Section: General Medicine

Case Report

EVANS SYNDROME AS THE PRESENTING MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS IN AN ADOLESCENT FEMALE: A CASE REPORT

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ABSTRACT

Evans syndrome (ES) represents a rare autoimmune condition involving concomitant autoimmune hemolytic anemia (AIHA) and immune thrombocytopenia (ITP), sometimes with neutropenia. Its emergence as the inaugural sign of systemic lupus erythematosus (SLE) is exceptional and may obscure timely diagnosis. Here, we detail the case of an 18-year-old woman who sought medical attention for persistent fever, a distinctive rash, and symmetric joint discomfort. Diagnostic evaluation disclosed pancytopenia, a positive direct antiglobulin test (DAT), antinuclear antibody (ANA) positivity, elevated anti-double-stranded DNA (anti-dsDNA) and anti-Smith D1 (anti-SmD1) antibodies, along with subtle proteinuria. These findings aligned with ES attributable to SLE. The case underscores the necessity of vigilant screening for underlying connective tissue disorders in young patients exhibiting multifaceted cytopenia and systemic symptoms to facilitate early therapeutic intervention.

Keywords: Evans syndrome (ES), Autoimmune hemolytic anemia (AIHA), Immune thrombocytopenia (ITP), Pancytopenia, Systemic lupus erythematosus (SLE), Secondary Evans syndrome, Menorrhagia, Fatigue, weakness, Pallor, Splenomegaly, Leukopenia, Thrombocytopenia, Positive Direct Antiglobulin Test (DAT), Spherocytes (PBS), Elevated ESR, ANA positive, Anti-dsDNA, Anti-SmD1 (SLE-specific).

INTRODUCTION

First delineated in 1951, Evans syndrome constitutes a chronic autoimmune disorder marked by the synchronous or successive development of AIHA and ITP, potentially compounded by immune-mediated neutropenia. Affecting fewer than 5% of individuals with AIHA, ES predominantly arises in the context of secondary triggers, including autoimmune rheumatic diseases like SLE, lymphoproliferative malignancies, or infectious processes. In paediatric and young adult cohorts, primary ES is infrequent, with up to 50% of cases linked to SLE, where it often signals heightened disease activity. When ES precedes overt SLE features, diagnostic delays can ensue, exacerbating morbidity through unchecked haemolysis, thrombocytopenic complications, or extramedullary hematopoiesis. This report illustrates such a scenario, emphasizing the pivotal role of comprehensive serological assessment in unraveling the etiology of refractory cytopenia.

Case Presentation

An 18-year-old female, previously in good health, presented with complaints of progressively heavy menstrual bleeding for the last 3 months. Her cycles occurred every 28–30 days and were characterized by excessive flow lasting 8–9 days, associated with passage of large clots. She required frequent pad changes (7–8 per day), which was a significant change compared to her baseline pattern of 4–5 days

of moderate bleeding. The excessive bleeding was accompanied by worsening fatigue, generalized weakness, and occasional dizziness, particularly on standing. She also reported difficulty concentrating on her daily academic activities. There was no history of intermenstrual spotting, post-coital bleeding, abdominal pain, weight loss, or easy bruising. She denied gum bleeding, recurrent epistaxis, hematemesis, or melena. There was no prior history of blood transfusions or use of anticoagulant medications. Her past medical and surgical history was unremarkable. There was no family history of bleeding disorders or hematologic malignancies.

On evaluation, she exhibited tachycardia (pulse 110 beats/min), BP (100/60 mmHg), afebrile, and tachypnoea (18 breaths/min), with preserved oxygenation (SpO_2 99% on ambient air). Physical survey revealed moderate pallor without jaundice, edema, or peripheral adenopathy. Neurological, cardiovascular, and respiratory assessments were unremarkable, though abdominal palpation confirmed moderate splenomegaly without hepatomegaly or ascites.

Investigations

Baseline laboratory investigations revealed severe anaemia. A urine pregnancy test (UPT) was performed and was negative. Ultrasonography (USG) of the pelvis and kidneys-ureters-bladder (KUB) showed no significant abnormality. Additional hematological, biochemical, and nutritional work-up is summarized in the tables below.

