Acquired Hemophilia A with Immune Thrombocytopenia in Adolescent Patient

Hyo Sun Kim[1], Jung Woo Han[1], Jae-Woo Song[4], Seung Min Hahn[1], Yoon Jung Shin[1], Sun Hee Kim[1], Moon Kyu Kim[2], Kun Soo Lee[3], and Chuhl Joo Lyu[1]

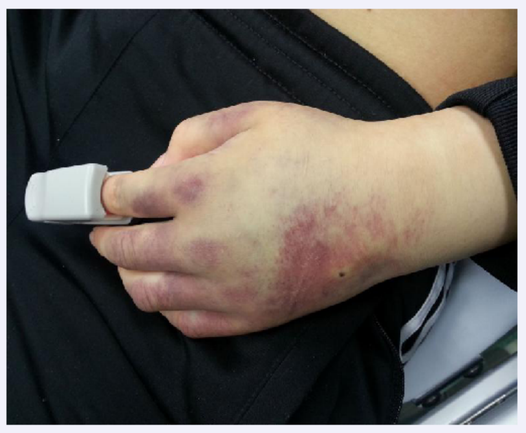
Department of Pediatric Hematology and Oncology, Yonsei University Health System, Seoul, Korea[1],
CHA Bundang Medical Center, CHA University, Seongnam, Korea[2], Kyungpook National University School of Medicine, Daegu, Korea[3],
Department of Laboratory Medicine, Yonsei University College of Medicine, Seoul, Korea[4]

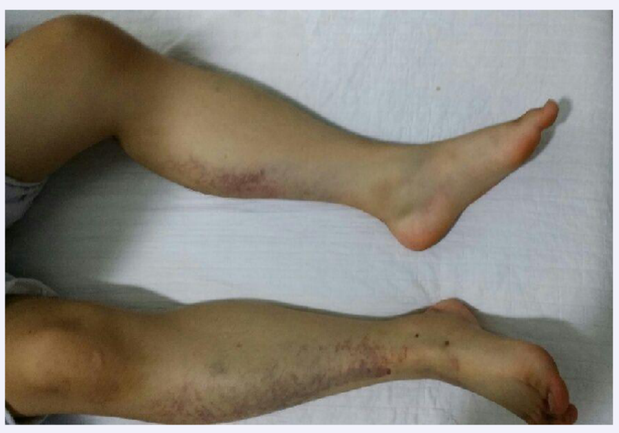
Background

- Acquired hemophilia A (AHA) is rare autoimmune disorder caused by autoantibodies against coagulation factor VIII (FVIII) in the nonhemophilic population.
- The age distribution of autoantibodies typically biphasic with small peak between 20 and 30 years (mainly postpartum inhibitors) and a major peak in patients aged 70 to 80 years. The incidence of AHA in children is very rare. Indeed, the incidence in children under 16 years has been estimated to be 0.045 per million/year compared with 14.7 per million/year in the elderly over 85 years.
- Also, immune thrombocytopenia is an acquired immune-mediated disorder caused by increased destruction of platelets opsonized by anti-platelet autoantibodies.
- We experienced a case of AHA with thrombocytopenia caused by autoantibody in 18 years old boy.

Case Report

- Patient : 18 years old , male
- Family history : none
- Past bleeding history : none
- Chief complaint : multiple bruises on both extremities
- He had no definite medication history.





- A initial activated partial thromboplastin time (APTT) was 136 seconds, the level of FVIII was 0.6 % and FVIII inhibitor was 13.1 Bethesda Units (BU). Moreover, platelet counts were low (34,000/uL), and we proved platelet-associated autoantibody. Other autoimmune diseases were ruled out.
- He was instantly treated with oral prednisolone (1mg/kg/d) and added to oral cyclophosphamide (2mg/kg/d) one week later due to elevation of FVIII inhibitor (14.7 BU). About seven weeks later after treatment, FVIII inhibitor was disappeared and APTT was normalized, we started to tapering off the medication. During treatment of inhibitor eradication, platelet counts were also increased and normalized.

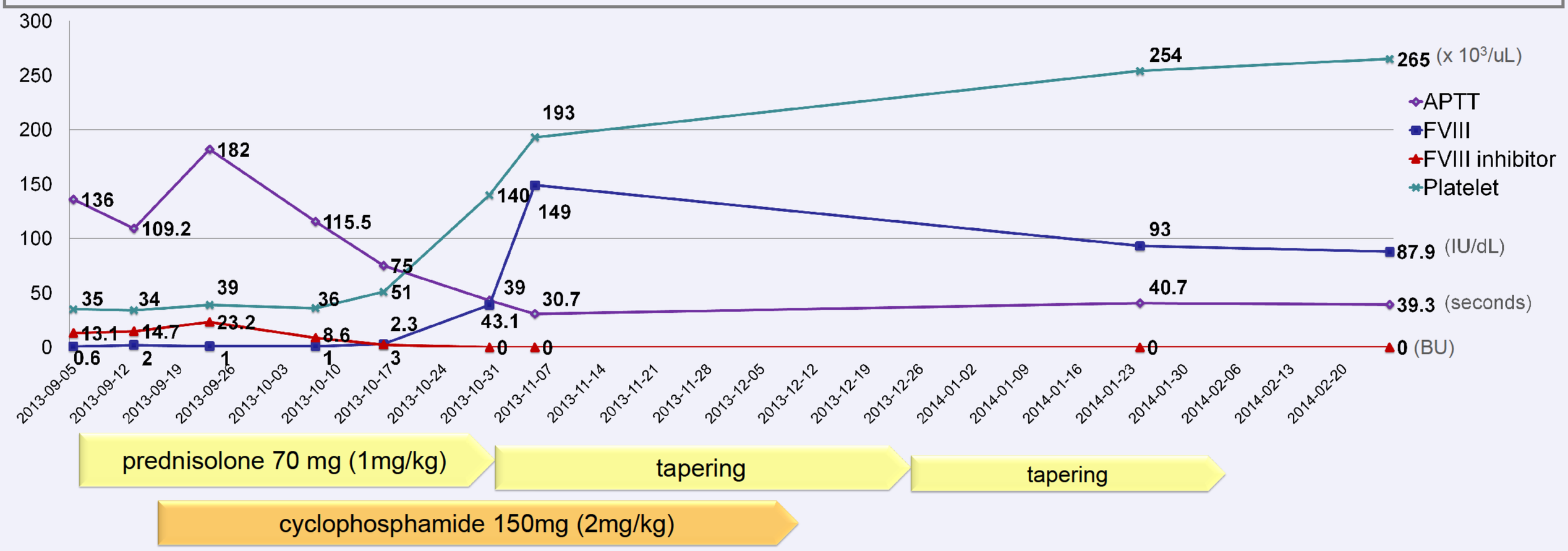


Figure 1. Alterations of laboratory finding according to immunosuppressive treatment for antibody eradication

Conclusions

- AHA patients should receive immunosuppressive therapy for inhibitor eradication immediately following the diagnosis. The most commonly used therapeutic strategy for complete inhibitor eradication is based on corticosteroids, alone or in combination with cyclophosphamide (approximately 70-80%).
- We report our experience as first successful antibody eradication for FVIII inhibitor and platelet-associated autoantibody simultaneously in adolescent AHA patient.

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Hyo Sun Kim
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