# The simultaneous incidence of acute pancreatitis and autoimmune hemolytic anemia: a rare duo in a patient with SLE

## Das gleichzeitige Auftreten von akuter Pankreatitis und autoimmunhämolytischer Anämie: ein seltenes Zusammentreffen bei einer Patientin mit SLE

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## Abstract

A young female presented with acute abdominal pain of two days duration consistent with acute pancreatitis. During her stay in the hospital she had a sudden drop in hemoglobin to 6 g/dl without any overt blood loss. On evaluation, it was evident that she had acute pancreatitis, in addition to displaying features of autoimmune hemolytic anemia. She had been a known case of systemic lupus erythematosus (SLE) and had discontinued her treatment. She was managed with methylprednisolone pulse therapy. Her clinical condition improved, and she has been regularly attending our clinic for the last 2 years. According to a literature search in Medline, it would appear that this is the first report of a case in which SLE with autoimmune hemolytic anemia has been associated with acute pancreatitis in a single case.

**Keywords:** systemic lupus erythematosus (SLE), acute pancreatitis, autoimmune hemolytic anemia, massive hemolysis

## Zusammenfassung

Eine junge Frau stellte sich mit akuten Bauchschmerzen von zwei Tagen Dauer, die auf eine akute Pankreatitis hinwiesen, vor. Während ihres Aufenthaltes im Krankenhaus erlitt sie einen plötzlichen Abfall des Hämoglobins auf 6 mg/dl ohne offenen Blutverlust. Die Untersuchung ergab, dass sie eine akute Pankreatitis hatte und zusätzlich Symptome einer autoimmunhämolytischen Anämie zeigte. Die Patientin hatte eine bekannte Vorgeschichte von systemischem Lupus erythematodes (SLE) mit Behandlungsabbruch. Sie wurde mit Methylprednisolon-Pulstherapie behandelt. Ihre klinischer Zustand verbesserte sich, und sie wird seit den letzten 2 Jahren regelmäßig in unserer Klinik untersucht. Nach einer in Medline durchgeführten Literaturrecherche scheint es, dass dies der erste Bericht über einen Fall ist, in dem SLE mit autoimmunhämolytischer Anämie und akuter Pankreatitis zusammen auftritt.

**Schlüsselwörter:** systemischer Lupus erythematodes (SLE), akute Pankreatitis, autoimmunhämolytische Anämie, massive Hämolyse

## Introduction

Systemic lupus erythematosus (SLE) is an autoimmune inflammatory disease characterized by the presence of a plethora of autoantibodies and immune complex formation targeted to various organs of the body. SLE presents with mucocutaneous, musculoskeletal and/or renal involvement. Hematological disturbances are also common in SLE. Anemia in SLE is multi-factorial and can be part of the disease process or a complication of the treatment itself. Autoimmune hemolytic anemia (AIHA) is often associated with thrombocytopenia, lupus nephritis, and central nervous system activity. In comparison, common gastrointestinal (GI) manifestations of SLE are non-specific and reflect either lupus of the GI tract or the effects of medication. Serious gastrointestinal manifestations of SLE can include mesenteric vasculitis, acute pancreatitis and intestinal pseudo-obstruction. This report describes

a rare combination of hematological and gastrointestinal complication found in a patient with SLE flare.

## Case description

A 22-year-old woman came to the emergency department of the King Fahad Medical City in Riyadh, a tertiary care center in Saudi Arabia, presenting with history of acute abdominal pain of 2 days duration. She described the pain as severe (score of 6/10). The pain was in the epigastrium with reference to the back. There was no history of offending drug intake, jaundice or fever. She had also had two episodes of non-bilious vomiting She had been diagnosed as suffering from SLE (American College of Rheumatology ACR, criteria 5/11) 5 years prior to the current presentation, but had discontinued treatment of her own volition. On examination she showed stable vitals, with mild epigastric tenderness and no organomegaly. Her systemic examination was unremarkable. She had a hemoglobin of 9.8 g/dl, a leucocytic count of 11,500 and a platelet count of $110\times10^3$. Her serum amylase was 380 IU (Ref. 110 IU) and the lipase levels were 850 IU. She was managed as a case of acute pancreatitis, was kept nil per mouth and was given normal saline intravenously. Her abdomen CT scan was consistent with grade B pancreatitis (Balthazar classification). She had a normal hepatobiliary system, and her serum lipids and serum calcium levels were also normal. On further evaluation, it appeared that she had an elevated ESR 66 mm/1$^{st}$ hr. Her antinuclear antibody (ANA) levels were elevated 1:60 (reference 1:35 IU/ml), as were her double stranded DNA (DsDNA) levels (>19 AU/ml). During the hospital stay, her hemoglobin dropped further to 6 g/dl. She had no bleeding, and further evaluation was suggestive of acute hemolysis (positive direct Coombs test, indirect hyper-bilirubinemia 8.6 g/dl, corrected retic count of 4%, elevated serum lactate dehydrogenase (LDH) 822 U/L). As a result of her past diagnosis of SLE, she was managed with pulse methylprednisolone 500 mg IV for 3 days. Her abdominal pain settled and her hemoglobin levels remained stable. She was later started on tab. prednisolone 40 mg once daily, which was tapered on follow-up. She has been asymptomatic for the past 2 years, and is regularly attending our clinic.

