Acquired von Willebrand’s Disease
Associated with Gastrointestinal Angiodysplasia and Monoclonal Gammopathy

Sir,

Acquired von Willebrand’s Disease (AVWD) is a rare bleeding disorder, usually occurs in elderly patients. We would like to present case with recurrent gastrointestinal bleeding diagnosed to have AVWD with inhibitors.

A 63 years male presented with a history of recurrent episodes of lower gastrointestinal bleeding for three years. He had also a history of occasional epistaxis and skin bleeds for last 3 months. Repeated endoscopic gastrointestinal examinations demonstrated the presence of multiple angiodysplastic lesions in the descending and sigmoid colon. Family history revealed no members with known bleeding disorders. On examination, he was pale and there were few scattered generalized petechiae. Other physical findings were unremarkable. Hemogram showed Hb of 7.4gm /dl, TLC of 6.4 x 10^9/L, platelet count of 230 x 10^9/L. Peripheral smear examination showed microcytic hypochromic anemia with no abnormal cells. Initial coagulation screening revealed a normal prothrombin time (PT); a prolonged activated partial thromboplastin time (APTT) and a prolonged Ivy bleeding time. At presentation, FVIII procoagulant activity (20%), vWF antigen levels (22%), and ristocetin cofactor activity (13%) were markedly reduced. The ratio of VWF:RCo : VWF:Ag was less than 0.7. Multimeric analysis revealed loss of high molecular weight (HMWM) and intermediate molecular weight multimers. An in-vitro Inhibitor against vWF of the patient’s plasma was demonstrated. A final diagnosis of AVWD – subtype 2A with inhibitors to VWF was made. The patient’s bleeding episodes were initially managed with cryoprecipitate replacement and desmopressin therapy to which he showed an adequate response in terms of clinical situation and his haemostatic parameters. However most of these conditions are associated with protein C deficiency. Priapism is an unusual manifestation of AVWD, when possible, also leads to the resolution of bleeding expression of angiodysplasia.2 Successful treatment of the underlying pathology associated with AVWD, when possible, also leads to the resolution of the bleeding diathesis. Immunosuppressive therapy in the form of steroid or cyclophosphamide showed some response in significant number of cases.3 Intravenous immunoglobulins have also shown some efficacy in the management of AVWD, especially cases associated with monoclonal gammopathies.1 Awareness of AVWD is essential for diagnosis and appropriate management.

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REFERENCES


Priapism: an Unusual Manifestation of Warfarin-induced Skin Necrosis with Protein C Deficiency

Sir,

Warfarin-induced skin necrosis is a rare condition. However most of these conditions are associated with protein C deficiency. Priapism is an unusual manifestation of warfarin-induced skin necrosis.

A 46 years male was presented with a history of pain and swelling of the left lower limb for 1-week duration. Doppler ultrasonography showed thrombosis of left peroneal and femoral vein. Patient was managed with...