

# Pleomorphic Sarcoma Presenting as Pancreatitis, Panniculitis and Polyarthrititis Syndrome



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## Introduction

**Pancreatitis, panniculitis and polyarthrititis (PPP) syndrome is a rare triad seen with pancreatic pathology**

- Pathogenesis is unknown but thought to be due to circulating pancreatic enzymes causing lipolysis
- Occurs in acute or chronic pancreatitis and pancreatic cancer<sup>1</sup>
- Roughly 50% of patients with PPP syndrome deny abdominal pain<sup>1</sup>

## Case Description

**An 18-year-old male presented for intermittent fevers and progressive joint pain.**

- Denied respiratory symptoms, abdominal pain and other infectious symptoms
- Physical exam demonstrated
  - Erythematous, tender nodules on shins (Image 1A)
  - Bilateral ankle, foot and hand swelling
- Initial labs were notable for elevated lipase 9,754 with mildly elevated AST and ALT

- CT demonstrated a 5.9 x 5 cm solid pancreatic mass with intracystic component
- Rheumatologic and infectious work up was unremarkable
- Skin biopsy demonstrated pancreatic panniculitis
- Aspirate during endoscopic ultrasound (EUS) showed undifferentiated pleomorphic sarcoma
- Initiated chemotherapy followed by surgical resection

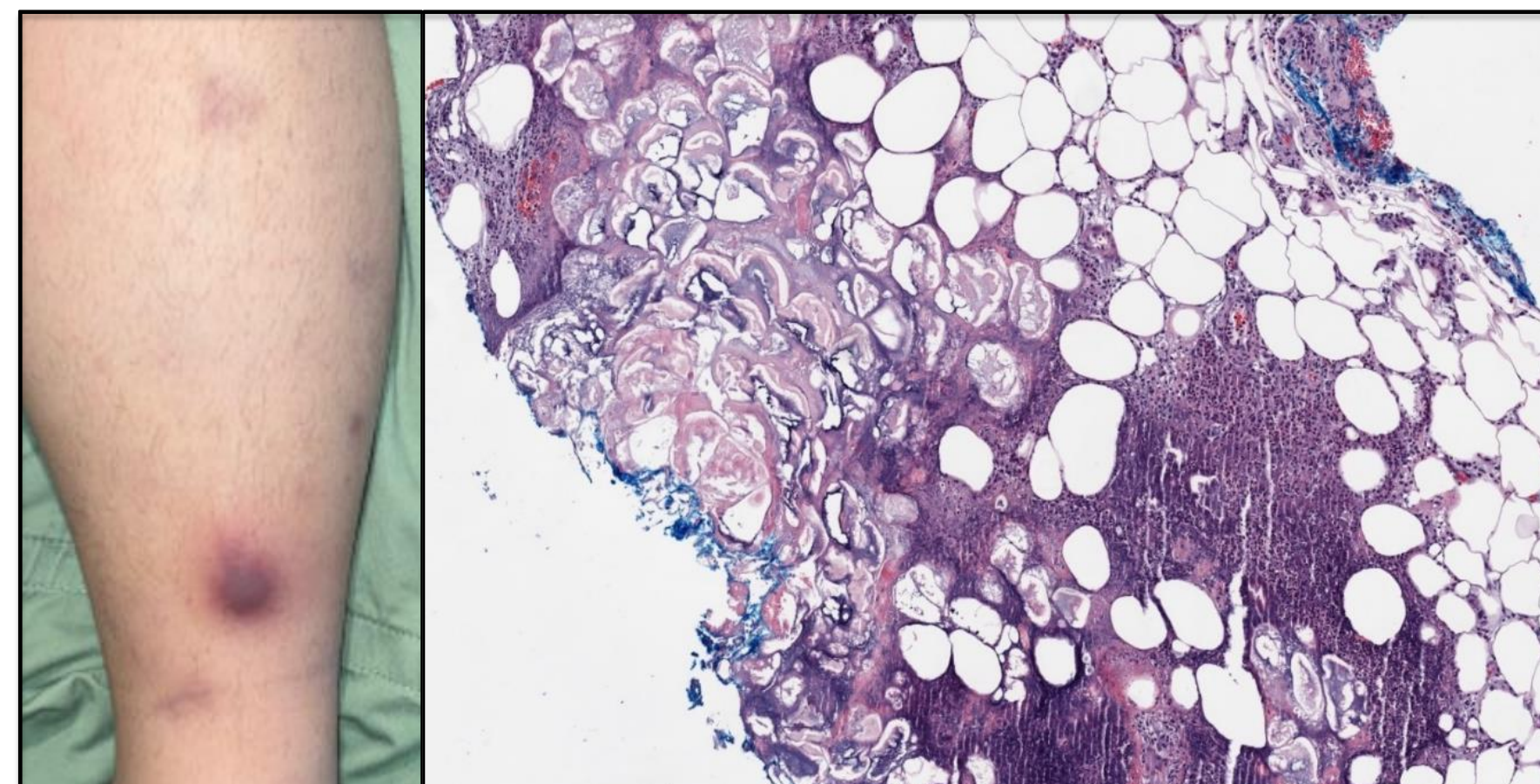


Image 1. A) Erythematous nodules on anterior left shin  
B) Skin biopsy demonstrating subcutaneous fat necrosis with necrotic adipocyte ghosts and basophilic material

