

Atypical Onset of Systemic Lupus Erythematosus with Acute Pancreatitis and Class II Lupus Nephritis: A Rare Case Report

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Abstract

Systemic Lupus Erythematosus (SLE) is a multisystem autoimmune disease with diverse clinical manifestations; gastrointestinal involvement is uncommon, and acute pancreatitis as the initial presentation of SLE is exceedingly rare, with its coexistence alongside lupus nephritis at disease onset posing a diagnostic challenge that may delay appropriate treatment. We report a 37-year-old female who presented with a two-week history of progressively worsening right upper quadrant abdominal pain, vomiting, low-grade fever, generalized edema, and abdominal distension. Laboratory investigations revealed leukopenia, anemia, elevated serum amylase and lipase levels, severe hypoalbuminemia, nephrotic-range proteinuria, and hypocomplementemia. Magnetic resonance cholangiopancreatography demonstrated edematous pancreatitis with a peripancreatic fluid collection, gross ascites, and bilateral pleural effusion. Common etiologies of acute pancreatitis, including gallstones, alcohol use, hypertriglyceridemia, viral hepatitis, and drug-induced causes, were excluded. Further autoimmune evaluation showed strongly positive antinuclear antibodies with markedly reduced complement levels, and renal biopsy confirmed class II lupus nephritis. Based on the clinical, serological, and histopathological findings, a diagnosis of SLE presenting as acute pancreatitis with concurrent lupus nephritis was established. The patient responded favorably to intravenous methylprednisolone, followed by oral corticosteroids, mycophenolate mofetil, and an angiotensin receptor blocker. This case underscores that acute pancreatitis may rarely represent the first manifestation of SLE and should be considered in patients with unexplained pancreatitis accompanied by multisystem features such as cytopenias, serositis, hypocomplementemia, or significant proteinuria, and highlights the value of early autoimmune evaluation and prompt immunosuppressive therapy in preventing disease progression and improving clinical outcomes

Keywords: Autoimmune Diseases; Hypocomplementemia; Nephrotic Syndrome; Proteinuria; Serositis.

Introduction

Case Presentation

A 37-year-old female presented with two weeks of progressively worsening dull, continuous epigastric and right-upper-quadrant pain, non-radiating, aggravated by oral intake and lying supine, partially relieved by leaning forward and analgesics. She had associated non-projectile, non-blood-stained vomiting for 7 days and low-grade intermittent fever for 5 days. She had no significant past medical, surgical, or social history: no abdominal trauma, ERCP, or pancreatic instrumentation, prior steroid or medication use, smoking, alcohol use, or drug allergies. She was initially admitted under General Surgery for suspected acute pancreatitis; after excluding a biliary or surgical cause, she was referred to General Medicine for associated leukopenia, fever, and serositis, with Nephrology involved after nephrotic-range proteinuria was detected. On examination, she was afebrile with stable vitals, pallor, and bilateral pitting edema with anasarca. The abdomen was moderately distended with right-sided tenderness and shifting dullness, without rebound tenderness, palpable masses, or organomegaly.

Laboratory Findings

The initial blood work revealed the following results:

The above mentioned findings are enclosed in **Table/Fig 1**.

