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Reply

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Acquired von Willebrand's disease has recently been reported as a cause of bleeding in patients with myeloproliferative disorders [1, 2]. Of 3 reported patients 2 had bleeding complications after splenectomy and 1 after tooth extraction. Decreased ristocetin co-factor activity (vWF:RCo), normal von Willebrand factor antigen (vWF:Ag) and normal factor VIII pro-coagulant activity (VWF:c) were noted in all 3 patients. On sodium dodecyl sulfate agarose electrophoretic analysis of vWF multimeric composition in 2 of these patients [2], alteration of the repeating von Willebrand factor (vWF) multimeric 'triplet' and decrease or absence of the larger vWF multimers, a pattern consistent with type II A von Willebrand's disease, was noted. Due to normal platelet adhesive index, normal platelet aggregation in response to ristocetin (final concentration 1.5 mg/ml), absence of platelet aggregation in response to half-strength ristocetin (final concentration 0.75 mg/ml), factor VIII C level of 2.25 U/ml (normal 0.5-1.5 U/ml) and prompt cessation of bleeding after thrombocytapheresis, diagnosis of acquired von Willebrand's disease was not seriously considered in our patient [3]. However, we agree with Dr. Fey that the diagnosis of acquired von Willebrand's disease could not be excluded without further studies. To resolve this question, we have performed factor VIII-vWF-related studies on patient's plasma specimen drawn at the time of admission to the VA Hospital, Oakland. Plasma ristocetin cofactor activity (factor VIII:RCo) was 1.44 U/ml (normal 0.5-1.5 U/ml). Plasma von Willebrand factor antigen (vWF:Ag) measured by Laurell immunoelectrophoresis was 2.25 U/ml (normal 0.5-1.5 U/ml). Two-dimensional immunoelectrophoresis of plasma von Willebrand factor showed normal multimeric composition. These studies exclude the diagnosis of acquired von Willebrand disease as a cause of this patient's massive abdominal wall hematoma.

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