中文題目：成功的以類固醇治療 A 型後天血友病: 案例報告

英文題目：Acquired Hemophilia A Successfully Treated With Corticosteroid : a case report

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Introduction

Acquired hemophilia A is a rare disease caused by autoantibody against coagulation factor VIII. It commonly affects elderly population with serious bleeding and atypical bleeding pattern. The clinical manifestation of acquired hemophilia A with high titer factor VIII inhibitor is usually massive hemorrhage into skin, soft tissue or mucosal area, which needs quick diagnosis and management. Symptomatic patients with low titer inhibitor and no major bleeding, however, maybe ignored or under diagnosed.

We describe a case of symptomatic acquired hemophilia A with low titer inhibitor successfully treated with corticosteroid.

Case Presentation

A 57-year-old Chinese woman, with a past medical history of gastric ulcer and type 2 diabetes mellitus, presented to our rheumatologic out-patient department with complaints of headache and hematomas spread all over her limbs for one month. She had four vaginal deliveries without any major bleeding episodes in the past.

In June, 2015, she was admitted to our Department of Rheumatology for pain and multiple hematomas developed over her four limbs (Figures 1 and 2). There was no mucosal bleeding, hemarthrosis, hematuria or melena during physical examination. Laboratory exams revealed a white blood cells count of 3360 cells/ μ L, a hemoglobin level of 12.5 g/dl, a platelet count of 220000 cells/ μ L. Coagulation profiles were prothrombin time (PT) 9.9 seconds (control 10.5 seconds), elevated aPTT of 42.4 seconds (control 24.5 seconds). An aPTT mixing study was performed with the aPTT correcting to 30.9 seconds. 2 hours incubation study showed aPTT elevated again to 36.7 seconds. Additional laboratory evaluation demonstrated factor VIII level 18% (normal 60-150%) and a factor VIII antibody was detected with a titer of 0.32 Bethesda Units (BU)/mL (normal < 0.05 BU/mL). Factor IX, factor XI and Von Willebrand factor levels were in normal limits. Anti-cardiolipin antibodies, lupus anticoagulant and anti-β2-glycoprotein I IgG were all negative. Given the isolated prolongation of aPTT, time dependent inhibition during mixing study, low factor VIII level, presence of factor VIII inhibitor and lack of prior history of major bleeding, a diagnosis of acquired hemophilia A was made.

The goal of management was to control of bleeding and eradicate the inhibitor. Treatment during hospitalization consisted of three cycles of fresh frozen plasma transfusion (FFP) 2 units/day, three cycles of intravenous methylprednisolone 80 mg/day in two divided doses. On the fourth day after treatment, her symptoms showed a significant relief, improvement of headache and pain over extremities, regression of ecchymosis over her limbs. She was discharged from using dexamethasone 4 mg/day and weekly followed up at outpatient room was arranged.

Normalization of aPTT and no new hematomas formation was noted during followed up. In September, 2015, dexamethasone was discontinued. She is currently being followed monthly with favorable condition.

Discussion

This case report shows the importance of analyzed the coagulation system, especially when patient experienced atypical symptoms, such as prolonged bleeding over wound, bleeding into soft tissue, or massive mucosal hemorrhage.

Clinical awareness of atypical symptoms and good collaboration with laboratory staff are important for early diagnosis. The patient experiences good response after corticosteroids treatment. Further studies with more patients are needed if this group of patient is different from patient with high titer inhibitor in prognosis and relapse rate, or it will transform from low titer inhibitor to high titer if without treatment. Further follow-up is also necessary to see if our treatment modality is adequate.