Necrotizing pancreatitis in a patient diagnosed with SLE; rare case report

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Introduction

Systemic Lupus Erythematosus (SLE), is a chronic autoimmune disease that can affect various organs and tissues throughout the body. In this autoimmune disease multiple organ systems including the skin, joints, kidneys, heart, pancreas, lungs, brain, blood cells, are involved (1).

Necrotizing pancreatitis (NP) is a severe and potentially life-threatening inflammatory condition affecting the pancreas. NP is characterized by tissue death, can occur in individuals with various underlying conditions, and SLE is among the potential contributors. While lupus primarily affects the joints, skin, kidneys, and other organs, it can also impact the pancreas. Pancreatic involvement in SLE may present with a range of symptoms, including abdominal pain, nausea, and vomiting (2).

Case presentation

We present a case report of a 17-year-old girl with problems that started a year ago in the form of multiple bruises on the whole body and bleeding from the gums, who was diagnosed with ITP (Immune thrombocytopenic purpura) and treated with corticosteroids and IVIG (Intravenous immune globulin). The patient had no problems until two months ago. Until she was hospitalized due to leukopenia and seizures. After investigations, proteinuria of 1100 mg/dL, lymphopenia and leukopenia, positive ANA and anti-ds DNA and reduced complement was detected (Table1). During hospitalization, due to abdominal pain and swelling and Shifting Dullness in the examination, CT scan of the abdomen and pelvis with oral and intravenous contrast is also performed for the patient (Figure1). Necrotizing pancreatitis is reported with involvement of less than 30% of the pancreas in addition to the presence of abundant free fluid in the abdomen and pelvis and bilateral pleural effusion. According to the seizure, MRI/MRV was performed for patient, MRI reported was "Diffuse hyperintensity is seen at subcortical white matter of high frontal lobes and parieto-occipital lobes bilaterally, is reported and also MRV is reported as normal. In order to investigate other causes of pancreatitis, MRCP was performed for the patient, which was reported as a normal. Finally, according to the history of thrombocytopenia, leukopenia and lymphopenia, proteinuria, seizures, positive ANA and Anti dsDNA and reduced complement, SLE was diagnosed and the patient was treated with methylprednisolone pulse for 3 days. And then she was treated with prednisolone at a dosage of 1 mg/kg and cyclophosphamide puls, with a dose of 500 mg. Other potential causes of pancreatitis were investigated, but no pathological findings were found. The patient's pancreatitis was justified in the context of SLE. During her hospitalization, she received routine treatment for pancreatitis. Currently, during the follow-up, cytopenia and ascites have improved, seizures have not recurred, and 24-hour proteinuria is at the level of 566 mg/dl. The patient is received the third dose of cyclophosphamide and 20 mg of prednisolone and 200 mg of hydroxychloroquine daily.

Discussion

In this case, we reported a 17-year-old person who constellation of her symptoms raises the suspicion of both SLE and necrotizing pancreatitis. Pancreatitis is a medical condition characterized by the inflammation of the pancreas; Symptoms of pancreatitis typically involve severe abdominal pain.

Based on the nature, duration, and characteristics of the inflammation affecting the pancreas, pancreatitis classified into 3 groups: Acute Pancreatitis (AP), Chronic Pancreatitis, Necrotizing Pancreatitis. Necrotizing pancreatitis is a severe form of AP characterized by the death of pancreatic tissue account for relatively high morbidity and mortality rate (3).

AP, as a result of SLE is more common rather than necrotizing pancreatitis (4). While AP manifests more frequently than necrotizing pancreatitis in patients with SLE, timely medical intervention in the acute inflammatory stage may attenuate progression to pancreatic necrosis in this population (5). SLE can affect small blood vessels, resulting in inflammation known as vasculitis (6). When this inflammation occurs in the blood vessels supplying the pancreas, it can restrict blood flow to the organ. This impaired perfusion deprives the pancreatic tissue of oxygen and nutrients. The ischemia and resulting dysfunction may then trigger the onset of AP in susceptible individuals (7). Beyond the pancreas, vasculitis damage from SLE can impact organs throughout the body.

The pathophysiological underpinnings of SLE and pancreatitis encompass intricate molecular, immunological, and genetic interactions (8). SLE has a substantial genetic component, which engenders immune perturbation aberrant B and T lymphocyte reactivity, cytokine disproportion, and epigenomic alterations critically regulate disease propagation. The hallmark interferon-alpha signaling cascade overload characterizes SLE immunopathology.

In comparison, pancreatitis molecular factors like genetic controllers of pancreatic enzymes, inflammatory mediators, and autoantibodies (notably in autoimmune pancreatitis) are operant. Premature pancreatic digestive enzyme activation, oxidative injury, and genetic liability collectively fuel pancreatitis' development.

Despite differing specifics, SLE and pancreatitis share an immunoregulatory breakdown and inflammation as

a cardinal unifier. Both conditions exhibit the loss of self-tolerance, enabling autoimmune injury and tissue damage(9). Comprehending the molecular intricacies is vital to further enable targeted therapeutics and superior handling of these compound autoimmune and inflammatory illnesses. Recent studies have been shown that patients with SLE often have higher rates of metabolic conditions like diabetes and hypertriglyceridemia (10). These are also independent risk factors for developing pancreatitis. Also, Cytokine imbalances due to overactive immune systems in SLE may promote inflammation in various organs, including theoretically the pancreas.

While numerous studies and reports extensively cover AP, there is a scarcity of literature and research focusing on necrotizing pancreatitis due to its rarity.

