Correspondence

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Spontaneous Massive Abdominal Wall Hematoma in Polycythemia vera

M.F.

Fey

Berne

M.F. Fey, MD, Central Hematology Laboratory, Inselspital, CH-3010 Berne (Switzerland)

In their recently published case report, Mittal and Meisler [1] describe a patient known to have polycythemia vera who, without a history of trauma, developed a hematoma within the abdominal wall. Apparently, they did not discover any particular hemostatic defect to be held responsible for this unusually dramatic spontaneous hemorrhage, since they report a complete coagulation profile, the platelet adhesive index and aggregation studies to have been normal. Whereas the occurrence of qualitative platelet defects in myeloproliferative disorders (MPD) is discussed, the authors do not mention the possibility of acquired von Willebrand's disease in this particular clinical setting [2, 3]. During the last few years we have seen a number of patients with MPD at our department who at one time or another were found to have a mostly transient decrease of factor VIII/von Willebrand factor (VIII:WF) [2]. In a series of 10 patients with polycythemia vera, 5 subjects had a reduced factor VIII/ristocetin cofactor (VIII:RC), the value in 1 case being as low as 2.5% (normal range 50-150%), and a prolonged template bleeding time, while their platelet counts varied between 200 and 2,000-10V1. It is noteworthy that in cases where these assays were done repeatedly, von Willebrand factor activity showed marked fluctuations, sometimes being normal, sometimes very low in the same patient. Tests for specific antibodies directed against factor VIII:WF were negative. Mittal and Meisler [1] do not specify whether measurement of von Willebrand factor was included in the coagulation profile. If not, acquired von Willebrand's disease should have seriously been considered among the possible causes of that patient's bleeding disorder. Had it indeed been responsible for that threatening hemorrhage, substitution of factor VIII by cryoprecipitate would have been a more appropriate management than plateletpheresis. It is important to bear in mind that the finding of acquired von Willebrand's disease in MPD need not be a constant one. At our department we therefore recommend that, while planning surgical procedures in these patients (e.g. splenectomy), laboratory screening should include VIII:WF assays shortly before the operation. Likewise, any bleeding episode might be due to transient von Willebrand's disease, even if assays performed several weeks or months earlier had yielded normal results.

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