

Anti-dsDNA and anti-Smith antibody double negative SLE presented by severe hemolytic anemia

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Introduction: Anti-dsDNA antibody and anti-Smith antibody are lupus-specific autoantibodies, typically positive in the majority of SLE patients, and anti-dsDNA antibody is associated with disease activity. Here, we report a case of SLE presenting with severe hemolytic anemia despite negative anti-dsDNA and anti-Smith antibodies.

Case: A 23-year-old woman presented with dyspnea and chest discomfort. She had a family history of a blood-related autoimmune disease, and her sister passed away from it during her teenage years. Her blood pressure was 109/58 mmHg, heart rate 121 bpm, respiratory rate 22 breaths/min, and body temperature 39.4°C. Laboratory tests showed a markedly low hemoglobin level (3.8 mg/dL) and hemolytic feature (total bilirubin 5.3 mg/dL, LDH 2241 U/L, haptoglobin <30 mg/dL, and direct Coomb's test 4+). Antinuclear antibody test revealed a speckled pattern (1:160). Anti-dsDNA and Smith antibodies were negative, but anti-SS-A and anti-RNP were positive. Complement levels decreased (C3 40.5 and C4 1.48 mg/dL). Anti cardiolipin antibody IgG (137 GPL-U/ml) and anti-beta2-glycoprotein 1 IgG (19.7 U/mL) were positive. She was diagnosed with SLE with severe hemolytic anemia based on the 2019 EULAR/ACR criteria (her total score was 12). During admission, she experienced severe visual impairment and retinal hemorrhage obstructing the macular was observed on optical coherence tomography (OCT) (figure 1a). Additionally, EKG showed T-wave inversion and second-degree atrioventricular block. No significant cardiac enzymes changes or abnormal coronary CT finding were noted. She received steroid pulse therapy (500mg for 3 days), prednisolone (1mg/kg), and 1 unite of packed RBC transfusion, resulting in improvement of hemoglobin levels, resolution of retinal hemorrhage (figure 1b), and normalization of heart rhythm.

Conclusion: The case highlights the importance of considering clinical manifestations as valuable diagnostic clues in rheumatic autoimmune diseases, even in the absence of positive disease-specific autoantibodies. Relying solely on antibody test results may lead to misdiagnosis or overlooked the correct diagnoses.

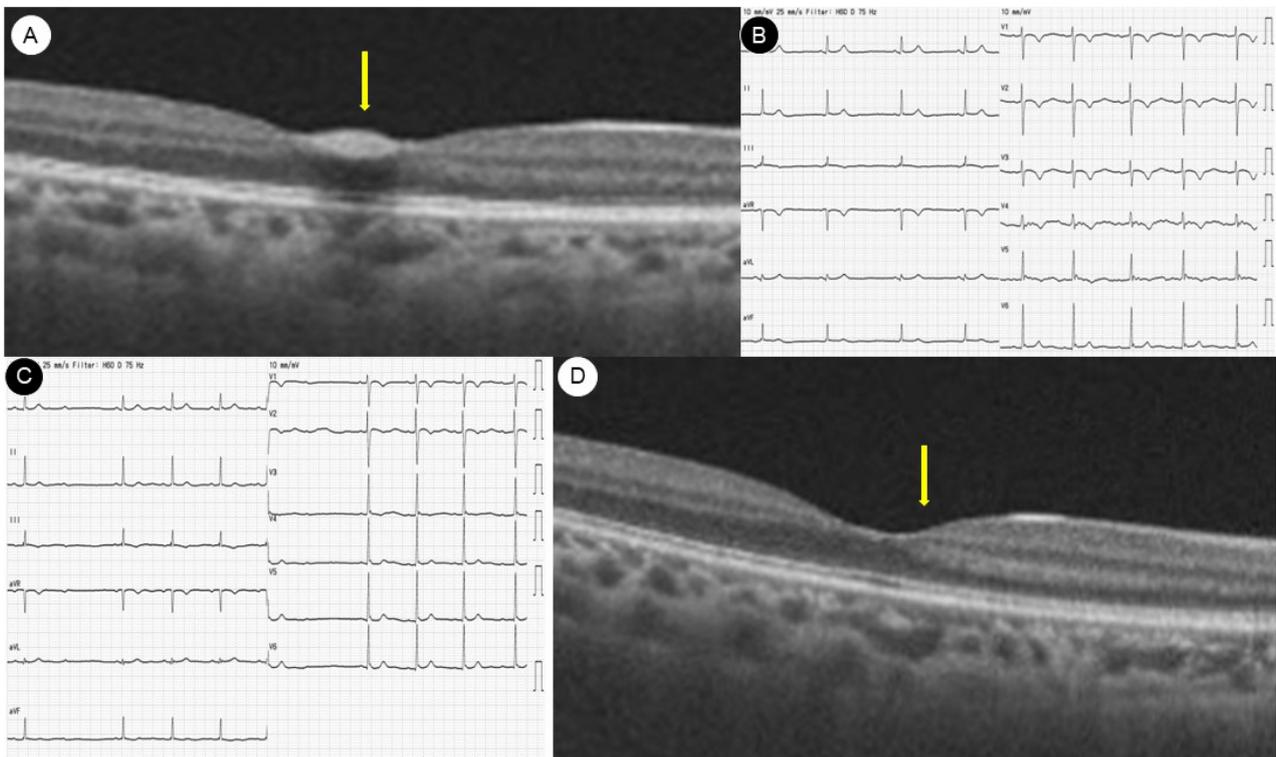


Figure 1. Retinal hemorrhage on optical coherence tomography (OCT) (A), T-wave inversion on electrocardiogram (B), second-degree atrioventricular block ©, and resolution of retinal hemorrhage on OCT after steroid pulse therapy (D)