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Systemic Lupus Erythematosus and Anti Phospholipid Syndrome in a Male Patient: A Case Report

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Abstract

Systemic Lupus Erythematosus (SLE) and Antiphospholipid Syndrome (APS) are two distinct autoimmune disorders characterized by dysregulated immune responses, organ involvement, and the presence of autoantibodies in the bloodstream. Primary Antiphospholipid Syndrome (APS) can occur among individuals who are generally healthy and have no history of an underlying systemic autoimmune illness. In contrast, several other systemic autoimmune diseases, including SLE, have the potential to cause secondary Antiphospholipid Syndrome (APS). While Systemic Lupus Erythematosus (SLE) and Antiphospholipid Syndrome (APS) affect women of childbearing age, there are only a few reported case studies documenting the presence of SLE or APS individually in male patients. However, the coexistence of both conditions in a male patient is extremely rare, with only a few documented cases reports available to date. This case report presents a unique occurrence of concurrent SLE and APS in a forty-year-old male patient, highlighting the rarity of such a presentation. The patient presented with per rectal and mucosal bleeding and had been on warfarin therapy for 16 years due to a history of recurrent Deep Vein Thrombosis (DVT), but was not evaluated further. Immune-related laboratory results revealed positive antinuclear antibodies, anti-dsDNA antibodies, anticardiolipin antibodies, lupus anticoagulants, and direct Coombs test. Concurrent SLE and APS were diagnosed by meeting the classification criteria for both diseases. This report adds to the medical literature and emphasizes the significance of considering concurrent SLE and APS as a potential diagnosis in males. Further research is needed to enhance our understanding of the underlying mechanisms and the best approaches for managing this rare coexistence.

Keywords: Systemic lupus erythematosus; Antiphospholipid syndrome; Deep vein thrombosis

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Introduction

Systemic Lupus Erythematosus (SLE) and Antiphospholipid Syndrome (APS) are two different autoimmune disorders characterized by dysregulated immune responses, the production of autoantibodies, and chronic inflammation [1,2]. APS primarily manifests as thrombotic events or pregnancy complications, while SLE involves immune complex deposition and inflammation in multiple organs, leading to various clinical symptoms, including thrombotic events [3,4]. Individuals with SLE and/or APS may experience heart valve issues, artery disease, lung hypertension, skin changes, low platelets, anemia, and kidney-related clotting problems. Kidney problems are common in both primary and secondary APS [5,6]. APS can occur independently or alongside Systemic Lupus (SLE) or other autoimmune disorders like Rheumatoid Arthritis (RA) or Sjogren's syndrome [7,8]. In Antiphospholipid Syndrome (APS), the major pathology involves the development of antibodies attacking phospholipids in cell membranes. This causes an increase in blood clotting as well as the formation of clots inside blood vessels [9,10]. APS typically presents with a variety of clinical manifestations, affecting veins, arteries, or small vessels through vascular thrombosis. Obstetric complications, including recurrent miscarriages, pre-eclampsia, and restricted fetal growth, are often associated with APS [3,10]. Additionally, APS encompasses neurological dysfunctions, systemic and pulmonary arterial hypertension, and endocardial [10]. Although these conditions primarily impact females, predominantly women of childbearing age, there have been occasional reports of male patients experiencing SLE and APS. However, the coexistence of these conditions together in a male patient is exceptionally uncommon [11,12]. This case report aims to describe an atypical presentation of SLE and APS in a male patient, shedding light on the complexities of diagnosis and management and the need for further research in understanding these associations.

The findings presented contribute to the existing literature, highlighting the importance of vigilant monitoring, early intervention, and personalized treatment approaches to optimize

outcomes in this unique clinical presentation.

