

PEDIATRICS CASE

Lauren Hoff, MS¹, David Bloom, MD²

¹ University of Michigan Medical School

² University of Michigan Department of Radiology

Case Presentation

History:

- 18-year-old male with history of asthma presented with 2-weeks of intermittent fevers, erythematous pruritic rash with subcutaneous nodules, night sweats, fatigue, and progressive ascending polyarthralgia.

Significant Physical Exam Findings:

- Sinus tachycardia
- Diffuse joint tenderness with bilateral effusions of the knees and ankles
- A few erythematous subcutaneous nodules of the anterior lower extremities

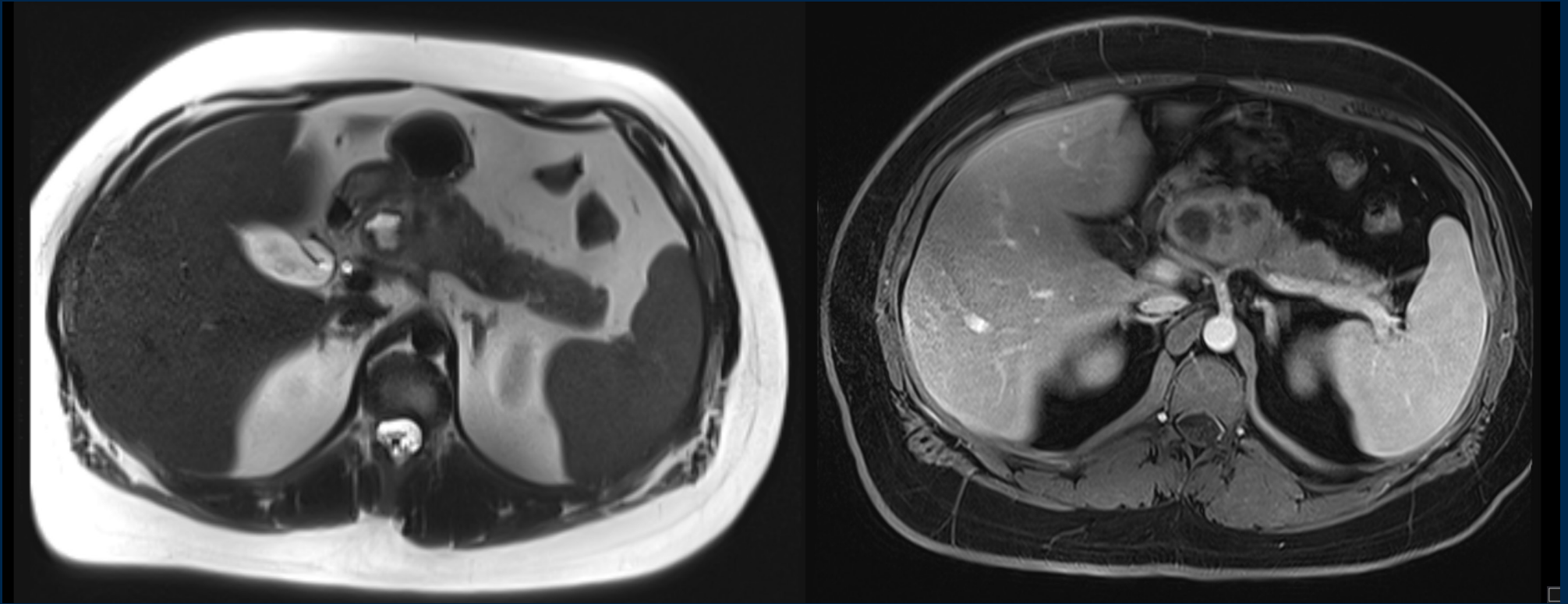
Significant Labs:

- Leukocytosis
- Normocytic anemia
- ANA positive, CRP elevated
- D-dimer > 5000
- Lipase >9000

Imaging:

- CT Pulmonary Angiogram – Diffuse ground-glass opacities, no evidence of pulmonary embolism.
- CT Abdomen/Pelvis prompted an MRI Abdomen – Let's take a look 😊