Table 1: Hematological and Inflammatory Markers

Parameter	Result	Reference Range/Interpretation
Hemoglobin (Hb)	5.3 g/dL	Severe anemia (<11 g/dL)
Total Leukocyte Count (TLC)	2.55–3.1 × 10^3/µL	Leukopenia (<4 × 10^3/µL)
Red Blood Cell Count (RBC)	2.02 × 10^6/µL	Decreased
Packed Cell Volume (PCV)	18.1%	Decreased
Platelet Count	84 × 10^3/µL	Thrombocytopenia (<150 × 10^3/µL)
Erythrocyte Sedimentation Rate (ESR)	86 mm/h	Elevated (>20 mm/h)
Reticulocyte Count	1%	Subdued response
Peripheral Blood Smear	Spherocytes present	Consistent with AIHA
Direct Antiglobulin Test (DAT)	Positive	Confirms AIHA

Table 2: Iron Studies and Nutritional Markers

Parameter	Result	Reference Range/Interpretation
Serum Iron	34 µg/dL	Low (<50 µg/dL)
Total Iron-Binding Capacity (TIBC)	241 µg/dL	Low (250–450 µg/dL)
Ferritin	659 ng/mL	High (>300 ng/mL), inflammatory
Vitamin D	15 ng/mL	Deficient (<20 ng/mL)
Vitamin B12	Normal	Within range

Table 3: Liver Function Tests (LFT)

Parameter	Result	Reference Range/Interpretation
Total Bilirubin	0.59 mg/dL	Mildly elevated (0.1–1.0 mg/dL)
Albumin	3.3 g/dL	Low (3.5–5.0 g/dL)
Albumin/Globulin (A/G) Ratio	0.9	Decreased (<1.0)

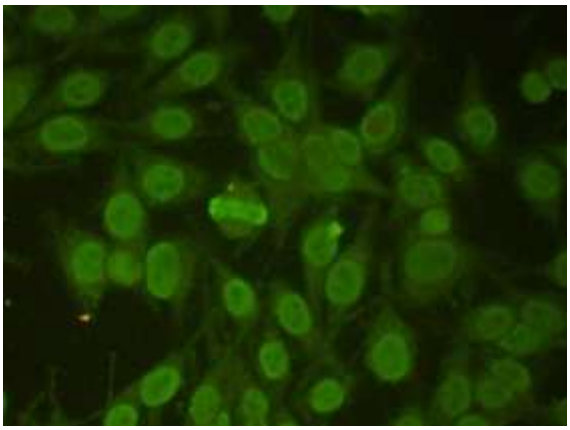
Table 4: Renal Function and Urinalysis

Parameter	Result	Reference Range/Interpretation
Renal Function Tests (RFT)	Within normal limits	Normative
Urinalysis	Trace proteinuria	Abnormal
24-Hour Urine Protein	149 mg	Mild proteinuria (<150 mg/24h borderline)

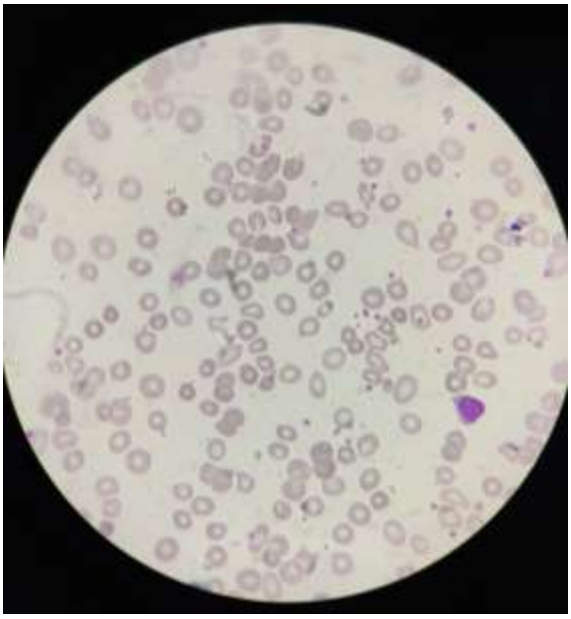
Table 5: Serological and Autoimmune Profile

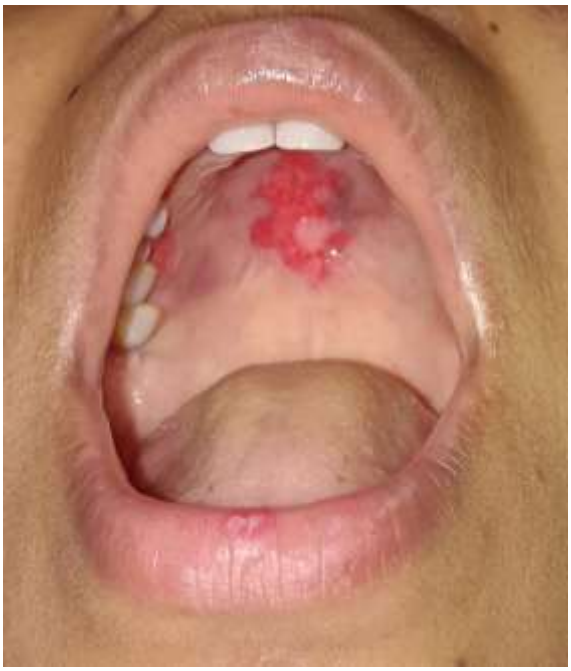
Parameter	Result	Reference Range/Interpretation
Viral Serology (HBsAg, HCV, HIV)	Negative	Nonreactive
Rheumatoid Factor (RF)	8.6 IU/mL	Negative (<12 IU/mL)
Antinuclear Antibody (ANA) by IFA	Positive, titer 1:80, homogeneous pattern	Positive (>1:40)
Anti-dsDNA	Weakly positive (2.22)	Abnormal
Anti-Nucleosome	+++ (4.09)	Positive
Anti-Histone	+++ (4.74)	Positive
Anti-SmD1	+++ (6.96)	Positive (SLE-specific)
Anti-U1-snRNP	+++ (5.52)	Positive
Other Extractable Nuclear Antigens (ENA)	Negative	Absent
Complement C3/C4	Decreased	Hypocomplementemia