Case Presentation

A 40-year-old male patient with no positive family history, who had been undergoing warfarin therapy for 16 years due to a history of recurrent Deep Vein Thrombosis (DVT) presented with a 1-day history of multiple episodes of rectal and mucosal bleeding, including bleeding from the gums and nose. The patient also reported significant joint pain, hair loss, frothy urine, and a substantial weight loss of approximately 10 kg over the past five years. Sixteen years ago, he was diagnosed with deep venous thrombosis when he experienced swelling, pain, and warmth in the right calf. Since then, he has been taking warfarin to manage recurrent DVT. Additionally, six years ago, he developed complaints of mood swings, severe depression, hallucinations, and altered talking. He was referred to a psychiatrist and prescribed SSRIs and benzodiazepines. On physical examination, the patient had a temperature of 98°F, blood pressure of 110/70 mmHg with no significant postural drop and a heart rate of 88 beats per minute. Further examination revealed severe anemia, sunken eyes, marfanoid habitus, arachnodactyly, receding hairline, hyper pigmented rash on cheeks sparing nasolabial folds and generalized muscle wasting evident with loose triceps folds, temporal wasting, and lipodystrophy, along with significant thenar and hypothenar wasting. During CNS examination, the patient exhibited decreased bulk and increased tone in all four limbs, along with decreased power. Active and passive movements of all the joints were intact. The patient had a stumbling gait with a stooped posture and resting tremors. The rest of the systematic examination was normal. He was admitted for a comprehensive evaluation, during which various blood investigations were conducted. The blood test results indicated normocytic anemia (Hb 4.9 g/dL) with a corrected reticulocyte count of 0.8%, leukopenia (leukocyte count 2200 cells/mm^3), and thrombocytopenia (96,000 cells/mm^3). The peripheral blood film displayed anisocytosis, poikilocytosis, macrocytes, polychromasia, and occasional hypersegmented neutrophils. Further investigations revealed a PT/INR of 16.8/1.62, an APTT of 38.9 sec, and an elevated C-reactive protein at 8.5 mg/dL (normal range: 0.3 to 1.0 mg/dL), and the ferritin level was 957 ng/mL. A direct Coombs test was performed, which came out positive. The serum iron profile indicated a serum iron level of 30 mcg/dL, TIBC of 550 mcg/dL, ferritin of 957 ng/mL, and transferrin saturation of 5.5%. Immunological investigations demonstrated positive findings, including an Antinuclear Antibody (ANA) titer of 1/5120, positive anti-dsDNA antibody at a level of 182.0 (>20), positive serum anticardiolipin IgG of 23.63 (>14.4), and lupus anticoagulant of 103.4 sec (31-44) with an LA ratio of 2.9 (0.8-1.1). Serum C3 levels were low, while C4 levels, as well as thyroid, liver, and renal functions, were within acceptable limits. Ultrasound abdomen and echocardiography results were normal. Additional tests for rheumatoid factor, CCP antibodies, serum Beta 2 glycoprotein, U1-RNP antibodies, SSA/Ro antibodies, SS-B/La antibodies, anti-smooth muscle antibodies, anti-mitochondrial antibodies, and Scl-70 antibodies all yielded negative results, as did viral serology. Urinalysis showed trace protein, and a spot urinary PCR was elevated 1.92. Doppler ultrasound of the lower limb revealed partial echogenic material in the right common femoral vein and popliteal vein, while the right saphenous vein on the right side was patent, as were all the veins on the left side. The chronicity of symptoms and notable diagnostic parameters led to a diagnosis of Systemic Lupus Erythematosus (SLE) and Antiphospholipid Syndrome (APS) as per the ACR-EULAR criteria and SLICC criteria. To assess the degree of kidney involvement, a renal biopsy was performed. The examined sections of the biopsy consisted mostly of cortex with a small amount of medulla. Approximately 10 glomeruli were included in the sample. Among these, one glomerulus showed ischemic solidification, while the rest exhibited minor changes. There was no thickening of the Glomerular Basement Membrane (GBM), no wire loop lesions, no segmental scarring, and no vasculopathy. A minimal focal tubular atrophy was observed. Immunofluorescence Microscopy (IMF) was conducted on fresh tissue and the results showed a completely negative panel, indicating the absence of immune complex deposits in the kidney tissue. Upon admission, the patient was started on pulse therapy with intravenous methylprednisolone 1gm once daily for five days. Following the completion of the pulse therapy, the patient was transitioned to an equivalent dose of intravenous dexamethasone. To address the neutropenic sepsis, the patient received intravenous Piperacillin/tazobactam 4.5 gm three times daily. Additionally, intravenous omeprazole 40 mg once daily as a proton pump inhibitor was given to reduce risk of peptic ulcer disease. The patient received oral medications including hydroxychloroquine 200 mg twice daily, folic acid 1 tablet twice daily, and ramipril 2.5 mg once daily. Hydroxychloroquine was prescribed for its immunomodulatory properties, while folic acid served as a supplementation strategy and ramipril, an Angiotensin-Converting Enzyme (ACE) inhibitor to improve proteinuria. As the patient's coagulation profile improved and bleeding symptoms resolved, the anticoagulant medication was initiated as subcutaneous injections of Clexane 40 mg twice daily. The patient responded positively to the given treatment. He was discharged on tab Deltacortril, tab hydroxychloroquine, tab folic acid, tab ramipril, tab mycophenolate mofetil and tab Rivaroxaban instead of warfarin due to its low risk of bleeding. One week following discharge, the patient presented with complaints of shortness of breath, dizziness, and lightheadedness for one day. Physical examination revealed tachycardia with a heart rate of 140 bpm, tachypnea with a respiratory rate of 35 bpm, and peripheral oxygen saturation (SpO_2) of 88% on room air. The rest of the physical examination was unremarkable. Given the severity of the patient's condition, urgent laboratory investigations were performed. Arterial Blood Gas (ABG) analysis demonstrated hypoxemia with hypocapnia, leading to respiratory alkalosis. Due to the deteriorating clinical status, the patient's intubation and mechanical ventilation were planned. However, tragically, the patient expired within an hour of hospital admission.