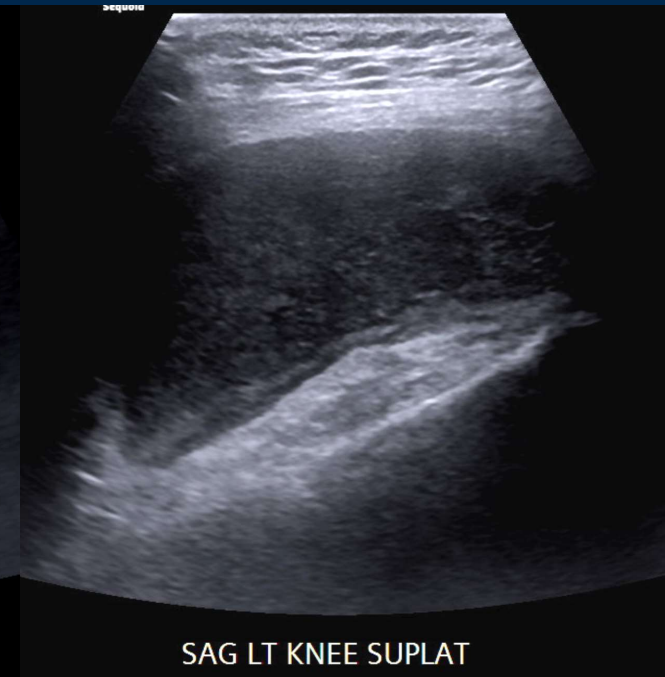
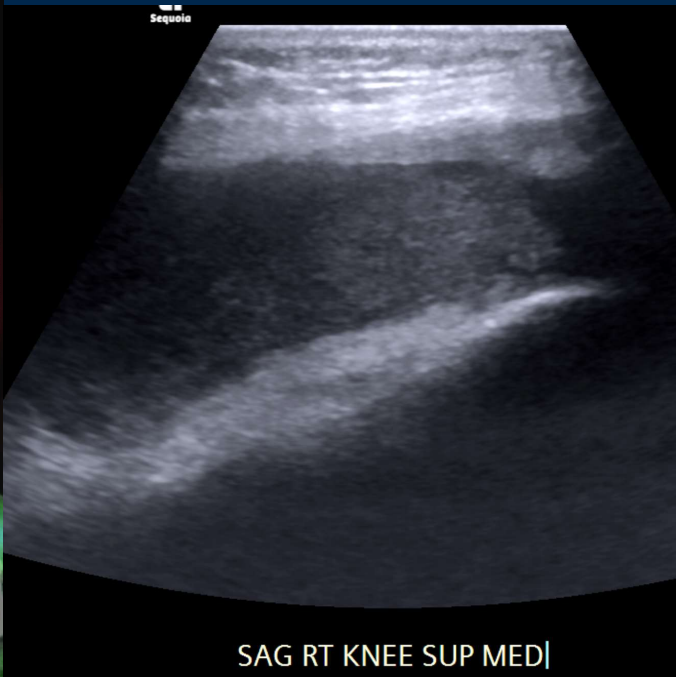
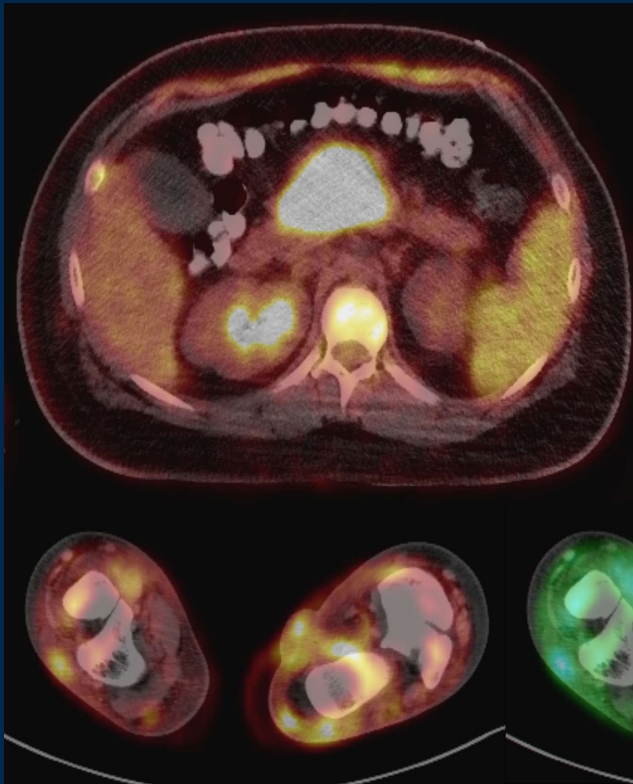
MRI



