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Case Report

# Evans syndrome in conjunction with rheumatoid arthritis and SLE: A unique case of autoimmune intersection ☆,☆☆

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ABSTRACT

Evans syndrome is a rare autoimmune disorder characterized by autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP), often linked with systemic lupus erythematosus (SLE). We present a case of a 25-year-old female with a history of rheumatoid arthritis (RA) who presented with new SLE symptoms, including left-sided weakness, pallor, and a photosensitive rash. Laboratory tests confirmed Evans syndrome, and MRI showed a cerebral infarction. Treatment with corticosteroids, hydroxychloroquine, and mycophenolate mofetil led to significant improvement. This case highlights the complexity of managing Evans syndrome in patients with coexisting autoimmune diseases like RA and SLE, emphasizing the need for early and aggressive treatment.



## Introduction

Evans syndrome, an autoimmune disorder marked by concurrent autoimmune hemolytic anemia and immune thrombocytopenia, is frequently associated with systemic lupus erythematosus (SLE) [1]. Evans syndrome was first identified by Evan and Duane in 1949 [2]. It is a condition with unknown etiology that results from an alteration of the immune system that produces multiple autoantibodies targeting red blood

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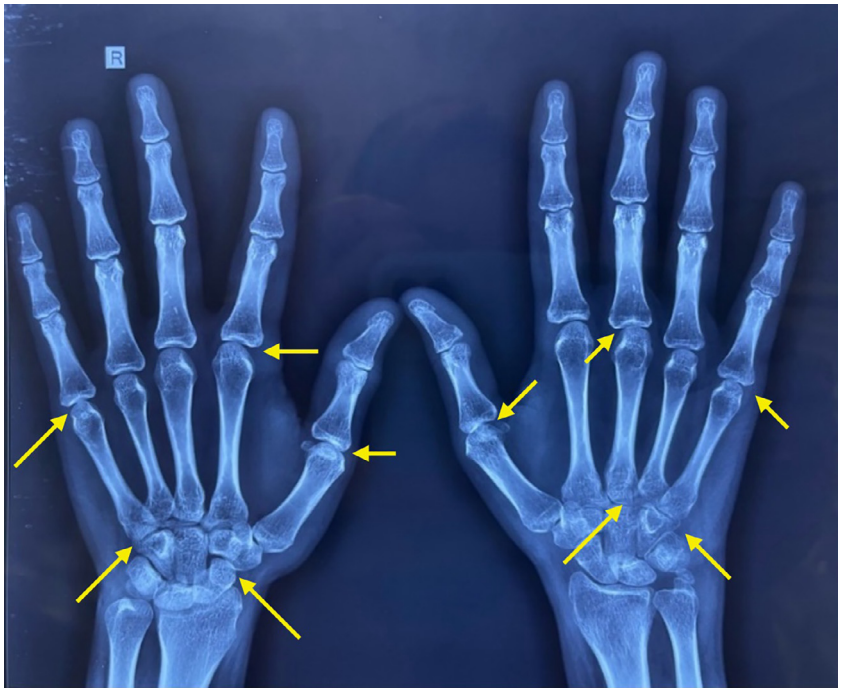


**Fig. 1 – Anteroposterior X-ray of the hands, showing osteopenia and joint space narrowing in the second, third and fifth MCP joints bilaterally, also in the carpometacarpal bilaterally and intercarpal.**

cells and platelets [3]. The great majority of patients with ES have a chronic relapsing course despite treatment, which is associated with significant morbidity and mortality [4].

This case report discusses a 25-year-old female with a history of rheumatoid arthritis (RA) who presented with new-onset symptoms suggestive of SLE, including left-sided weakness, severe pallor, and a photosensitive rash. Laboratory findings confirmed Evans syndrome, with severe anemia, thrombocytopenia, and a positive direct Coombs test. MRI revealed an infarction in the region supplied by the right middle cerebral artery. The overlap of RA and SLE, along with the development of Evans syndrome, posed significant diagnostic and therapeutic challenges [5,6]. Treatment with intravenous methylprednisolone, followed by prednisone, hydroxychloroquine, and mycophenolate mofetil, led to significant clinical improvement. This case highlights the complexity of managing Evans syndrome in patients with pre-existing autoimmune diseases like RA and underscores the importance of early, aggressive treatment in such scenarios.

## Case presentation

A 25-year-old female presented with a 2-day history of left-sided weakness, more pronounced in the upper limb, accompanied by slurred speech, headache, and severe pallor. She also reported a recent onset of fatigue, joint pain, and a photosensitive rash over her cheeks.

Her medical history includes rheumatoid arthritis (RA), diagnosed 2 years prior. This diagnosis was confirmed through clinical symptoms, serological tests, and imaging studies, which revealed positive anti-citrullinated protein antibodies (ACPA), elevated rheumatoid factor (RF), and radiographic imaging of her hands at that time, showing periarticular osteopenia and joint space narrowing in the second, third and fifth MCP joints bilaterally. Also, in the carpometacarpal bilaterally and intercapals, characteristic of RA as shown in (Fig. 1). Her condition had been well-controlled on methotrexate, with stable disease until recently, when she developed new symptoms, raising concerns about a potential overlap syndrome or the emergence of systemic lupus erythematosus (SLE) in the context of her established RA.

