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Bleeding in Anti-Phospholipid Syndrome

Tzeng Lin Wong[1,2], Sargunan Sockalingam[1], Jasmin Raja[1], Fariz Yahya[1]

[1] Division of Rheumatology, Department and Faculty of Medicine, Universiti Malaya

[2] Division of Rheumatology, Department and Faculty of Medicine, Universiti Putra Malaysia

Introduction

Lupus anticoagulant-hypoprothrombinaemia syndrome (LAHPS) is a rare coagulopathy characterized by acquired factor II deficiency due to presence of lupus anticoagulant, resulting in mild to severe bleeding diathesis.

Report

A 39-year-old woman with a history of systemic lupus erythematosus (SLE) with hematological involvement since 2008 has been managed successfully with steroids alone. In 2012, she was diagnosed with antiphospholipid syndrome (APS) after two first-trimester miscarriages, evidenced by positive lupus anticoagulant and elevated anticardiolipin antibodies. Her coagulation profile showed normal prothrombin time (PT) but prolonged activated partial thromboplastin time (APTT) that was not corrected by a mixing test. She experienced recurrent severe immune thrombocytopenic purpura in 2013 and 2017, managed with IV rituximab and IV immunoglobulin, respectively. Mycophenolate mofetil 500 mg BD stabilized her blood counts.

In 2018, she developed menorrhagia and abnormal uterine bleeding. Despite treatment with a Mirena IUD and hormonal therapy, her symptoms worsened in 2023, requiring multiple blood transfusions. Coagulation tests showed prolonged PT and APTT with a low factor II level (23.7%), confirming LAHPS. Other tests indicated normal fibrinogen and von Willebrand factor activity, with low factors VIII, IX, and XII likely due to lupus anticoagulant interference. Bleeding resolved after IV rituximab 1g biweekly followed by mycophenolate mofetil 1000 mg BD.

LAHPS involves acquired factor II deficiency due to antiphospholipid antibodies. In this syndrome, these antibodies target factor II, resulting in hypoprothrombinaemia and an increased bleeding risk.

LAHPS should be considered when antiphospholipid antibodies are present, and both APTT and PT are prolonged. Although rare, the syndrome can lead to severe bleeding. 55% of LAHPS cases are associated with autoimmune conditions, with 40% of cases involving SLE. 33% being infection-related, and 5% linked to malignancies.

No standardized guidelines exist. Treatment includes supportive care and immunosuppressive therapy. Corticosteroids are the first-line agents, while refractory cases may require cyclophosphamide, azathioprine, mycophenolate mofetil, IV immunoglobulin, or rituximab. The combination of rituximab and corticosteroids has shown efficacy. In this case, the patient responded well to rituximab and maintained remission with mycophenolate mofetil.

Conclusion

This case underscores the importance of recognizing LAHPS in patients with unexplained prolonged PT, APTT and bleeding history, especially with antiphospholipid syndrome. Autoimmune- associated LAHPS has a favourable prognosis with timely treatment. Early and aggressive immunosuppression is crucial to prevent haemorrhagic complications.