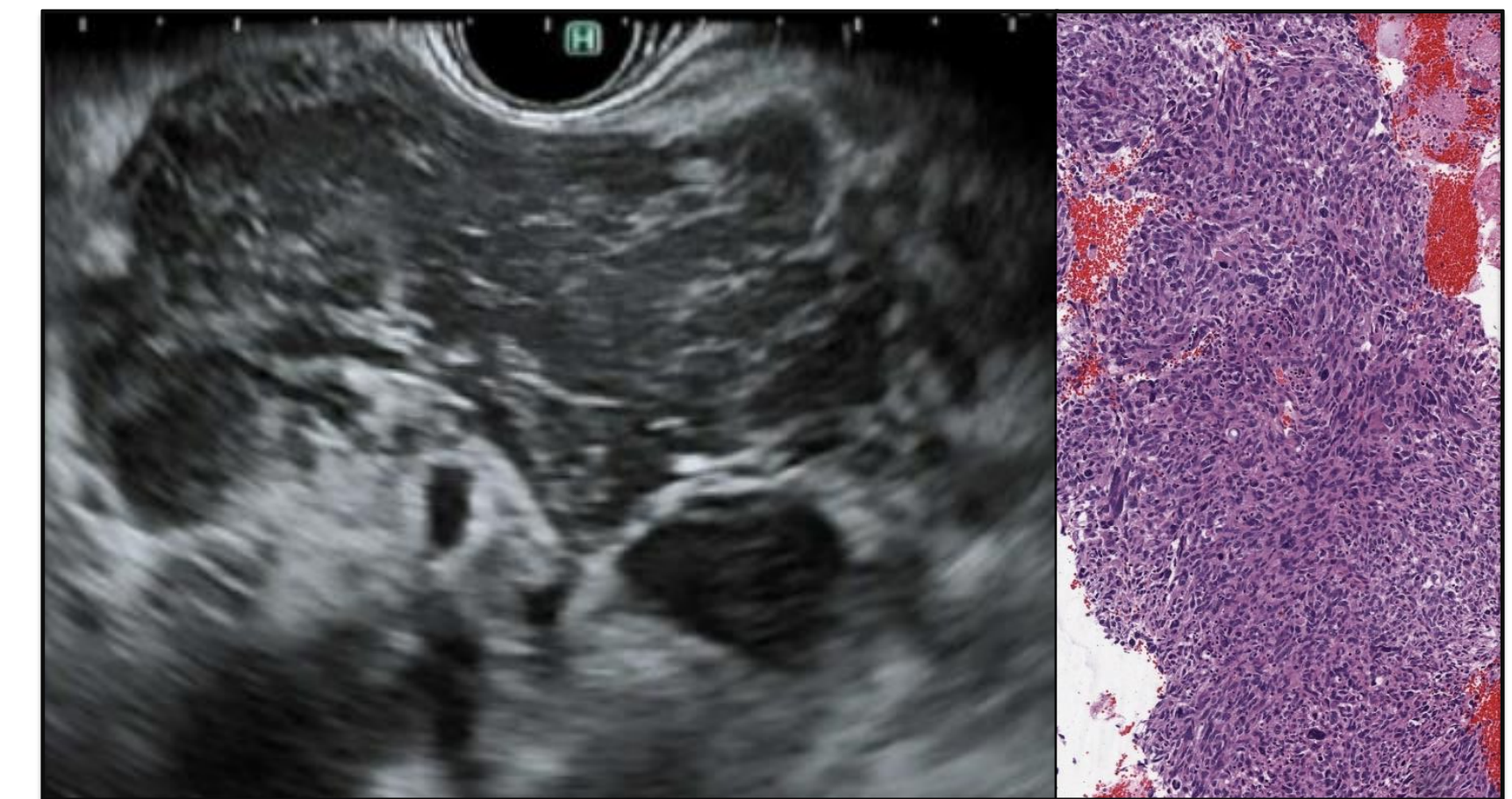


Image 2. A) Pancreatic mass on EUS B) Histology with frequent bizarre mitotic features and anaplastic cytology consistent with undifferentiated pleomorphic sarcoma

## Discussion

PPP syndrome is an important manifestation of pancreatic cancer

- Resolution relies on treatment of disease process
- High mortality estimated at over 25% of patients<sup>1</sup>

## Conclusion

Prompt recognition of the clinical features of PPP syndrome can aid in identifying pancreatic pathology

## References

1. Betrains A, Rosseels W, Van Mieghem E, Vanderschueren S, Nijs J. Clinical characteristics, treatment, and outcome of pancreatitis, panniculitis, and polyarthrititis syndrome: a case-based review. *Clin Rheumatol.* 2021;40(4):1625-1633. doi:10.1007/s10067-020-05333-8