Background

Systemic Lupus Erythematosus (SLE) is a complex autoimmune disorder characterized by its multisystem involvement and an elusive etiological origin. Among the recognized associations of SLE are Evans syndrome and secondary anti-phospholipid syndrome (APLS), both of which contribute to the clinical complexity of this autoimmune entity. However, the simultaneous occurrence of all three conditions in a single patient is a rare phenomenon, presenting significant diagnostic and therapeutic dilemmas. In this medical article, we present a case where Evans syndrome, featuring an unusual direct antiglobulin test (DAT) profile, coexisted with SLE and APLS, and discuss the successful management of this intricate triad of autoimmune disorders.

Case Summary

A 22-year-old female with a history of APS on warfarin presented with a month-long progression of shortness of breath and fatigue. While ruling out common causes like massive pulmonary embolism, further investigation revealed severe anaemia and autoimmune hemolytic anaemia (AIHA) along with immune-mediated platelet destruction, leading to a diagnosis of Evans syndrome. Additionally, a mesenteric angiogram and computed tomography pulmonary angiogram (CTPA) showed possible portal vein thrombosis and splenic infarction and segmental pulmonary embolism respectively. The patient received intravenous immunoglobulin (IV Ig), methylprednisolone, and IV rituximab. Due to worsening thrombocytopenia, therapeutic plasma exchange was performed, followed by IV cyclophosphamide, resulting in gradual improvements in both haemoglobin and platelet counts.