Case Report

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Concurrent occurrence of acute pancreatitis and intracerebral hemorrhage as presenting manifestations in lupus: a case report

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Introduction and importance: Systemic lupus erythematosus (SLE) is a rare autoimmune condition that may affect almost every organ system and has a wide range of disease severity. It is characterized by a spectrum of clinical manifestation, a plethora of autoantibodies, and immune complex formation. The symptoms can come from any organ system, alone or in a group, and they can be of any severity, which makes diagnosis and prognosis difficult.

Case presentation: The authors hereby present the case of an 18-year-old female with chief complaints of fever, abdominal pain, headache, vomiting, and loss of vision. She was diagnosed with acute pancreatitis (AP) and intracerebral hemorrhage (ICH) with an etiology linked to SLE. SLICC criterion was used to diagnosed SLE while ATLANTA criteria for AP and neuro-radiological findings for ICH. Emergency temporo- parietal-occipital-osteoplastic craniotomy was done for ICH as well as started with immunosuppressive therapy for SLE. On the 18th day of admission, she was discharge with maintenance medications for SLE. While the vision took over a month to come to a premorbid state, she was clinically improved within 2 weeks of admission.

Clinical discussion: Clinical manifestation of SLE vary greatly. AP and intracranial bleeding are few of the rare presentation of SLE. Acute presentation of both conditions in an otherwise healthy individual in the initial course of disease left the clinician with a wide array of differentials. Literature shows very little evidence of co-occurrence of ICH and pancreatitis as an initial manifestation in SLE patients. The exclusive diagnosis of these potentially fatal condition is made holistically with clinical, biochemical, and radiological parameters.

Conclusion: SLE may present with atypical, life-threatening initial manifestations. Early diagnosis and timely intervention in therapy can lead to successful management. The treating physician must consider, SLE when a straightforward diagnosis is associated with inexplicable multiple concomitant abnormalities, especially in young women.

Keywords: acute pancreatitis, case report, cerebral hemorrhage, systemic lupus erythematosus

Introduction

SLE is a complex, polymorphic, chronic autoimmune connective tissue disorder. It is a multisystem inflammatory disease of unknown etiology with the predominance of prevalence in women of childbearing age. Whatever the underlying cause, it is characterized by a global loss of self-tolerance with activation of auto-reactive T and B cells leading to the production of pathogenic autoantibodies, immune complex formation, and deposition which triggers complement and other mediators of inflammation, leading to tissue injury and organ damage[1]. Virtually damage to any cell is possible, leading to the possibility of heterogeneous manifestations depending upon the organ involvement of variable severity, ranging from mild to very severe and life-threatening. This makes the clinical profile of lupus often challenging[2].

HIGHLIGHTS

- Systemic lupus erythematosus (SLE) related pancreatitis and intracranial bleeding as initial manifestations are documented barely, individually.
- No paper documents the concurrent occurrence of SLE-related pancreatitis and intracranial bleeding at presentation, both of which are potentially fatal conditions. They are exclusive diagnoses made based on clinical, biochemical, and radiological parameters.
- SLE may present with atypical life-threatening initial manifestations. Early diagnosis and timely intervention of therapy can lead to successful management. The treating physician must consider SLE when a straightforward diagnosis is associated with inexplicable multiple concomitant abnormalities, especially in young women.

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This study reports a rare, atypical case of concurrent occurrence of SLE-related acute pancreatitis and intracerebral bleeding as initial manifestations, both of which are potentially life-threatening conditions if not diagnosed and managed rightly. This case report follows Surgical CAse REport (SCARE) guidelines[3].

Case report

An 18-year-old female was referred to our hospital when she experiences sudden onset of a throbbing headache in the occipital region, which is accompanied by loss of vision in the left eye as well as a decline in her consciousness. Prior to coming to our hospital, she had been admitted twice in the past 10 days for complaints; fever, abdominal pain, vomiting, severe headache with no significant improvement. As stated by patient party, patient had on and off fever for 20 days, associated with chills and rigor. She had been vomiting for 8 days ago with acute onset of dull left upper quadrant abdominal pain.

