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From Cytopenia to Clot: Evans Syndrome Presenting as Acute Portal Vein Thrombosis with Rapid-Onset Ascites

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ABSTRACT

Evans syndrome is a rare autoimmune condition characterized by the coexistence of autoimmune hemolytic anemia and immune thrombocytopenia, with a paradoxical predisposition to thrombosis. We report a 51-year-old male presenting with rapidly progressive ascites and bilateral pedal edema, subsequently diagnosed with acute portal vein thrombosis leading to non-cirrhotic portal hypertension. Laboratory evaluation demonstrated hemolysis and thrombocytopenia, confirming Evans syndrome after exclusion of secondary causes. Despite thrombocytopenia, prothrombotic mechanisms including endothelial dysfunction, platelet activation, and hemolysis-associated hypercoagulability likely contributed to thrombosis. The patient was managed with supportive therapy and planned anticoagulation following platelet stabilization. This case underscores the importance of recognizing thrombotic complications in autoimmune cytopenias and highlights the need for early diagnosis and individualized management to optimize outcomes.

INTRODUCTION

Evans syndrome is an uncommon autoimmune disorder defined by the coexistence or sequential development of Autoimmune Hemolytic Anemia (AIHA) and Immune Thrombocytopenia (ITP), with or without immune neutropenia [1,2]. The estimated prevalence is less than 1 per 1,000,000, and the condition is characterized by a chronic, relapsing course with substantial clinical heterogeneity [1]. Evans syndrome may occur as a primary entity or secondary to Systemic Lupus Erythematosus (SLE), Antiphospholipid Syndrome (APS), lymphoproliferative disorders, or primary immunodeficiencies [2,3]. Although cytopenias remain the hallmark,

accumulating evidence reveals a paradoxical prothrombotic milieu in Evans syndrome. This arises from several interrelated mechanisms: platelet activation despite reduced counts, release of procoagulant microparticles during hemolysis, free hemoglobin–mediated nitric oxide scavenging leading to vasoconstriction and endothelial dysfunction, and chronic inflammation promoting a hypercoagulable state [4,5]. The incidence of thromboembolism in immune thrombocytopenia alone has been reported at 1.5 times to 2.5 times that of the general population, and concurrent AIHA may amplify this risk further [6,7].

Portal Vein Thrombosis (PVT) most commonly occurs in the context of hepatic cirrhosis, intra-abdominal malignancy, myeloproliferative neoplasms, or inherited thrombophilia's [8]. Non-cirrhotic PVT, however, is increasingly recognized in systemic inflammatory and autoimmune conditions [9]. The occurrence of acute PVT in Evans syndrome is exceedingly rare, with only isolated reports in the literature, and poses unique diagnostic and therapeutic challenges particularly when rapid-onset portal hypertension and ascites mimic decompensated cirrhosis [10].

We present a case of Evans syndrome manifesting as acute extrahepatic portal vein thrombosis with rapid-onset non-cirrhotic portal hypertension, emphasizing the need for heightened clinical suspicion of thrombotic complications in autoimmune cytopenias.

CASE DESCRIPTION

A 51-year-old male presented with rapidly progressive bilateral lower limb swelling of 4 days' duration and abdominal distension for 2 days. The swelling initially involved the feet and, over a short period, progressed proximally to involve both legs up to the knees. It was pitting in nature and associated with a sensation of tightness and discomfort, without local warmth or erythema. Within 48 hours of onset of lower limb edema, the patient developed abdominal distension, which increased rapidly, resulting in a feeling of fullness and mild diffuse abdominal discomfort. There was no history of hematemesis, melena, fever, or jaundice. The patient also reported reduced appetite and disturbed sleep.

On examination, the patient was conscious, oriented, and hemodynamically stable. Bilateral pitting pedal edema up to the knees was noted. Abdominal examination revealed tense distension with positive shifting dullness, consistent with ascites. There was no abdominal tenderness. Cardiovascular and respiratory system examinations were unremarkable. Neurological examination revealed features suggestive of early hepatic encephalopathy (Grade I), including mild irritability and altered sleep wake cycle. Laboratory investigations (Table 1) revealed anemia and thrombocytopenia. Peripheral smear findings, along with further hematological evaluation, were suggestive of autoimmune hemolytic anemia with immune thrombocytopenia, consistent with Evans syndrome. Liver function tests were largely preserved except for mild hypoalbuminemia. Renal parameters were within normal limits, and glycated hemoglobin (HbA1c) was 5.4%. Ultrasound abdomen with Doppler demonstrated moderate ascites, splenomegaly, and features of portal hypertension. There was evidence of thrombosis within the portal vein with markedly reduced flow, suggestive of acute portal vein thrombosis. The hepatic parenchyma appeared preserved on imaging, with no definitive radiological features of cirrhosis. Upper gastrointestinal endoscopy revealed Grade I–II esophageal varices.