Table/Fig 1: Laboratory Parameters

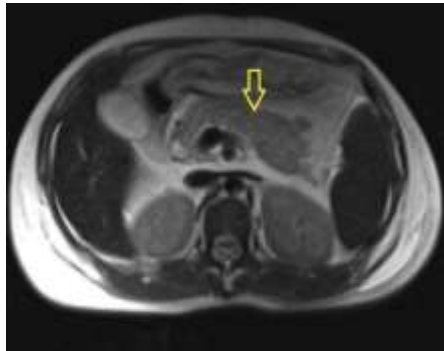
Test Category	Parameter	Result	Normal Values
Complete Hemogram	Hb	9 g/dL	M: 13.8-17.2, F: 12.1-15.1 g/dL
	WBC	2,720 cells/mcL	4,500-11,000 cells/mcL
	Platelet Count	1,78,000 cells/mcL	150,000-450,000 cells/mcL
	ESR	92 mm/hr	M: 0-15, F: 0-20 mm/hr
Liver Function	ALT	563 IU/L	7-56 U/L
	AST	188 IU/L	10-40 U/L
	ALP	879 U/L	44-147 U/L
	Albumin	1.4 g/dL	3.5-5.0 g/dL
	Total Protein	3.4 g/dL	6.3-7.9 g/dL
Electrolytes	Serum Calcium	7.8 mg/dL	8.5-10.5 mg/dL
Renal Function	Serum Creatinine	0.4 mg/dL	M: 0.74-1.35, F: 0.59-1.04 mg/dL
	eGFR	131 mL/min/1.73 m ²	≥90 mL/min/1.73 m ²
Pancreatic Enzymes	Serum Amylase	260 U/L	0-86 U/L
	Serum Lipase	735.8 U/L	13-60 U/L
Lipid Profile	Triglycerides	59 mg/dL	<150 mg/dL
Complement	C3	30.1 mg/dL	90-180 mg/dL
	C4	3.2 mg/dL	10-40 mg/dL
Autoimmune Serology	ANA (titer, pattern)	Positive 4+, 1:100, homogeneous	Negative
	Anti-dsDNA	Negative	Negative
	Anti-Smith (Anti-Sm)	Negative	Negative
	APLA panel	Normal	Normal
Urine Analysis	Protein/Creatinine Ratio	36.76	<0.2
	24hr Urine Protein	3800 mg/24 hrs	<150 mg/24 hrs
	RBCs	Plenty /HPF	0-4/HPF
Hemolysis Workup	Peripheral Smear	Normocytic normochromic RBCs, moderate anisopoikilocytosis, occasional fragmented RBCs and microspherocytes	No significant morphological abnormality
	Serum Haptoglobin	Normal	Normal
	Direct Coombs Test	Negative	Negative
	Lupus Anticoagulant	Not detected	Not detected
Viral Serology	HIV, HBsAg, HCV	All non-reactive	Non-reactive

Hb: hemoglobin; WBC: white blood cell count; ESR: erythrocyte sedimentation rate; ALT: alanine aminotransferase; AST: aspartate aminotransferase; ALP: alkaline phosphatase; eGFR: estimated glomerular filtration rate; C3: complement-3; C4: complement-4; ANA: anti-nuclear antibody; dsDNA: double-stranded DNA; Sm: Smith antigen; APLA: antiphospholipid antibody; RBC: red blood cells; HPF: high power field; HIV: human immunodeficiency virus;

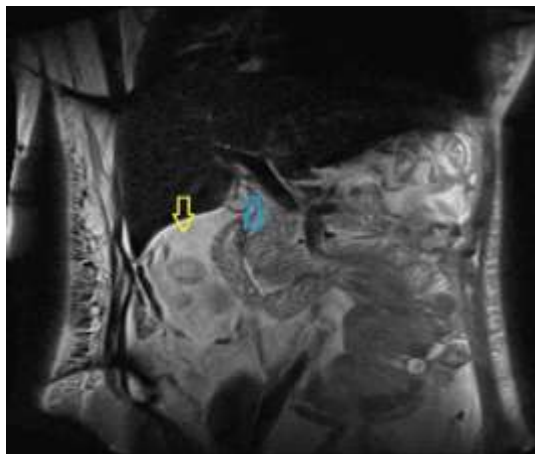
HBsAg: hepatitis B surface antigen; HCV: hepatitis C virus.

Imaging

Abdominal ultrasonography showed a bulky, edematous pancreas with a peripancreatic fluid collection, moderate-to-gross ascites, and bilateral minimal pleural effusion; both kidneys were normal with no cholelithiasis or biliary dilatation. MRCP confirmed a bulky pancreas with increased T2-weighted signal intensity, consistent with acute pancreatitis (**Table/Fig 2**).

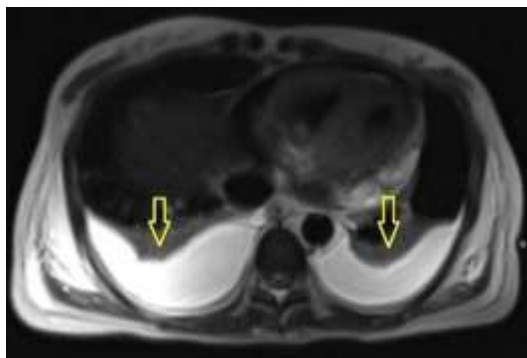


Table/Fig 2: SE axial view of MRI WITH MRCP Abdomen illustrating edematous pancreas (yellow arrow) Additionally, there was evidence of gross ascites (**Table/Fig 3**).