MRI Findings

- Heterogeneous pancreatic mass within the head and proximal body of the pancreas measuring 6.2 x 6.5 x 4.5 cm. The mass demonstrates cystic change, necrosis, and evidence of prior hemorrhage.
- These findings likely represent a pancreatic neoplasm.
- There are subcutaneous nodules present which are unlikely to represent metastatic disease.
- No liver metastases.

Follow-Up PET/CT and Ultrasound



PET/CT and Ultrasound Findings

- PET/CT
 - Intense hypermetabolism in the pancreatic mass may reflect active malignancy or infection/inflammation.
 - Multifocal subcutaneous soft tissue nodules with hypermetabolism may reflect multifocal infectious or inflammatory pathology.
- Ultrasound
 - Large bilateral complex knee joint effusions/synovial thickening, suggesting polyarthritis.

Differential Diagnosis of Pancreatic Mass

1. Solid Pseudopapillary Tumor of the Pancreas
2. Pancreatic Lymphoma
3. Pancreatic Sarcoma

Differential Diagnosis

Solid Pseudopapillary Tumor of Pancreas

- Rare and usually benign neoplasm that is most often seen in young females. Usually asymptomatic at presentation.
- Radiographic features:
 - CT – well-defined lesion with varying cystic and solid components.
 - MRI - well-defined lesion, low signal intensity on T1, high signal intensity on T2.
- Preferentially located at the pancreatic tail.

Differential Diagnosis

Pancreatic Lymphoma (Focal Form)

- Most often arises in middle-aged patients and is more common in the immunocompromised.
- Symptoms may include abdominal pain, palpable abdominal mass, weight loss, obstructive jaundice, or acute pancreatitis.
- Radiographic features:
 - CT – minimal enhancement, peripancreatic lymph node enlargement, uniform low attenuation.
 - MRI – low signal intensity on T1 and intermediate signal intensity on T2, faint contrast enhancement.
- Preferentially located at the pancreatic head.

Differential Diagnosis

Pancreatic Sarcoma

- Primary leiomyosarcomas, fibrosarcomas, undifferentiated pleomorphic sarcomas, follicular dendritic cell sarcomas, and carcinosarcomas have been described.

We'll talk about one of these subtypes in detail...

Diagnosis

- Skin punch biopsy – panniculitis.
- Pancreatic FNA – **high-grade undifferentiated pleomorphic sarcoma.**
- There was also clinical and laboratory evidence of acute pancreatitis.

Discussion

- To date, there are 16 previously-described cases of **undifferentiated pleomorphic sarcoma of the pancreas**.
- Symptoms at presentation tend to include epigastric pain, nausea, and vomiting.
- Most often arises in middle-aged patients. Previously, the youngest patient was 37 years old.
- The tumor may arise in any region of the pancreas.
- Radiographic features:
 - CT – large, heterogeneous, low-attenuation or multinodular mass with cystic components and necrosis.
 - MRI – low signal intensity on T1 and high signal intensity on T2, heterogeneous enhancement
- Diagnosis is typically made by immunohistochemistry.

Discussion

- Rarely, pancreatic disorders such as acute pancreatitis, chronic pancreatitis, and pancreatic carcinoma have been accompanied or preceded by panniculitis and polyarthrititis. This triad is known as **Pancreatitis, Panniculitis, and Polyarthrititis (PPP) Syndrome**.
- To our knowledge, this is the first case of pancreatic sarcoma associated with PPP Syndrome.

Patient Outcome

- Patient has completed 3 cycles of chemotherapy with dexrazoxane, doxorubicin, ifosfamide, and mesna.
- Complications including worsening pancreatitis have occurred while completing chemotherapy.
- Tumor Board has recommended a pancreaticoduodenectomy.

References

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