

Category: Présentation de cas clinique / Clinical case presentation

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Title: A Special Case of Antiphospholipid Syndrome

Objective(s): We present the case of a 30-year-old female patient of West African origin known for menorrhagia.

Result(s): In 2016, she was diagnosed with a triple positive antiphospholipid antibody syndrome (APLS) with significant history of venous thromboembolism causing chronic thromboembolic disease. She required an inferior vena cava filter and different anticoagulants over time, including warfarin. In 2019, a diagnosis of systemic lupus erythematosus (SLE) was confirmed in the context of fever, discoid rash, hemolytic anemia, hypocomplementemia, and serologies that included ANA (1:160 speckled), anti-RNP, -Sm and -dsDNA. She was initially treated with hydroxychloroquine, prednisone, and intravenous immunoglobulin (IVIg). Mycophenolate mofetil (MMF) was introduced later due to pericarditis, arthralgias, and suspected lupus nephritis.

In November 2023, she was admitted to the Intensive Care Unit for a ruptured hemorrhagic ovarian cyst with hemodynamic compromise. Despite anticoagulation reversal and withdrawal, numerous blood transfusions and laparotomies, her abdominal bleeding could not be controlled. At this stage, rheumatology and hematology were consulted. She had active SLE with a discoid rash, arthritis, hypocomplementemia, and positive anti-dsDNA and Coombs test. Keeping in mind her lupus anticoagulant affecting coagulation testing, her International Normalized Ratio was 1.8-1.9, activated partial thromboplastin time 100-130 seconds, prothrombin time (PT) 20 seconds; and her factor II levels were persistently low despite serial dilutions. These abnormalities persisted after anticoagulation reversal and prothrombin complex concentrate administration. Based on this presentation, a diagnosis of Lupus Anticoagulant Hypoprothrombinemia Syndrome (LAHPS) was established. She received rituximab 375 mg/m² weekly for 4 weeks, prednisone 1 mg/kg/day, and IVIg, resulting in normalization of factor II levels and lupus activity parameters. She was successfully discharged from the hospital with no further bleeding episodes 8 weeks later.

LAHPS is a rare bleeding diathesis, often associated with SLE, primary APLS, infections, or malignancy, caused by acquired autoantibodies against factor II, which increase its clearance. It should be suspected in the case of a prolonged PT, low factor II, and lupus anticoagulant positivity in presence of hemorrhagic, rather than thrombotic, complications. No established treatment guidelines exist for LAHPS, but severe cases and those associated with SLE or APLS reported in the literature were generally treated with high glucocorticoid doses alongside supportive care, most often normalizing factor II levels and PT within one month. Reports of successful treatment with cyclophosphamide, azathioprine, IVIg, plasmapheresis, hydroxychloroquine, cyclosporin, MMF, danazol, or rituximab have also been published.

Conclusion(s): Failure to recognize LAHPS as a complication of SLE can be fatal.