


**Hematological evaluation of acquired von Willebrand syndrome before oral surgery: Management of an unusual case**

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**Table 1: Results of coagulation blood tests of the patient with IgM-MGUS and AvWS**

Laboratory tests	Laboratory values	Normal range
Bleeding time (BT), Ivy method (min)	18	5-7
PT (ratio)	1.02	0.90-1.14
aPTT (ratio)	1.48	0.82-1.19
aPTT 1:1 mixing study (ratio)	1.02	0.82-1.19
Factor VIII:C (IU/dL)	25	62-156
vWF:Ag (IU/dL)	28	52-148
vWF:RCo (IU/dL)	5	49-151
RIPA (mg/mL)	1.03	0.70-1.20
vWF propeptide (IU/dL)	60	49-158
vWF (propeptide/Ag) ratio	2.14	0.7-1.9
vWF high molecular weight multimers in plasma	Undetectable	Detectable

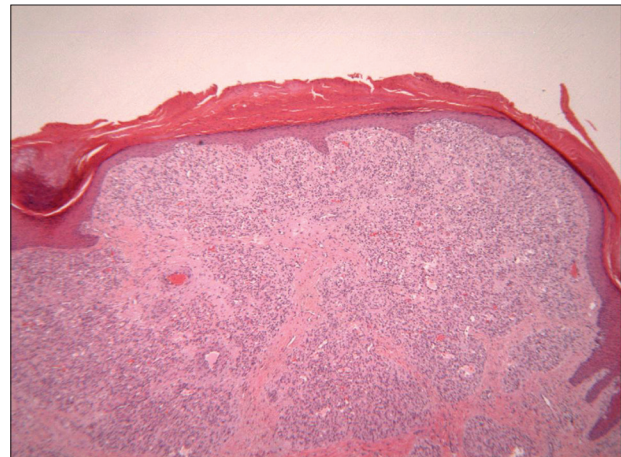
Sir,

Patients with monoclonal gammopathies of uncertain significance (MGUS) may develop an acquired bleeding disorder similar to congenital von Willebrand's disease (vWD), called acquired von Willebrand syndrome (AvWS).<sup>[1]</sup>

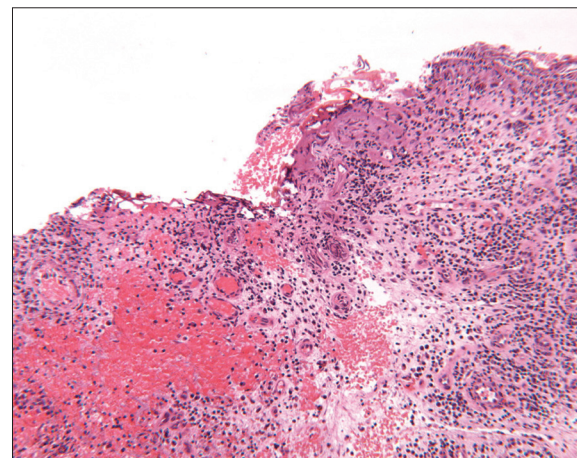
An 88-year-old male patient presented to our department for bleeding episodes from a nodule on the tongue caused by dental trauma. Blood tests showed a mild form of iron-deficiency anemia. Serum protein and immunofixation electrophoresis revealed a monoclonal IgM-κ component. Bence Jones proteinuria was undetectable. A diagnosis of IgM-MGUS was made by his hematologist. Liver and/or kidney diseases were absent. The patient was not taking any medications that could have altered blood coagulation. The family history of bleeding was negative. The coagulation tests are summarized in Table 1. The patient was referred to the Hemophilia and Thrombosis Center of the same hospital to characterize the vWD type. A diagnosis of vWD type 2A was made. In order to distinguish between an inherited vWD and an AvWS, a quantitative analysis of the plasma vWF propeptide (pro-vWF:Ag) was performed using ELISA technique. Normal plasma levels of pro-vWF:Ag were found together with an increased ratio of pro-vWF:Ag to vWF:Ag, indicating an accelerated clearance of vWF from blood circulation. These findings were suggestive of AvWS.<sup>[1]</sup>

Treatment with 1-deamino-8-D-arginine-vasopressin (DDAVP) which was administered intravenously at a dose of 0.3 µg/kg did not show a significant improvement in bleeding time (BT), plasma levels of factor VIII, and vWF. Thus, he received intravenous infusions of factor VIII/vWF concentrates (Haemate-P; Centeon, Marburg, Germany) at a daily dose of 40 IU/kg until the third postoperative day for an average of five administrations, achieving a shortened BT (from more than 18 min to 8 min) as well as transient correction of factor VIII and vWF plasma levels.

The oral lesion excision was carried out using a diode laser (wavelength 808 nm) set at 1.5 W in the pulsating wave mode. Oral rinses with 10 mL of 4.8% tranexamic acid solution were



**Figure 1:** Cytologically bland lobular capillary proliferation with variable cellularity. The overlying epithelium is flattened and forms a peripheral collarette (×4, H&E)



**Figure 2:** Pyogenic granuloma with thermal artifacts of laser irradiation, consisting of an outer layer of carbonization and an inner layer of coagulation with vessel sclerosis (×10, H&E)

administered four times a day for seven postoperative days.<sup>[2]</sup> Histological analysis reported a pyogenic granuloma [Figure 1].

As far as we know, no consensus exists on the therapeutic management of AvWS in relation to oral soft tissues surgery.<sup>[2,3]</sup> The unusual dyscrasia reported here highlights that some AvWS patients may be unresponsive to DDAVP. Thus, the replacement therapy with factor VIII/VWF concentrates remains, the sole option of treatment in AvWS patients with IgM-MGUS, although this will produce transitory effects<sup>[4]</sup> as was the case in our patient. The photochemistry affinity of the diode lasers to hemoglobin can further help to achieve clot stabilization, by promoting the formation of induced sclerosis laser on the vascular network of the chorion [Figure 2].

Dentists may have a role in secondary prevention through the detection of latent AvWS, because they are able to manage minor bleeding problems – often encountered in their daily practice – during oral surgery among dental patients.

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