Diagnostic paracentesis yielded clear ascitic fluid. Analysis was consistent with portal hypertensive ascites, with no evidence of infection (Table 2). Further evaluation revealed features of hemolysis, including elevated lactate dehydrogenase (LDH: 860 U/L), indirect hyperbilirubinemia (indirect bilirubin: 2.8 mg/dL), and reticulocytosis

(reticulocyte count: 5.6%). Direct antiglobulin (Coombs) test was positive, confirming autoimmune hemolytic anemia. In the presence of concurrent thrombocytopenia and after exclusion of secondary causes, a diagnosis of Evans syndrome was established with acute extrahepatic portal vein thrombosis leading to rapidly progressive non-cirrhotic portal hypertension and ascites. The patient was initiated on supportive management, including albumin infusion, with further evaluation planned for underlying autoimmune and hypercoagulable states. Careful consideration for anticoagulation was undertaken in view of the coexisting thrombocytopenia.

Table 1: Summary of laboratory investigations.

Parameter	Result	Reference Range
Hemoglobin	7.8 g/dL	13.0–17.0 g/dL
Mean Corpuscular Volume (MCV)	102 fL	80–100 fL
Reticulocyte count	5.60%	0.5–2.5%
Platelet count	38 × 10^9/L	150–400 × 10^9/L
White blood cell count	6.2 × 10^9/L	4.0–11.0 × 10^9/L
Total bilirubin	3.6 mg/dL	0.1–1.2 mg/dL
Indirect bilirubin	2.8 mg/dL	0.1–0.8 mg/dL
Lactate Dehydrogenase (LDH)	860 U/L	140–280 U/L
Haptoglobin	<10 mg/dL	30–200 mg/dL
Direct Antiglobulin Test (DAT)	Positive (IgG + C3d)	Negative
Serum albumin	2.9 g/dL	3.5–5.0 g/dL
Aspartate Aminotransferase (AST)	42 U/L	10–40 U/L
Alanine Aminotransferase (ALT)	38 U/L	7–56 U/L
Alkaline Phosphatase (ALP)	98 U/L	44–147 U/L
International Normalized Ratio (INR)	1.1	0.8–1.2
Serum creatinine	0.9 mg/dL	0.7–1.3 mg/dL
Blood Urea Nitrogen (BUN)	14 mg/dL	7–20 mg/dL
HbA1c	5.40%	<5.7%
Antinuclear Antibody (ANA)	Negative	Negative
Anti-double-stranded DNA	Negative	Negative
Lupus anticoagulant	Negative	Negative
Anticardiolipin antibodies (IgG/IgM)	Negative	Negative
Anti-β2-glycoprotein I (IgG/IgM)	Negative	Negative
JAK2 V617F mutation	Not detected	Not detected
Hepatitis B surface antigen	Negative	Negative
Anti-hepatitis C antibody	Negative	Negative
HIV 1/2 antibody	Negative	Negative

Table 2: Ascitic fluid analysis.

Parameter	Result	Interpretation
Appearance	Clear, straw-colored	—
Total protein	1.8 g/dL	Transudative
Serum-Ascites Albumin Gradient (SAAG)	1.5 g/dL	≥ 1.1 g/dL indicates portal hypertension
Cell count	120 cells/mm^3 (lymphocyte predominant)	No spontaneous bacterial peritonitis
Polymorphonuclear leukocytes	18 cells/mm^3	<250 cells/mm^3
Gram stain and culture	Negative	No infection
Cytology	Negative for malignant cells	—
Adenosine Deaminase (ADA)	8 U/L	<40 U/L (tuberculosis unlikely)

