Acquired Hemophilia A associated with Systemic Lupus Erythematosus

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Acquired hemophilia A (AHA) is a very rare hemorrhagic disorder caused by autoantibodies against factor VIII (FVIII). AHA presents extensive clinical manifestation and associated with many other clinical states. This is a first case of AHA associated with systemic lupus erythematosus (SLE) in Korea. Eighty-year-old woman visited the hospital because of multiple bruises on upper and lower extremities, gross hematuria. There were large ecchymosis and swelling on buttock area. Anemia (Hb 9.6 g/dL) and normal platelet counts were seen. Initial coagulation results showed prolonged aPTT (68.5 sec) and normal PT. From mixing test, FVIII activity was decreased (2%), FVIII inhibitor (FVIII-I) titer was increased (74.4 BU) and lupus anticoagulant was negative. Antinuclear antibody (ANA) and anti beta2 glycoprotein I antibody (anti beta2 GPI) were positive. AHA was diagnosed on the basis of late onset bleeding and increased FVIII-I. In addition, the patient meets the criteria of SLE[oral ulcer, proteinuria, ANA (+), anti beta2 GPI (+)]. We start oral prednisolone for FVIII-I eradication. After 4 months, patient’s bleeding tendency, aPTT and FVIII-I were decreased (47.3 sec, 1.24 BU), and FVIII activity was increased (10%, Figure 1). Treatment principles of AHA are to control the bleeding, eradicate the inhibitor, and treat underlying disease. In this patient, bleeding tendency improved and the titers of FVIII-I decreased after treatment with steroid. Because of old age and combined disease (SLE), we would like to minimize steroid dose and add hydroxychloroquine.

The fluctuation of serum anti ds-DNA antibody level has strong relationship with lupus flare up

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Objective: Patients with Systemic Lupus Erythematosus (SLE) may experience flare of disease activity. The aim of this study was to assess clinical features of flare, focusing on the relationship with serially assessed anti-double stranded DNA antibody serum levels by Farr assay, through the analysis of a monocenter cohort of SLE patients and a literature review. Methods: We analyzed 159 patients who visited outpatients or admitted to Dong-a university hospital between march 2012 and march 2015. They were diagnosed to SLE, according to the revised 1997 ACR criteria and serially assessed serum anti-dsDNA every three months. Among subjects, 62 subjects were excluded because of drop-out. So 97 patients were enrolled and classified into three groups. 29 patients showed significant increase or decrease of serum anti-ds-DNA level (group 1), another group, 33 patients maintained high level anti-ds-DNA (group 2), the other group, 35 patients maintained lower level anti-ds-DNA (group 3). Informed consents were obtained from all individual participants in the study. Results: The mean SLEDAI score change was 4.21 in 29 patients who showed fluctuation of serum anti-dsDNA level. In contrast, SLEDAI score showed no significant change (0.47, 0.51) in group 2 and group 3(\(p=0.000\)). The 14 patient who showed increased serum anti-dsDNA level, had hematologic and cardiopulmonary exacerbation. 15 patients, who showed decreased serum anti-ds-DNA level, had worsened musculoskeletal and mucocutaneous manifestation. Conclusions: The increased SLEDAI score is associated with change anti ds-DNA antibody level and increased anti-ds-DNA level subjects had more severe clinical manifestation than decreased anti-ds-DNA level subjects.