Table/Fig 3: Coronal view of T2 MRI WITH MRCP Abdomen illustrating the gross ascites(yellow arrow) and edematous head of the pancreas (blue arrow)

Also, it revealed bilateral moderate pleural effusion (**Table/Fig 4**).



Table/Fig 4: Axial STIR View of MRI WITH MRCP Abdomen illustrating moderate pleural effusion (yellow arrows)

CT of the Chest revealed bilateral moderate pleural effusion with mild pericardial effusion (Table/Fig 5).



Table/Fig 5: Axial view of PLAIN CT CHEST illustrating mild pericardial effusion (yellow arrows)

Echocardiography showed preserved left ventricular systolic function (EF ~60%) with trace pericardial effusion, no wall motion abnormality, and no pulmonary arterial hypertension.

Autoimmune panel and Additional Investigations

Autoimmune serology (ANA, anti-dsDNA, anti-Smith, APLA panel), complement levels, hemolysis work-up, and viral serology are summarized in Table/Fig 1; complement factors C3 and C4 were both markedly reduced. A kidney biopsy was performed, confirming class II lupus nephritis.

Diagnosis and Treatment

Based on clinical, laboratory, and histological findings, the patient was diagnosed with SLE with class II lupus nephritis and SLE-associated acute pancreatitis. She received intravenous methylprednisolone 500 mg once daily for three days as pulse therapy, followed by oral prednisolone 40 mg once daily with a planned taper, showing significant clinical improvement and hemodynamic stability throughout admission. Hydroxychloroquine, considered as first-line steroid-sparing therapy, was not tolerated by the patient, and mycophenolate mofetil was selected instead for renal and systemic disease control. She was discharged on tapering oral prednisolone, mycophenolate mofetil 500 mg twice daily, and telmisartan 40 mg once daily (an angiotensin receptor blocker, to reduce proteinuria), with advice to follow up in the General Medicine and Nephrology outpatient clinics after one week with repeat complete blood count, renal function tests, and urine protein-creatinine ratio. At follow-up, abdominal symptoms and fever had resolved, with reduced anasarca and improved function. Renal function was stable (creatinine unchanged) and albumin improved ~1 g/dL; protein-creatinine ratio not repeated. Corticosteroids and mycophenolate mofetil were well tolerated aside from mild GERD-type symptoms; blood pressure 100/60 mmHg.

Discussion

SLE is a chronic autoimmune disorder with multisystem inflammation and immune dysregulation [1]. Renal, musculoskeletal, hematological, and mucocutaneous involvement predominate; gastrointestinal disease, including acute pancreatitis (0.2–8.2% of cases), is less common but carries a high-stakes association with disease activity and poorer outcomes [2-5]. Its nonspecific presentation overlaps with far more common causes, so the diagnosis is frequently missed without a pre-existing SLE diagnosis, as here [6]. Diagnosis rested on characteristic abdominal pain, markedly elevated amylase/lipase, and MRCP evidence of pancreatic edema with a peripancreatic fluid collection and ascites, without biliary obstruction or structural abnormality [7].

Further evaluation revealed leukopenia, anemia, hypocomplementemia, nephrotic-range proteinuria, pleural effusion, and ANA positivity (homogeneous pattern). Although anti-dsDNA and anti-Smith were negative, the clinical and laboratory findings met the 2019 EULAR/ACR classification criteria for SLE with a cumulative score of 20, well above the threshold of 10 [12] (Table/Fig 6). Kidney biopsy confirmed class II lupus nephritis.

Entry criterion (positive ANA $\geq 1:80$, HEp-2): met (titer 1:100, homogeneous pattern). Only the highest-weighted

criterion within each domain is counted; a criterion is scored only if not better explained by another diagnosis.

