CASE STUDY

99. The Lupus You Didn't See Coming: Secondary Evans Syndrome with Fulminant Autoimmune Myocarditis in a Young Woman

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Background: Some cases in medicine feel like a thriller, where every test, every symptom, and every intervention builds suspense. We present a young woman whose illness unfolded like a high-stakes medical drama: chest pain, cardiogenic shock, hemolysis, and mystery antibodies all converging in a race against time.

The Case: A 25-year-old female arrived at the emergency room with seven days of dragging left-sided chest pain radiating to the neck, back, and arms, accompanied by shortness of breath (NYHA II–III), palpitations, sweating, orthopnea, and facial puffiness. One day prior, she developed black stools and non-bilious vomiting. On arrival, she was in cardiogenic shock, requiring noradrenaline infusion.

ECG revealed ST-elevation in aVR with global ST-depression, and echocardiography demonstrated global hypokinesia with moderate left ventricular dysfunction. Labs were dramatic: hemoglobin 6 g/dL, highly elevated troponin I, deranged liver function tests, unconjugated hyperbilirubinemia, positive direct Coombs test, and low complement levels (C3 and C4).

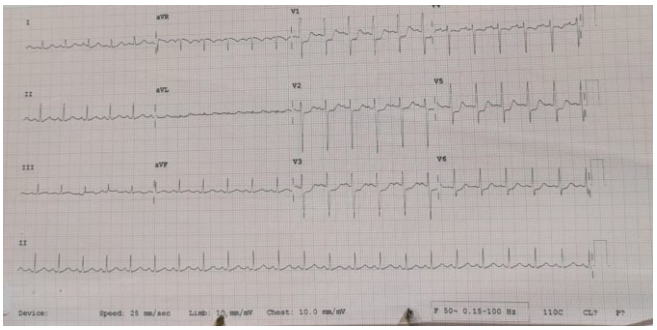
Immediate management included intravenous steroids, multiple blood transfusions, and plasma exchange. Serology revealed strong anti-histone antibody positivity with a completely negative ANA profile (anti-dsDNA, anti-Sm, anti-RNP). Coronary angiography was normal, confirming myocarditis as the source of troponin elevation. Ultrasound showed hepatosplenomegaly, and history uncovered a recent mismatched blood transfusion, complicating the hemolytic picture.

Through rapid multidisciplinary coordination including critical care, cardiology, hematology, rheumatology, and nephrology, the patient stabilized, hemoglobin improved, and she was discharged on a carefully tailored immunosuppressive regimen.

Conclusion:
This case is a diagnostic rollercoaster. Anti-histone positivity initially suggested drug-induced lupus, yet hypocomplementemia, secondary Evans syndrome, and fulminant myocarditis pointed toward an atypical or early-onset SLE. The presentation mimicked acute coronary syndrome, adding urgency and tension. It underscores that anti-histone antibodies should never be interpreted in isolation, and highlights the importance of integrating clinical, serological, and imaging data in complex autoimmune syndromes.

Some patients turn the ER into a thriller. Fulminant autoimmune disorders can masquerade as common emergencies, putting lives at immediate risk. Early recognition, rapid multidisciplinary intervention, and careful interpretation of serology are the keys to survival and successful outcomes. This is an incredibly rare presentation of anti-histone positive lupus, with ours being the second reported case in literature.

Figure 1. lead ECG Strip Demonstrating Global ST Depression.



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