These findings collectively indicated pancytopenia, hemolytic anemia, splenomegaly, positive autoantibodies, and mild renal involvement, consistent with ES secondary to SLE per the 2019 EULAR/ACR criteria.



Name of Antigen	Results	Index Value
ds DNA	Weakly positive (+)	2.22
Nucleosome	Strong Positive (+++)	4.09
Histones	Strong Positive(+++)	4.74
SmD1	Strong Positive (+++)	6.96
PCNA	Negative (-)	0.26
P0	Negative (-)	0.43
SS-A/ Ro60KD	Negative (-)	0.22
SS-A/ Ro52KD	Negative (-)	0.17
SS-B/La	Negative (-)	0.57
CENP - B	Negative (-)	0.17
Scl70	Negative (-)	0.22
U1-snRNP	Strong Positive (+++)	5.52
AMA M2	Negative (-)	0.57
Jo-1	Negative (-)	0.30
PM-Scl	Negative (-)	0.17
Mi-2	Negative (-)	0.17
Ku	Negative (-)	0.65
DFS 70	Borderline (0)	1.09





Management

Therapy commenced with high-dose intravenous methylprednisolone (1 g daily for 3 days), seamlessly tapering to oral prednisolone (1 mg/kg/day). Hydroxychloroquine (200 mg twice daily) was introduced for SLE stewardship. Supportive measures encompassed two units of packed red blood cells for symptomatic anemia. Multidisciplinary input from hematology and rheumatology guided care. Within 2 weeks, she achieved hematologic remission (hemoglobin 9.8 g/dL; platelets 180 × 10^3/μL; leukocytes 5.2 × 10^3/μL), rash resolution, and arthralgia abatement. She was discharged with tapered glucocorticoids, hydroxychloroquine, and scheduled surveillance for renal and serologic progression.

DISCUSSION

ES embodies a formidable autoimmune diathesis, with pediatric incidence approximating 1:1,000,000 and a propensity for SLE comorbidity in 20–40% of instances. Hematologic derangements in SLE, encompassing anemia (50–70%), leukopenia (20–40%), and thrombocytopenia (15–30%), stem from multifactorial pathways: autoantibody opsonization, splenic sequestration, and cytokine-driven apoptosis. In this adolescent, ES heralded SLE, mirroring sporadic reports where cytopenias antedate canonical lupus stigmata by months, confounding initial appraisals toward infections, malignancies, or idiopathic aplasia.

Discriminative challenges include macrophage activation syndrome or paraneoplastic cytopenias, yet the absence of hyperferritinemia extremes, viral titers, and marrow dysplasia, coupled with lupus-

specific autoantibodies, steered toward SLE. Anti-SmD1 and anti-dsDNA positivities confer diagnostic specificity (>95%) and prognostic gravity for nephritis, respectively, while anti-nucleosomal and anti-histone reactivities intimate chromatin epitope exposure in nascent autoimmunity. The modest proteinuria portended class II/III lupus nephritis, warranting serial eGFR and proteinuria tracking. Negative rheumatoid factor obviated juvenile idiopathic arthritis mimicry.

Corticosteroids remain cornerstone for ES-SLE, yielding 60–80% initial responses, augmented by antimalarials for cutaneous/joint control and relapse prophylaxis. Refractory subsets may necessitate rituximab or mycophenolate, with 30% relapse risk underscoring lifelong vigilance. This vignette advocates proactive autoantibody interrogation in cytopenic youths with enigmatic systemic complaints to expedite SLE unmasking.

CONCLUSION

ES as SLE's sentinel event in adolescence is an uncommon yet instructive paradigm, demanding astute hematologic-rheumatologic synergy. Expeditious autoantibody delineation, proteinuria quantification, and rheumatoid exclusion are imperative for averting sequelae. Heightened clinician awareness may curtail diagnostic lags, optimizing outcomes in this vulnerable demographic.

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