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A rare presentation of factor VIII inhibitor with lupus anticoagulant and lupus enteritis in a case of antiphospholipid syndrome

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Acquired hemophilia or factor VIII (FVIII) deficiency, caused by FVIII inhibitor antibodies is a rare condition. The concomitant presence of lupus anticoagulant and FVIII inhibitor is exceedingly rare. We report a case of Acquired FVIII inhibitor with lupus anticoagulant and lupus enteritis in a case of Antiphospholipid syndrome (APLA). These patients are at high risk of spontaneous bleeding, hence need to be identified and monitored closely. A 51 years female with Systemic Lupus Erythematosis since 16 years (from 2001) presented with abdominal pain of three months duration, not responding to symptomatic treatment. She was scheduled for endoscopy and was advised prior coagulation screening. Her activated partial thromboplastin time (APTT) was prolonged, immediate and delayed mixing studies, using normal plasma, showed no correction of APTT. This suggested the presence of an inhibitor. A Dilute Russel's Viper Venom Test (DRVVT) confirmed presence of lupus anticoagulant. However, due to her long standing history of APLA and clinical suspicion, a factor VIII assay was performed. The patient was found to show concomitant FVIII inhibitor. The abdominal pain, following endoscopic examination revealed lupus enteritis. She has chronic hemolytic anemia since 2001 with spherocytes, reticulocytosis and Direct Antiglobulin Test (DAT) positive at the time of presentation. On clinical grounds, she was evaluated for autoimmune disorders and was found to be ANA and SSRo positive. The DRVVT was negative at this time and the coagulation studies were normal. She was diagnosed and treated for systemic lupus erythematosus/Sjogren's syndrome with autoimmune hemolytic anemia. The lupus enteritis is responding well to Prednisolone and she is being closely monitored for her hematological condition.

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