Discussion

Systemic Lupus Erythematosus (SLE) and Antiphospholipid Syndrome (APS) are autoimmune disorders characterized by a wide range of clinical presentations, yet their underlying cause remains incompletely understood. These conditions are serologically characterized by the presence of autoantibodies targeting specific proteins within the body [10,11]. Notably, antiphospholipid antibodies (aPL), including Lupus Anticoagulant (LA) or anticardiolipin antibodies (aCL), have been reported in 30% to 50% of SLE patients [13]. SLE itself serves as an independent risk factor for arterial and venous thrombotic events, and the presence of antiphospholipid antibodies (aPL) further increases the overall thrombotic risk [14]. Additionally, persistent inflammation triggers the upregulation of tissue factors, activating the procoagulant pathway while impeding anticoagulation and fibrinolysis [15]. In the presented case, the initial identifiable symptom of SLE was the occurrence of deep vein thrombosis without an apparent cause. However, SLE can also manifest in various other forms of thromboembolic events in different

locations, such as cerebral venous thrombosis and pulmonary embolism, followed by subsequent deep vein [16]. It is important to note that hypercoagulability is often an initial symptom, although in some cases, it may be preceded by other symptoms such as fever, weight loss, general malaise, body swelling and generalized joint pain [3]. The purpose of this case study is to provide light on an unusual occurrence of Antiphospholipid Syndrome (APS) and Systemic Lupus Erythematosus (SLE) in a male patient. This case study provides new information for the medical literature and emphasizes the importance of considering Systemic Lupus Erythematosus (SLE) and Antiphospholipid Syndrome (APS) as potential diagnoses in males. In this case, the patient was treated with rivaroxaban instead of warfarin due to the presence of bleeding upon admission and as a safety measure [17]. In general, DOACs are safer and more effective than warfarin, but in patients with antiphospholipid syndrome, warfarin remains the best option to prevent VTE recurrence [18,19]. Hence, in cases where patients decline warfarin treatment, have a low tolerance to warfarin, or experience inadequate anticoagulant control despite adherence, the use of apixaban can be taken into consideration [20]. This highlights the need for more research to improve our understanding of the underlying mechanisms and find the best methods for managing this unusual coexistence. While a clear tendency towards females is evident, we emphasize the importance of considering the possibility of Systemic Lupus Erythematosus (SLE) and Antiphospholipid Syndrome (APS) in males as well, given the severe trajectory of the clinical presentation.

Conclusion

We present a rare case of a forty-year-old male patient who had both Systemic Lupus Erythematosus (SLE) and Antiphospholipid Syndrome (APS) concurrently. This case highlights the rarity of such a presentation, particularly in male patients. It is important to remember that even in males, the diagnosis of concurrent SLE and APS should be considered. The diagnosis was confirmed on the patient's clinical symptoms and meeting the classification criteria for both disorders. A comprehensive management approach was employed, and this case contributes to the existing literature by emphasizing the need for further research to improve our understanding of the underlying mechanisms and treatment. By presenting this case, we aim to inform healthcare professionals about the possibility of SLE and APS occurring in male patients and how to properly manage Antiphospholipid Syndrome (APS).

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