In past history, she tested positive for severe acute respiratory syndrome coronavirus 2 (2019-n-CoV) via Real-time Reverse Transcription – Polymerase Chain 1 month ago, during which she had mild painful swelling of the right knee and right ankle. However, she did not seek any medical attention for it. She was symptomatically better and tested negative after 15 days of home isolation. She denied prior cholelithiasis, hypertension, diabetes, alcohol consumption, illicit drug use, abdominal trauma, use of over-the-counter drugs, or herbal remedies. Her family history is unremarkable for autoimmune disorders and heritable conditions.

On examination, she had a Glasgow coma scale score of 12 (eye-opening: 3, verbal response: 3, motor response: 5). The blood pressure was 160/80 mmHg, and the temperature was 103F. Her abdomen was soft with mild tenderness in the left upper quadrant. Normal bowel sounds without rebound tenderness, guarding, rigidity, and contusions were noted. Bilateral basal crepitation was heard over the bilateral lower lung fields. The power of 5/5 was present in all four limbs. On ophthalmic examination, unaided visual acuity was limited to hand movement in the left eye.

Laboratory blood investigations revealed anemia (Hemoglobin: 9.2 gm/dl). Packed cell Volume and Red Blood Cell count were both decreased with values of 29.1% and 3.2 million/ cu.mm, respectively. The total leukocyte count was 7200/cumm. The differential count was Neutrophils 80%, lymphocytes 10%, eosinophils 5%, and monocytes 5%. The platelets count was normal. Liver function test showed hypoproteinemia (serum total protein: 8.6 gm/dl) and hypoalbuminemia (serum albumin: 3 gm/dl). However, total bilirubin, direct bilirubin, and liver enzymes were normal. In renal function tests, serum urea, creatinine, and sodium values were within the normal range. However, serum potassium was low (3.2 meq/l). Serum amylase and lipase were elevated (825 U/L and 500 U/L, respectively). The erythrocyte sedimentation rate was high (60 mm/h). Prothrombin time/international normalized ratio, urine routine examination, 24 h urine creatinine, and microalbumin were within normal limits Table 1.

On radiological examination, the following findings were made:

The findings were consistent with early subacute intraparenchymal hematoma. The diagnosis of acute pancreatitis and intracerebral bleeding was made based on ATLANTA criteria and neuro-radiological findings, respectively.

An emergency left temporo-parietal-occipital osteoplastic craniotomy with evacuation of hematoma with lax duraplasty was performed by a team of neurosurgeons. Approximately, 100 ml of dark brown hematoma was evacuated from the left parietal region. The histopathology report was consistent with hematoma.

Meanwhile, further investigations to establish the cause and rule out complications were carried out. This young woman's acute evidence of simultaneous clinical presentation prompted our team to consider the possibility of a connective tissue disorder. As a result, a workup was performed, and the results of that workup are as follows:

A rheumatology consultation was done. Based on clinical and laboratory criteria, the diagnosis of SLE was made. Also, the diagnosis of SLE-related pancreatitis and intracerebral bleeding was made after ruling out the non-SLE causes of them, respectively. Together with the values of the panel for antiphospholipid syndrome and MR venogram findings, an association of antiphospholipid syndrome (APLA) was made. To rule out if there was renal involvement of lupus, a renal doppler evaluation was performed, which revealed normal findings.

Postoperatively, the patient's treatment was started in the ICU. The patient was managed symptomatically with strict blood pressure control, medical measures to lower intracranial pressure and started on immunosuppressive therapy (prednisolone, hydroxychloroquine, and mycophenolate mofetil).

Anemia was persistent despite repeated whole blood transfusions and further workup was done. Coombs' test was positive and a peripheral blood smear showed predominantly normocytic normochromic red blood cells with anisopoikilocytosis. A few ovalocytes and macrocytes were seen. SLE associated hemolytic anemia was assumed and the patient was managed with an increased dose of glucocorticoids and blood transfusions.