## Discussion

The diagnosis of acute pancreatitis is an important differential in the evaluation of abdominal pain in a patient with SLE. The patients usually present with pain of variable intensity. Several factors have been implicated in the pathogenesis of this condition, such as vasculitis, drugs and antiphospholipid antibodies. The role of corticosteroids as an etiologic factor remains controversial. Ben et al. described acute pancreatitis in six SLE patients, with favorable outcomes following steroid treatment [1]. Recently, data on 4,053 SLE patients from China showed the overall prevalence of pancreatitis to be 0.67%, an annual incidence of 0.56% with a mortality rate of 37.04%. The authors of this study concluded that a combination of SLE activity and multiple organ system involvement were predictors both of severity and mortality due to acute pancreatitis in SLE patients. In this study, further glucocorticoid treatment was considered to lead to a better outcome in a majority of SLE patients [2].

The index case had been diagnosed with SLE 5 years prior to her current presentation and she had discontinued treatment of her own accord. It is quite possible that, in her case, stopping the maintenance dose of steroids was responsible for both the acute pancreatitis and the acute autoimmune hemolytic anemia (AIHA). The association of acute pancreatitis in a cohort study of 551 SLE patients included the withdrawal of a maintenance dose of steroids, the presence of seizures and of arthritis, and the authors of this study found no independent predictor of this complication [3]. In yet another study, hypertriglyceridemia appeared to be a strong associate of pancreatitis in SLE patients. Authors in this study further observed that SLE patients with psychosis and pleurisy were at increased risk for pancreatitis [4]. The index case had a normal lipid profile and no CNS or pleural involvement.

The combination of SLE and acute pancreatitis is a very serious complication, which could even be fatal, as reported by Cairoli et al. [5]. The spectrum of SLE is not solely restricted to acute pancreatitis as the association of SLE with chronic pancreatitis has been reported as well [6]. In addition to her acute pancreatitis, the index case also developed features of acute autoimmune hemolytic anemia during her hospital stay. Overt autoimmune hemolytic anemia has been noted up to 10% of SLE patients [7]. AIHA may manifest itself in SLE patients at the time of diagnosis, or within the first year following diagnosis. In this case, however, the index case presented with this complication 5 years after the diagnosis. AIHA is often associated with thrombocytopenia, lupus nephritis and CNS activity. To address the natural history and treatment response in AIHA among SLE patients, Gomrad et al. [8] studied 26 patients with severe isolated AIHA. The authors observed that AIHA is a serious complication of SLE, requiring urgent and appropriate management. In their study, patients responded to steroids in 96% of the cases and a recurrence rate of only 3 per 100 person-years was observed. Furthermore, in the same study, 7 patients (27%) experienced a relapse of AIHA and required immunosupressants. Based on their results, the authors recommended steroids as the first line of treatment, and advocated additional therapy with rituximab, danzol in refractory cases. Our index case responded well to the steroid treatment, with no relapse over the two years of follow-up.

Massive hemolysis due to any reason has been found to be another cause of acute pancreatitis. Drum et al. [9] retrospectively studied acute hemolysis due to different reasons in forty patients with drop of hematocrit greater than 12% over a period of 12 hours. Pancreatitis was observed to be a complication of massive hemolysis with

a prevalence of above 20%. The authors were of the opinion that the back pain often found in acute hemolysis may originate from the pancreas rather than the kidneys. Experimental models have supported massive hemolysis as a cause of acute pancreatitis, and, in addition, histological changes following massive hemolysis have been observed in the pancreas in these experimental models [10].

Having said this, it is pertinent to mention here that the index case presented with acute pancreatitis and developed severe hemolysis during her stay in hospital, so there seems to be no temporal relation between the two (acute pancreatitis with hemolysis). However, it is possible that low grade hemolysis taking place a few days before the manifestation of the acute pancreatitis actually precipitated the attack, as she had no other established risk factor of acute pancreatitis. We cannot rule this out with any certainty. However, it is certain that due to massive hemolysis during her hospital stay, her pancreatitis could have worsened had she not been managed in time.

In conclusion, it may be said that the present case brings to the fore the multiple complications of SLE, a clinical scenario due to the generation of multiple autoantibodies directed towards the pancreas and the hemopoietic system. Acute vasculitis with its varied clinical presentation is often the commonest cause of abdominal pain in SLE, but acute pancreatitis and acute autoimmune hemolytic anemia, with its consequences, should be kept in mind in a given case of SLE. Further, this case emphasizes the need for close follow-up and proper health education in an SLE patient in order to circumvent any complication.

# Notes

## Competing interests

The author declares that he has no competing interests.

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