中文題目：處理後天性A型血友病患者的出血問題及其併發症

英文題目：The management of acute bleeding and associated complication in a patient with Acquired hemophilia A.

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Introduction:

Acquired hemophilia A (AHA) is a rare but potentially life-threatening autoimmune disorder that is characterized by the abrupt onset of bleeding in patients with negative family and personal history of bleeding. Treatment should be control bleeding with bypassing agents, and eradication of the inhibitor with immunosuppress agents.

Abstract:

We reported a case of 67-year-old man, presented to KMUH ER because of ecchymosis of right thigh and progression in these 3-4 days. Lab data showed normocytic anemia, and isolated prolongation of activated partial thromboplastin time (aPTT). PTT mixing test showed 87.9sec, clotting factors VIII< 1%, and bethesda unit=141B.U. FFP transfusion and oral steroid prednisolone were used. But due to difficult approach to his peripheral vein for blood sampling, artery puncture at right femoral artery was done. After 2 days, the patient had hematoma over his right inguinal area. Right leg computed tomography (CT) highly suspect abscess formation over iliopsoas muscle. Therefore, recombinant human coagulation Factor VIIa (rFVIIa), accompanied with immunosuppression agents Cyclophosphamides and IV form Methylprednisolone were used. Then, FEIBA (aPCC, Activated prothrombin complex concentrate) was applied to him. However, fever with bacteremia noted after several days, and blood culture report reveals Salmonella group D1 (not typhi). For infection, antibiotics was used for a period (14 days). After his condition improved, he was discharged.

One month later, he presented to ER again because of increasing abdomen circumference with fever. Abdominal CT showed abscess around right paoas and iliopsoas muscles. CT-guided PTAD (percutaneous abdominal drain) was arranged. Then pus culture of abdominal abscess yields Salmonella D1(non typhi), the same pathogen as previous blood culture report. He then had antibiotics treatment with Ertapenam for 14 days, and was discharge with oral form antibiotics. He regular follows up at hematology OPD, and lab data reveal normal PTT, without bleeding episode.

Conclusions:

1. Acquired hemophilia A has high risk of bleeding; if coagulopathy had not been corrected, we should always avoid unnecessary invasive procedure, including puncture of artery.
2. We should be aware of complication about bleeding; as for compartment syndrome associated with hematoma over extremities and intra-abdominal abscess related to hematoma, early detection and proper treatment are important.