% express CD34 and actin
 - MUC4 is normally tested in these cases as perineuriomas may closely resemble low-grade fibromyxoid sarcomas
 - MUC4 has very high sensitivity for low-grade fibromyxoid sarcomas and can help with differentiation



Case Discussion-Extraneural Perineurioma

• Clinical Management

- These masses have a very minimal risk of metastasis and a low recurrence rate after removal
- If patient desires symptomatic relief, the tumor can be excised surgically.
- However, the benign nature of the neoplasm does not necessitate surgical management unless otherwise indicated.



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