Two months before presentation, the patient developed facial skin lesions that were red, painful, and swollen, eventually progressing to purulent discharge, minor bleeding, and scarring, as shown in (Fig. 2). She also experienced painless oral ulcers, hair loss, Raynaud's phenomenon, headaches, arthritis, fatigue, and subjective fevers during this period.

On examination, the patient was conscious and oriented but appeared severely pale and had oral ulcers, which are common in SLE. The neurological assessment revealed left-sided weakness with power graded at 3/6 in the upper limb and 5/6 in the lower limb, along with a left extensor plantar response. Sensory examination was unremarkable, with intact cranial nerves and no signs of meningeal irritation. Given the neurological symptoms, an MRI was promptly conducted, revealing an infarction in the right middle cerebral artery ter-

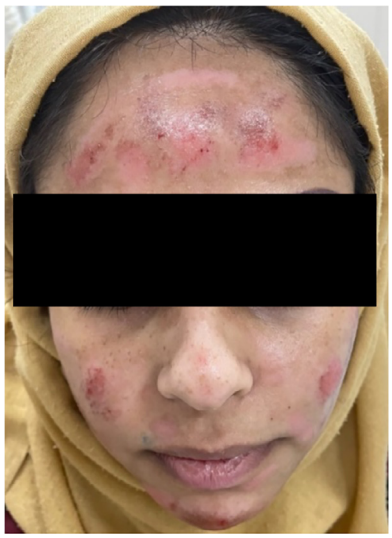

**Fig. 2 – A photograph of the face reveals photo distributed, scaly erythematous plaques with hypopigmented atrophic scarring.**

ritory, as shown in (Fig. 3), which explained her neurological deficits.

Given her severe pallor, laboratory investigations were conducted, revealing severe anemia with a hemoglobin level of 4.2 g/dL, a mean corpuscular volume (MCV) of 110 fL, and a platelet count of 37 × 10^9/L. A positive direct Coombs test confirmed autoimmune hemolytic anemia, while tests for paroxysmal nocturnal hemoglobinuria (PNH) were negative.

In this case, the diagnosis of Evans syndrome was made through exclusion, as is typical for this condition. Blood tests indicated mild anemia, reticulocytosis, and thrombocytopenia, with a peripheral blood smear showing polychromatic erythrocytes. Elevated LDH levels pointed to hemolysis. The positive Coombs test and antibody screening suggested warm-type autoimmune hemolytic anemia (AIHA). The simultaneous occurrence of AIHA and immune thrombocytopenia (ITP), both without an identifiable cause, led to the confirmation of Evans syndrome.

Treatment was initiated with pulse intravenous methylprednisolone for 3 days, followed by a maintenance regimen of oral prednisone, hydroxychloroquine, and mycophenolate mofetil. Within the first week of treatment, the patient exhibited significant improvement in both hematological parame-