There was gradual clinical improvement in the patient and she was discharge on the 18th day of admission. Her vision in the left eye gradually improved over a month to a premorbid state. The patient has been on monthly follow-up for 14 months regularly until now. She reports no persistence of symptoms. An MRI of the brain with an MR Venogram done on 6 and 12 months follow-up shows no mass, bleed, or collection. However, chronic

Table 1

Radiological findings of abdominal computed tomography (CT) and MRI

Test with image	Findings
Abdominal computed tomography (CT)	A mildly bulky body of the pancreas with the rest being normal in size, outline, and attenuation. (As shown on arrowhead) Splenomegaly with a spleen size of 12.9 cm in craniocaudal span with the normal outline, attenuation.
MRI of abdomen (Plain T2 weighted)	Bulky and edematous pancreatic tail with a smooth outline and minimal peripancreatic fat stranding suggestive of mild focal pancreatitis. (As shown on arrowhead).

Table 2

Radiological findings of Magnetic Resonance (MR) imaging of head and orbit (plain T2 weighted) and MR venogram

Test with image	Findings
Magnetic Resonance (MR) imaging of head and orbit (plain T2 weighted)	Early subacute intraparenchymal hematoma measuring 17 × 13 × 12 mm in the left occipital lobe and cystic encephalomalacia with hemorrhagic foci in the right frontoparietal lobe. (As shown on arrowhead) No obvious abnormalities were seen in bilateral optic nerves and pathways.
MR Venogram	Asymmetrical narrowing of the lumen size of the sigmoid and transverse sinus on the left side with the minimal flow within the lumen. (As shown on arrowhead) No flow was seen within the lumen of the left Intra-Jugular Vein. Findings were suggestive of chronic thrombosis.

thrombosis is persistent. Repeat values of serum amylase and lipase were within normal limits Table 2.

Currently, the patient is maintaining well on prednisone, hydroxychloroquine, mycophenolate mofetil, losartan, and warfarin.

Discussion

Systemic lupus erythematosus, also known as SLE, is a type of autoimmune disease that can affect multiple body systems at once. The condition can manifest itself in a variety of phenotypes, with clinical manifestations ranging from mild mucocutaneous manifestations to severe involvement in multiple organ systems and the central nervous system[4].

Gastrointestinal system involvement is not a rare entity in SLE. Over 30–40% of patients at some point in life experience GI symptoms[5]. SLE occasionally involves the pancreas. Immune complex-induced vasculitis is the most often documented mechanism, albeit the underlying pathophysiology is unclear. It is crucial to know that pancreatitis in SLE can be the manifestation of the disease process. Subclinical pancreatitis is substantially more common than symptomatic pancreatitis in terms of incidence, and the use of steroid in SLE may conceal the pain of pancreatitis[6]. While our case was symptomatic and the diagnosis was made mostly based on clinical signs and symptoms, together with noticeably raised lipase and amylase levels, and MRI findings.

Another, most intricate manifestations of the disease is neuropsychiatric SLE (NPSLE)[7]. According to estimates, 43–75% of SLE patients also have neuropsychiatric syndromes, with headache, mood disorder, cognitive dysfunction, seizure, and acute cerebrovascular illness being the most prevalent NPSLE disorders[8]. It is appropriate to take into account multifactorial pathways related to antibody-induced damage, antiphospholipid prothrombotic impact, noninflammatory vasculopathy, and cytokines-mediated cytotoxicity when discussing etiology[8]. Cerebrovascular diseases are particularly concerning because they have an estimated prevalence of 5–15% among SLE patients and influence their overall health and outcome[9]. As a part of NPSLE, ischemia stroke is more often encountered rather than a bleed, which was not in line to our case. In between 0.4 and 7% of instances, cerebral hemorrhage is noted, and it is typically brought on by natural or unavoidable causes (e.g. arterial hypertension, thrombocytopenia, or anticoagulation, aneurysm), however, an independent risk factor for cerebral bleeding was discovered to be thrombocytopenia[9,10]. The prognosis was quite good as 65% showed excellent recovery at discharge, which is similar to our case[9].