To the best of our knowledge, although there are several case reports of lupus-associated AP, only one case of lupus-associated necrotizing pancreatitis has been reported. In July 2014, a 37-year-old woman of African descent, previously diagnosed with SLE, presented with sudden abdominal pain, accompanied by nausea and vomiting. Despite being under ongoing treatment with mycophenolate mofetil (MMF), prednisolone, hydroxychloroquine (HCQ), and belimumab, she encountered recurrent episodes of AP. Diagnostic assessments, including laboratory tests and imaging, verified the presence of pancreatitis with 20% necrosis. The medical team suspected SLE-induced pancreatitis, prompting the administration of high-dose steroids. This intervention yielded favorable responses in both clinical symptoms and biochemical markers. Following this, the patient's ongoing care plan incorporated rituximab for enhanced disease management. A subsequent follow-up examination indicated notable improvement in the pancreatitis condition. This case highlights the complexities of addressing complications associated with SLE and underscores the effectiveness of tailored approaches, including high-dose steroids and rituximab, in managing AP and achieving positive patient outcomes (11).

Also, retrospective studies indicate that 0.9% to over 5% of SLE patients suffer from AP. Among 264 SLE patients studied, predominantly female with an average age of 31.4 years, abdominal pain was a consistent symptom. AP was more common in those with shorter disease duration, high activity scores, and multiorgan involvement. Diagnosis as "idiopathic" SLE-related AP was based on guidelines, excluding other causes. Managing SLE-related AP is challenging due to the lack of a standardized approach, and the use of glucocorticoids is debated due to potential complications. Monitoring serum lipase levels after high-dose steroids is suggested. Some studies suggest a positive outcome with plasma exchange, and the occurrence of AP in SLE may signal an association with macrophage activation syndrome, with reported mortality rates of up to one third of cases (12).

Conclusion

In conclusion, our article presented a female with SLE, developed necrotizing pancreatitis, which is a rare complication of SLE.

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Author contributions

All authors approved the final manuscript.

Availability of data and material

Considering that this study is about a rare disease availability to patients and data may hard to some extent.

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Competing interests

The authors declare that they have no competing interests

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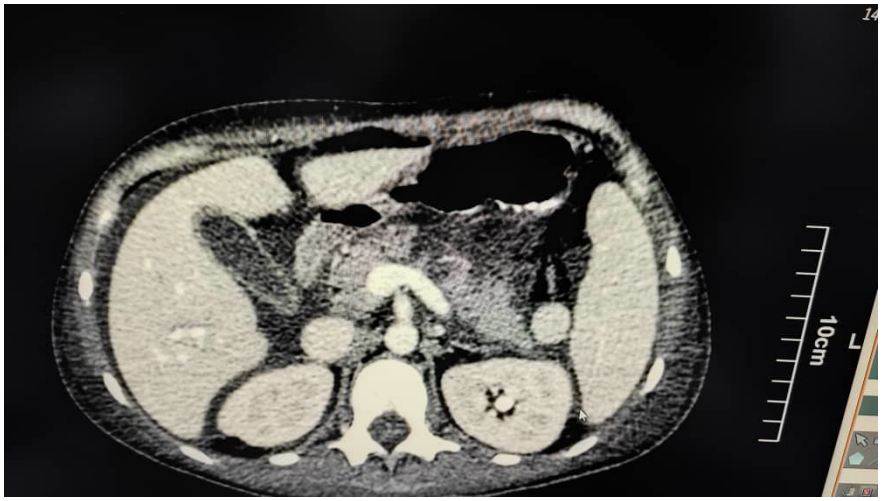

Figure1. Necrotizing pancreatitis

Table 1: Laboratory parameters of the patient

Test	Result	Normal range	Unit
ESR	40/20	For males: 0-15 mm/hr For females: 0-20 mm/hr	mm/hr
CRP	Negative	Less than 10 mg/L	mg/L
F ANA	12	1	-
Anti dsDNA	349	30	-
Antiphospholipid	Negative	-	-
C3	0.4	0.9 – 1.8	g/dL
C4	0.03	0.1 -0 .4	g/dL
CH50	30	50-150	g/dL
BUN	24	8-20	mg/dL
Ca	normal	-	-
Ph	normal	-	-
TG	normal	-	-
Creatinine	0.8	adult males: 0.6 - 1 adult females: 1.3 0.5 - 1.1	mg/dL
AST	31	10-40	U/L
ALT	27	7-56	U/L
ALKP	144	44-147	U/L
Amylase	normal	1 to 8	U/L
Lipase	360 u/l	100	U/L
Baseline WBC	2500	4,500 to 11,000	cells/μL

Test	Result	Normal range	Unit
Final WBC	4660		
Baseline HB	7.5 g/dl	males: 13.8 to 17.2 g/dL females: 12.1 to 15.1 g/dL	g/dL
Final HB	11 g/dl	80 to 100	Femtoliters
PLT	226000	150,000 to 450,000	platelets/μL
Baseline Urine protein 24 hr	1083 mg/dl		
Final Urine protein 24 hr	566 mg/dl		

ESR: Erythrocyte Sedimentation Rate, CRP: C-Reactive Protein, F ANA: Fluorescent Antinuclear Antibody, Anti dsDNA: Anti-Double-Stranded DNA Antibodies, C3: Complement Component 3, C4: Complement Component 4, CH50: Complement Total Blood Test AST: Aspartate Aminotransferase, ALT: Alanine Aminotransferase, ALKP: Alkaline Phosphatase, LDH: Lactate Dehydrogenase, WBC: White Blood Cell Count, HB: Hemoglobin, PLT: Platelet Count, BUN: Blood Urea Nitrogen, CA: Calcium, Ph: Phosphors, TG: Triglyceride

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