DISCUSSION

This case highlights important clinical principles related to Evans syndrome, particularly its paradoxical association with thrombosis. Although thrombocytopenia would be expected to reduce thrombotic risk, evidence shows that immune-mediated platelet destruction can instead create a prothrombotic state. Several mechanisms contribute to this phenomenon. Platelet destruction leads to the release of phosphatidylserine-rich microparticles, which provide a catalytic surface for thrombin generation and accelerate coagulation [4,5]. Concurrently, hemolysis in Autoimmune Hemolytic Anemia (AIHA) promotes hypercoagulability through multiple pathways. Free hemoglobin scavenges nitric oxide, resulting in vasoconstriction and endothelial activation, while byproducts such as arginase and adenosine diphosphate enhance platelet aggregation [6,7]. In addition, chronic inflammation and circulating autoantibodies contribute to endothelial dysfunction by upregulating adhesion molecules and tissue factor expression [5]. Splenomegaly, commonly observed in Evans syndrome, further contributes by promoting venous stasis within the splanchnic circulation [8]. Together, these processes fulfill Virchow's triad of stasis, endothelial injury, and hypercoagulability, thereby predisposing patients to thrombosis despite low platelet counts [9].

Acute non-cirrhotic Portal Vein Thrombosis (PVT) represents a diagnostic challenge because its clinical presentation can closely mimic decompensated cirrhosis. Features such as ascites, splenomegaly, and varices often lead to diagnostic confusion [10]. However, several distinguishing features can aid differentiation. In this case, hepatic synthetic function was preserved, as evidenced by a normal International Normalized Ratio (INR) and only mildly reduced albumin levels attributable to ascites. Imaging findings further supported a non-cirrhotic etiology, with no evidence of hepatic nodularity, regenerative nodules, or surface irregularity [11]. The rapid onset of symptoms over days, rather than the gradual progression typical of cirrhosis, also favored an acute vascular process. Importantly, the presence of an underlying hematological disorder with known thrombotic risk strengthened suspicion for PVT [12]. Maintaining a high index of suspicion is therefore essential, particularly in patients with autoimmune or prothrombotic conditions. Doppler ultrasound remains the first-line diagnostic modality, while contrast-enhanced computed tomography or magnetic resonance imaging provides confirmation and delineates thrombus extent [13].

Management of PVT in Evans syndrome presents a therapeutic dilemma due to the competing risks of thrombosis and bleeding. Anticoagulation is the standard treatment for acute PVT, as it promotes recanalization and prevents thrombus progression, thereby reducing complications such as portal hypertension and intestinal ischemia [14]. However, thrombocytopenia increases bleeding risk, particularly in the presence of esophageal varices. Despite this, available evidence supports anticoagulation in patients with immune thrombocytopenia once platelet counts exceed 30–50 $\times 10^9$/L, alongside treatment of the underlying autoimmune disorder [6]. In this patient, corticosteroid therapy resulted in sufficient platelet recovery to allow safe initiation of anticoagulation. Close monitoring, individualized dosing, and avoidance of antiplatelet agents are essential to balance risks and benefits [15]. Exclusion of secondary causes is a critical component of Evans syndrome evaluation, as it may influence both management and prognosis. In this case, investigations for systemic lupus erythematosus, antiphospholipid syndrome, viral hepatitis, human immunodeficiency virus infection, and JAK2-positive myeloproliferative neoplasms were negative. No evidence of lymphoproliferative disorders was found on peripheral smear or bone marrow examination, although continued surveillance is warranted given known associations [3].

Thrombotic complications in AIHA are increasingly recognized, with reported venous thromboembolism rates of 11% to 15% in some cohorts [4]. However, splanchnic vein thrombosis, particularly acute PVT presenting as rapid-onset portal hypertension, remains rare and is largely described in isolated case reports [10]. This case therefore adds to the limited literature and underscores the importance of recognizing the prothrombotic potential of autoimmune cytopenias. Early diagnosis and a carefully balanced therapeutic approach are essential to optimize outcomes in such complex clinical scenarios.

CONCLUSION

This case demonstrates that Evans syndrome, despite its defining cytopenias, can be complicated by serious thrombotic events, including acute portal vein thrombosis with rapid onset non-cirrhotic portal hypertension. The clinical presentation, ascites, splenomegaly, and esophageal varices may masquerade as decompensated cirrhosis, potentially delaying accurate diagnosis. Clinicians should maintain a high index of suspicion for thrombotic complications in autoimmune cytopenias and pursue appropriate imaging when atypical features are present. Management requires a multidisciplinary approach, balancing the need for anticoagulation against bleeding risk in the context of thrombocytopenia.

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