Poster Board P5-044

0739 AN INHIBITOR TO FACTOR VIII ANTIGEN PRESENTING AS ACQUIRED VON WILLEBRAND DISEASE

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A patient with clinical and laboratory evidence of acquired von Willebrand disease is described. He presented with the recent onset of spontaneous hemorrhage and demonstrated a prolonged bleeding time, reduced factor VIII:C, and undetectable factor VIII:AG and factor VIIIR:WF. Following transfusion of cryoprecipitate, there was a smaller than expected immediate increase in factor VIII:C, factor VIIIR:WF and factor VIIIR:AG, with rapid return to baseline levels, and no secondary rise in factor VIII:C. An inhibitor could be demonstrated in the patient's plasma which markedly decreased the level of factor VIIIR:AG in normal plasma while only weakly decreasing the activity of factor VIIIR:WF. Activity against factor VIII:C was demonstrable in vitro using a concentrated inhibitor preparation. The inhibitor was contained in the IgG fraction of plasma and lacked precipitating properties. This inhibitor, which demonstrates a specificity not previously seen in spontaneous anti-factor VIII antibodies, provides further evidence for the separate identity of the antigen and von Willebrand factor sites of the factor VIII molecule. It also indicates that inhibition of ristocetin-induced aggregation of platelets is not the most sensitive method for the detection of an inhibitor in all cases of acquired von Willebrand disease.

P5-045

0740 COAGULATION DISORDERS IN FULMINANT HEPATITIS TREATED BY DIALYSIS

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A young man, 28 years old, was admitted to Infectious Division with transaminases over 2500 U/ml, reduced cortical activity and decerebrate posturing. A prolonged APTT, Prothrombin below 15%, very low levels of Factor II, V, VII (5-20%), AT III by S-2238 (17%), Antiplasmin (20%) and Plasminogen (5%) by S-2251, Prekallikrein by S-2302 (5%), Fibrinogen (70mg%) but very high VIII AHF (4.5U/ml), VIII AGN (4.0U/ml) and VIII VWF (3.8U/ml) were recorded. After daily dialysis sessions with polyacrilonitrile membrane (RP 6) a marked improvement was observed. The patient awoke while prothrombin and platelet recovery took place, fibrinogen, VIII AHF and Factor V rose over normal value to 600mg%, 7.5U/ml and 3U/ml rispectively. Antiplasmin, Plasminogen, PKK showed a slow but costant improvement. Unfortunatly a venous thrombosis and sepsis set in during the 2nd day after dialysis, with a rapid decrease of platelets. Heparin infusion 1 mg/kg b.w. was infused every 6 hrs. After 15day platelet returned to normal value and VIII AHF to initial level but it was still higher (4U/ml) than normal value after 2 mounths from recovery.

P5-046

0741 HOMOLOGOUS ANTIBODIES TO FACTOR IX SHORTEN THE BOVINE THROMBOPLASTIN COAGULATION TIME OF HUMAN PLASMA

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We have previously demonstrated that neutralization of factor IX in normal plasma by heterologous antisera shortens the one stage prothrombin time determined with bovine thromboplastin. In this study we demonstrate a similar effect of a homologous antibody. Addition of 1/5 volume of plasma from a patient with hemophilia B- and an acquired inhibitor to factor IX (titre 5 U/ml) gave a shortening of the prothrombin time of plasma from 7 normal persons (mean 34 sec) compared to the prothrombin time determined after addition of 1/5 volume of control plasma from a patient with hemophilia B- and no acquired inhibitor (mean 39 sec). Addition of inhibitor plasma had no effect on the prothrombin time of plasma from 6 patients with hemophilia B-. Complexes between factor IX and the human inhibitor could be demonstrated both before and after the coagulation with bovine thromboplastin. These complexes were demonstrated as a factor IX antigen with a reduced electrophoretic mobility in crossed immunoelectrophoresis against a rabbit antiserum to factor IX. The results indicate that normal factor IX loses the ability to act as an inhibitor in the coagulation with bovine thromboplastin after having formed a soluble complex with a homologous antibody.