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A Case of Small Lymphocytic Lymphoma After Treatment of Cold Agglutinin Disease

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Cold agglutinin disease (CAD) is a most common autoimmune hemolytic anemia (AIHA) induced by cold antibody. CAD represents approximately 16-32% of AIHA cases and causative cold autoantibodies commonly show specificity against the I antigen.. CAD is commonly related with lymphatic neoplasia or infection. Primary cold agglutinin disease will change to malignant lymphoma after several years. We experienced a case of small lymphocytic lymphoma after treatment of primary cold agglutinin disease. The patient was a sixty seven year old man who complained of neck mass. Node biopsy revealed diffuse neoplastic proliferation of small sized monotonous lymphoid cells with polygonal or oval shaped nuclei. Immunohistochemical stain revealed CD20 and CD5 positive. We teated this patient with chlorambucinl and prednisolone. Follow up chest and neck CT scan after treatment revealed stable disease state. We report a case of small lymphocytic lymphoma after treatment of cold agglutinin disease and review the literature.

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A case of Von Willebrand's disease type 2N diagnosed by using gene mutation analysis

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Von Willebrand's disease is a hemorrhagic disorder caused by deficiency or dysfunction of von Willebrand factor (vWF), and von Willebrand's disease type 2N is caused by homozygous or compound heterozygous vWF mutation that impairs binding to factor VIII.

A 39-year-old Nepalese woman admitted for evaluation of coagulation problem. She has been experienced deep muscle and joint bleeding since childhood, and one of her brothers had similar to hers. She has been suffered from irregular menstruation interval and menorrhagia. She treated for intramuscle bleeding on right thigh 4 months ago.

On laboratory test, hemoglobin was 12.8g/dL, and WBC was 8600/μL, and platelet was 283,000/μL. Bleeding time was 2min (Normal range: 1-4) and prothrombin time was 14.9sec (Normal range: 11-14), and activated partial thrombin time was 51sec (Normal range: 24-38), which was corrected by mixing with normal plasma after 2hours incubation. Coagulation profiles were factor VIII activity 4.85% (Normal range: 50-150), factor IX 73% (Normal range: 60-140), vWF related Ag 71.30% (Normal range: 47-197), and vWF ristocetin cofactor activity 130.4% (Normal range: 52-154). On vWF gene mutation analysis, we found a homozygous mutation (C2446T) in Exon 19[2:2446 C>T (p. R816w) (homozygous)]. Finally, she was diagnosed as von Willebrand's disease type 2N, and we recommended that she must be treated with factor VIII-vWF concentrate when she has bleeding problem or needs operation.

Patients with von Willebrand's disease type 2N have normal levels of vWF related Ag and vWF ristocetin cofactor activity but only factor VIII is decreased. For these reasons, von Willebrand's disease type 2N can be mistaken for a hemophilia A. Gene analysis would be needed for differential diagnosis of von Willebrand's disease with hemophilia A.