Table/Fig 6: 2019 EULAR/ACR Classification Criteria Fulfilled by the Present Case

Domain	Criterion Met	Weight	Points Scored
Constitutional	Fever $\geq 38.3^{\circ}\text{C}$ – not confirmed (recorded temperature 98.5°F)	2	0
Hematologic	Leukopenia (WBC 2,720/mcL)	3	3
Serosal	Pleural and pericardial effusion	5	5
Renal	Biopsy-proven Class II lupus nephritis	8	8
Complement proteins	Low C3 and low C4	4	4
Neuropsychiatric / Mucocutaneous / Musculoskeletal / Antiphospholipid antibodies / SLE-specific antibodies	Not met in any of these domains	—	0
Total			20

The differential diagnosis also included thrombotic microangiopathy, prompted by anemia and occasional fragmented red cells on peripheral smear; however, the platelet count was normal throughout with no schistocyte-predominant picture, and a directed work-up was not pursued absent clinical suspicion. Viral hepatitis- and HIV-associated pancreatitis were excluded by non-reactive serology; hypercalcemia-induced pancreatitis was excluded by a normal albumin-corrected serum calcium (7.8 mg/dL measured, ~ 9.9 mg/dL corrected); and IgG4-related pancreatitis was considered unlikely given the absence of a focal mass, capsule-like rim, or ductal narrowing on MRCP, with no features of an overlapping connective tissue disease.

An unusual feature was severe hypoalbuminemia and nephrotic-range proteinuria despite biopsy-confirmed class II lupus nephritis, typically associated with only mild proteinuria; sampling limitation or early, patchy membranous change cannot be excluded. Immunofluorescence and electron microscopy were unavailable at our resource-limited institute, precluding formal activity/chronicity indexing; classification rests on light microscopy and clinical correlation alone, a recognized limitation of this report.

Pathogenesis of lupus-associated pancreatitis is incompletely understood but is thought to involve small-vessel vasculitis, immune-complex deposition, complement activation, and thrombosis [5,10]; drug-induced pancreatitis (corticosteroids, azathioprine, diuretics) is a recognized alternative in established SLE, though our patient had no prior exposure [11]. Pancreatic involvement remains uncommon even in large cohorts: only 4% of 1,811 patients in the Hopkins Lupus Cohort developed SLE-related pancreatitis [8], and a comparative review found an overall prevalence of 1.34%, five-fold higher in pediatric- than adult-onset disease [9] — placing our fourth-decade patient among the minority of adult-onset sentinel cases. Of the Hopkins Cohort's strongest risk factors — hypertriglyceridemia, psychosis, pleurisy, gastritis, anemia [8] — she shared only pleurisy and anemia, with normal triglycerides (59 mg/dL) and no neuropsychiatric involvement, suggesting an immune-complex/complement-mediated mechanism, consistent with markedly low C3/C4. Mortality was 14.8% in adult-onset versus 53.8% in

pediatric-onset disease, with episode severity — not age — the strongest predictor of death [9]; her uncomplicated course and prompt corticosteroid response place her favorably, though improvement after corticosteroids is compatible with, rather than proof of, an immune-mediated mechanism.

Acute pancreatitis as an initial SLE manifestation was documented only nine times in the English literature as of the earliest indexed report [15]; closer to home, Bairy et al. later reported three such cases in this journal [16]. Jia et al. described a 23-year-old woman whose preceding leg swelling proved, in retrospect, an early clue to systemic disease [4] — as in our patient, an overlooked extra-pancreatic feature (anasarca and leukopenia, here) prompted the autoimmune work-up. Gupta et al. reported a 30-year-old woman with initial-presentation pancreatitis, class II nephritis, and 2.5 g/24h proteinuria, improving on corticosteroids and an ACE inhibitor [13]; Li and Li described a 47-year-old woman with sentinel pancreatitis and biopsy-proven class II nephritis complicated by antiphospholipid syndrome [14]. Our patient shares this triad but is distinguished by nephrotic syndrome disproportionate to that histology — albumin 1.4 g/dL, anasarca, and 3.8 g/24h proteinuria — with more pronounced hypocomplementemia, leukopenia, and serosal involvement, absent anti-dsDNA, anti-Smith, or antiphospholipid positivity: a combination not previously reported.

Conclusion

This case underscores the importance of early recognition of SLE-associated acute pancreatitis: once common causes are excluded, prompt pulse corticosteroid therapy should be strongly considered, as it can prevent further pancreatic damage and improve prognosis. A thorough autoimmune work-up identified concurrent class II lupus nephritis, illustrating the multisystem nature of SLE. Acute pancreatitis of unclear etiology, particularly with multisystem features, should prompt consideration of SLE.

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