The hematological abnormalities in our case are thrombosis and anemia. Thrombosis was present on initial workup as an asymmetrical narrowing of the lumen size of the sigmoid and transverse sinus of the left side evident on the MR Venogram. Subsequent evaluation after 6 months showed similar findings suggestive of chronic thrombosis with no clues of acute thrombosis. Similarly, there was persistence of autoimmune hemolytic anemia days following treatment, which can be ascribed to anticardiolipin antibody and lupus anticoagulant and is well supported by other literature[11]. Hematologic and gastrointestinal complications had a substantial correlation with CNS involvement; however, not much literature is evident on databases[12]. Antiphospholipid antibodies were also positive in our case. These antibodies are evident in healthy young individuals with a prevalence below 5%. As with aging, the titers increase, however, more increment will be observed in a patient with a chronic coexisting illness like SLE[13].

The primary objective in treating individuals diagnosed with SLE is to minimize disease activity, safeguard organ function, and prevent permanent damage. To alleviate the symptoms associated with lupus, healthcare providers often prescribe nonsteroidal anti-inflammatory drugs (NSAIDs), anti-malarial medications, and corticosteroids. As the disease progresses and symptoms worsen, higher doses of corticosteroids and immunosuppressive drugs are commonly administered to slow down or halt its progression[14]. However, from the point of view of the patient, one of the primary goals is to preserve the patient's functional ability and quality of life Table 3.

Approximately half of individuals diagnosed with SLE experience symptoms related to their digestive system. However,

Table 3

Laboratory findings of workup for connective tissue disorder

Test	Result	Reference Range
Antinuclear Antibody (ANA)	Positive	
Anti-ds DNA	Positive (> 800 IU/ml)	< 100
Anti-Smith	Positive (46.9 RU/ml)	Positive: > 11
Anti-Histone	Positive (115.3 RU/ml)	Positive: > 11
Anti-Ribosomal	Positive (31.3 RU/ml)	Positive: > 11
Complement C3	Low (71.6 mg/dl)	85.00–160.0
Complement C4	Low (< 8 mg/dl)	10.00–40.00
Beta 2 Glycoprotein, IgG serum	Positive (21.19 SGU)	< 20.00
Beta 2 Glycoprotein, IgM serum	Negative (5.67 SGU)	< 20.00
Cardiolipin antibody, IgG	High (20.05 GPL)	< 15.00
Cardiolipin antibody, IgM	High (16.85 MPL)	< 12.50
CRP Quantitative	High (10.7 mg/dl)	< 10
Serum IgG4	Normal (0.51 g/l)	0.049–1.9851

lupus pancreatitis is not common, and it is even rarer for acute pancreatitis to be the initial indication of the disease[6]. Similarly, only a small number of papers discuss intracranial hemorrhage as the initial symptom of lupus, making it a rare occurrence. To the best of our knowledge, no published paper in the English literature has reported the simultaneous presentation of acute pancreatitis and intracerebral hemorrhage as the initial manifestation. Consequently, this case report is exceptionally uncommon and distinctive because it involves the simultaneous occurrence of two potentially severe conditions—acute pancreatitis and intracerebral bleed - as the initial symptoms, with a subsequent diagnosis of SLE.

Conclusion

The early manifestations of SLE may occasionally exhibit unusual and potentially life-threatening symptoms. However, timely diagnosis and intervention in treatment can lead to a favorable management outcome. When confronted with a clear diagnosis combined with the presence of multiple concurrent unexplained abnormalities, particularly in young women, it is incumbent upon the attending physician to diligently assess the possibility of a connective tissue disorder.

Ethical approval

NA.

Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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Conflicts of interest disclsoure

The authors declare that they have no financial conflict of interest with regard to the content of this report.

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