Currently, there is no standard treatment for patients with T-LGL. For asymptomatic T-LGL patients with an indolent course, a wait-and-see approach can be considered [11]. In our series, all 4 patients are alive and stable, with a slowly declining trend of platelets during the more than 3-year follow-up. T-LGL clonality could not be confirmed by a T-cell receptor gene rearrangement in our cases because our facility lacks this capacity. The clinical features and laboratory findings of the T-LGL patients in our study were similar to that reported in the literature. It remains unclear if the incidence is truly low or the disease has been underdiagnosed because most cases are asymptomatic on presentation. This raises the importance of reviewing the peripheral smears in asymptomatic patients who have persistent lymphocytosis or neutropenia. Systematic long-term follow-up studies need to be performed.

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Second case of postpartum acquired hemophilia A in a Korean female

TO THE EDITOR: Acquired hemophilia A (AHA) is a very rare condition with an incidence of about 1.5 per million of the general population per year [1]. It develops in middle age due to autoantibody production directly against the factor VIII (FVIII) and occurs in both sexes equally. The underlying conditions associated with AHA are other autoimmune diseases, malignancy, drugs and pregnancy. However, about half of AHA has no specific cause, thus they are called idiopathic. Postpartum AHA is a special category with distinct clinical manifestation contributing to 7-21% of patients with AHA [2-4]. In Korea, the first case of postpartum AHA in a 40 years old female has been reported by Lee *et al.* [5]. To the best of our knowledge, the present case report is the second one in Korea.

A previously healthy 18 years old female had several minor bruises on hands and feet at 5 months after delivery. There was no specific complication immediately after delivery. She previously didn't take any drugs which could result in AHA and her family history was nonspecific. The hemorrhagic symptoms were aggravated during the process of time and she visited a private medical doctor at 8 months after delivery. At the medical clinic, she was only supplemented with iron to treat her iron deficiency anemia.

Because of a persistent migrating painful swelling on her knees and ankle joints, multiple bruises and menorrhagia, she was transferred to our institute at 9 months after delivery.

Her initial laboratory test results were as follows: PT 10.2 sec (normal range, 10–14 sec), INR 0.95 (normal range, 0.85–1.50), aPTT 84.3 sec (normal range, 20–40 sec), FVIII activity 1.4% (moderately decreased), and anti-FVIII antibody 28 Bedestha units (BU). Thus, she was confirmed as postpartum AHA. To manage her persisting hemorrhagic symptoms, activated prothrombin complex concentrate, FEIBA (Baxter, Westlake Village, CA, USA) was infused to her based on the dose for joint hemorrhage (50–100 units/kg every 12 hours until pain/disabilities are improved, maximum 200 units/kg/day) and her bleeding symptoms resolved. We closely followed up her at the outpatient clinic with on-demand FEIBA injection. After 2 months, the last follow up PT/aPTT were 11.2/95.1 sec, FVIII activity 1.5% and anti-FVIII antibody 40 BU. We consider giving corticosteroid to her additionally.

According to European Acquired Haemophilia (EACH2) registry, 42 cases of postpartum AHA developed from 2003 to 2008 in 13 European countries [4]. In other words, the medical personnel in hematology or obstetrics can experience one postpartum AHA patient every year or biyearly in one country. Because time to diagnosis of postpartum AHA was 21–120 days after delivery, pregnant women should be observed closely approximately 1–3 months after delivery [4].

Usually, AHA is often misunderstood as another bleeding disorder like disseminated intravascular coagulation because a hemorrhage into the skin (purpura) or soft tissue is the most common presenting sign of AHA, while a hemarthrosis, the hole mark of a congenital hemophilia, occurs rarely [1, 2]. According to EACH2, hemarthrosis was also a rare hemorrhagic symptom of postpartum AHA contributing about 5%. Subcutaneous (45%) or mucosal bleeding (43%) was most common. On the other hand, in case of our report, the patient was mistaken as to have rheumatic disease because the migrating painful swelling of her joints due to hemarthrosis was the main symptom.

The treatment of acute bleeding in case of postpartum AHA is not different from the general form. Two bypassing agents, FEIBA and the recombinant activated factor VII, NovoSeven (Novo Nordisk, Princeton, NJ, USA) are the main therapeutic agents to control acute hemorrhagic symptoms. The overall complete response rate of hemorrhages using FEIBA was 86% with the infusion of 75 units/kg every 8 to 12 hours [6]. And the efficacy rate using NovoSeven was 95% as first line therapy for AHA [7]. NovoSeven has some advantages that it's blood born viral transmission is shut out and the risk of thromboembolism is lower than that of FEIBA [8, 9].

The combination of bypassing agents for stopping the hemorrhage and immunosuppressive therapy is usually used together to eradicate the autoantibodies of FVIII in the general form of AHA. Prednisolone (prednisone), cyclophosphamide, azathioprine, 6-mercaptopurine, rituximab (anti-CD 20 monoclonal antibody), mycophenolate or cyclosporin have been used for treatment [1, 3, 10-12].

However, the immunosuppression may be individually reserved considering the advantages and disadvantages in case of postpartum AHA because of its more favorable prognosis compared to the general form. In retrospective reviews of postpartum AHAs, 86–100% of patients showed complete remission and survival rate was 97–100% [4, 12]. Further, there was no statistical difference of the time to complete remission between the postpartum AHA patients had immunosuppressive therapy and the rest of cohort [4]. On the other hand, the course of a general AHA is quite different. The relapse rate was about 20% after the cessation of immunosuppression and the survival rate was about 60% [1].

In some international survey, the recurrence rate of postpartum AHA was 0% [13, 14]. However, there is a controversy over the recurrence rate of anti-FVIII antibody in case of subsequent pregnancy. Large-scaled study about the recurrence rate is not available yet. Thus the patients who experienced postpartum AHA should be observed carefully after subsequent pregnancies.

In the previous Korean case of postpartum AHA reported by Lee *et al.* [5], the patient was treated with a bypassing agent plus corticosteroid. In the present case, we are using on-demand FEIBA infusion for control of acute bleeding and postponed the immunosuppression considering a spontaneous remission. In our present case, the hemorrhagic symptoms of the patient were now solved using FEIBA. We expect that case presentations will help to increase the awareness on this rare condition among the medical personnel in the hematologic and obstetric field.

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