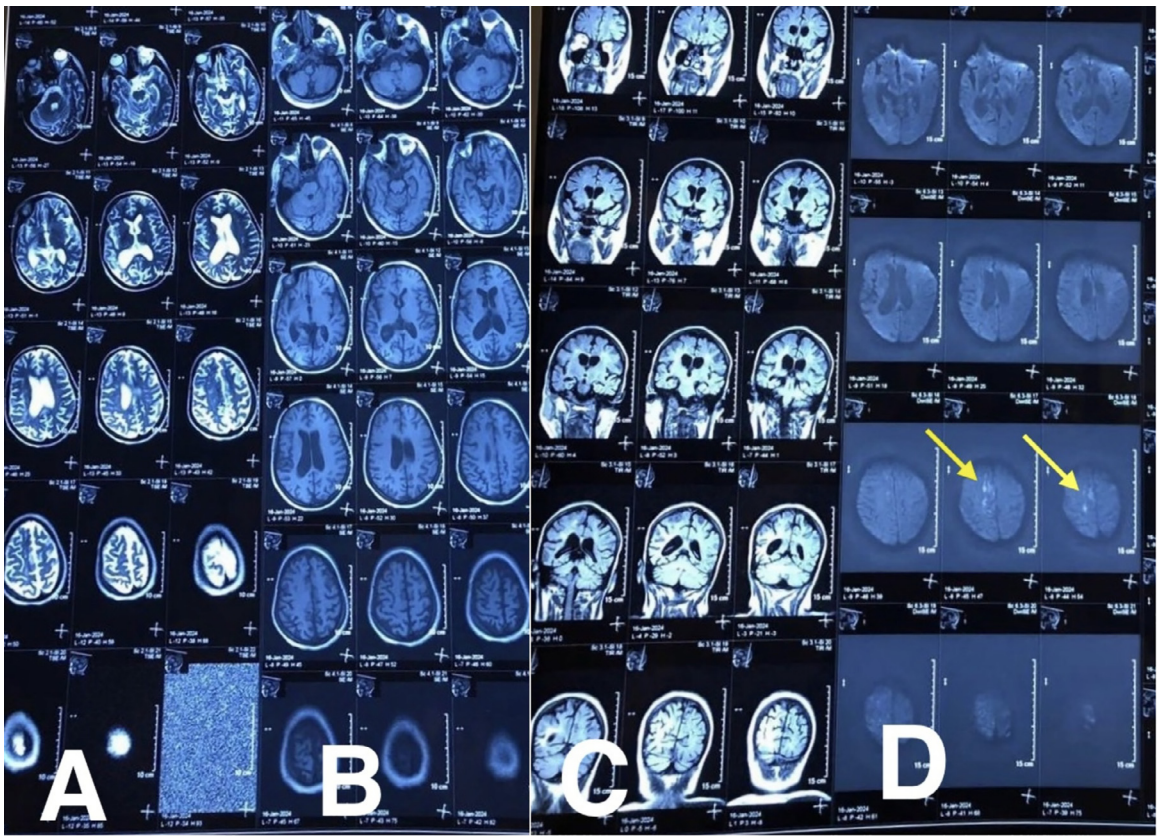


**Fig. 3 – Brain MRI demonstrating multiple bands of abnormal signal intensity predominantly involving the cortical surface of the right fronto-parietal and occipital lobes. The lesions appear hyperintense on T2-weighted (A) and FLAIR sequences (C), hypointense on T1-weighted images (B), and hyperintense on DWI (D), consistent with hyperacute infarction within the territory of the right middle cerebral artery.**

ters and systemic symptoms, underscoring the importance of early and aggressive immunosuppressive therapy in managing Evans syndrome associated with SLE, particularly in a patient with concurrent RA.

## Discussion

Evans syndrome is a rare autoimmune disorder characterized by the simultaneous or sequential occurrence of autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP). It often presents in association with other autoimmune conditions, such as systemic lupus erythematosus (SLE). The syndrome is challenging to manage due to its chronic and relapsing nature, requiring a multidisciplinary approach involving corticosteroids, immunosuppressants, and close monitoring. [7]

The patient's presentation with neurological symptoms, including left-sided weakness and slurred speech, is atypical for Evans syndrome but can occur in the context of SLE-related neuropsychiatric manifestations. The presence of severe anemia and thrombocytopenia, coupled with a positive direct Coombs test, confirmed the diagnosis of Evans syndrome [8]. Treatment primarily involved immunosuppressive therapy, which included corticosteroids, hydroxychloroquine, and mycophenolate mofetil, leading to a marked improvement in clinical and laboratory findings [9].

Patients with Evans syndrome (ES) in the context of systemic lupus erythematosus (SLE) often present with other autoimmune diseases. In our case, the patient has rheumatoid arthritis (RA), which is also an autoimmune disease [10]. This aligns with research showing a tendency for additional autoimmune conditions in such patients. However, another study found that most of the associations are with conditions like antiphospholipid syndrome, Sjögren's syndrome, systemic sclerosis, and Takayasu's arteritis. The presence of RA in our case, therefore, highlights the variability of autoimmune associations in ES and underscores the uniqueness of this case [11].

A related case report, "Evans Syndrome in the Context of Incomplete Systemic Lupus Erythematosus" (Abu Kar et al.), describes a similar case where Evans syndrome was associated with incomplete SLE, highlighting the diagnostic challenges in such cases. The patient, like ours, required immunosuppressive therapy to achieve remission. This case emphasizes the complexity of Evans syndrome when multiple autoimmune conditions coexist and underscores the importance of aggressive, individualized treatment. Including this report provides valuable context for managing the unpredictable nature of Evans syndrome in such complex clinical scenarios [12].

Compared to similar cases in the literature, this case's unique aspects include the concurrent occurrence of rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), and Evans syndrome (ES), along with cerebral infarction. These features underscore the critical need for early recognition. Given the rarity of ES, particularly in the context of both SLE and RA, a high index of suspicion is essential, along with a carefully tailored therapeutic approach to manage the complex interplay of autoimmune processes.

## Conclusion

This case highlights the critical importance of considering Evans syndrome in patients presenting with severe anemia, thrombocytopenia, and symptoms suggestive of an autoimmune disorder. The association with SLE further complicates the clinical picture, requiring a multidisciplinary approach to diagnosis and management. Early intervention with appropriate immunosuppressive therapy can significantly improve patient outcomes, as demonstrated in this case. The unusual presentation underscores the need for awareness and thorough investigation in similar clinical scenarios.

## Ethics approval

Our institution does not require ethical approval for reporting individual cases or case series.

## Patient consent

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

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