Bilateral peripheral retinal hemorrhaging secondary to undiagnosed von Willebrand disease

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INTRODUCTION

Von Willebrand disease, an inherited clotting disorder, is caused by either a deficiency or mutation in the von Willebrand factor protein. Three types exist with type 1 being the mildest. Symptoms are directly related to the clotting deficiencies and can include frequent nosebleeds, easy bruising, and heavy periods in women. However, many patients can be asymptomatic and therefore the condition is likely under diagnosed. We report a rare case of bilateral peripheral retinal hemorrhaging secondary to undiagnosed von Willebrand disease.

CASE HISTORY

A 57-year-old black female presented for a comprehensive eye exam. Her medical history included hypertension and type two diabetes with all conditions being controlled with medications. Her last HbA1C was 5.7 with her blood pressure being 135/75 at examination. Best corrected visual acuity was 20/20 OD and OS. External examination, entrance testing, and slit lamp examination findings were unremarkable. IOP's were 16 mmHg OD/OS with Goldmann applanation tonometry. Dilated fundus examination revealed optic discs with 0.25/0.25 cup to disc ratio with clear maculas OD and OS. Peripheral retinal examination revealed scattered pinpoint hemorrhaging and micro aneurysms more so in the temporal quadrants of each eye. Based on the bilateral nature of the hemorrhaging and controlled blood sugar levels the patient was referred for extensive blood work including but not limited to: complete blood count with differentials, sickle cell, ESR, CRP, PT, PTT, INR, and auto immune testing. Lab testing was all unremarkable except for low levels of von Willebrand factor. Hematology consultation confirmed the diagnosis of type 1 von Willebrand disease. Consultation with a retina specialist and subsequent fluorescein angiography was unremarkable. As the patient was asymptomatic with no other retinal findings present, she continues to be monitored on a semi-annual basis.

DISCUSSION

Although rare, von Willebrand retinopathy, secondary to decreased von Willebrand factor, ultimately leads to blood vessel destabilization which can manifest as retinal hemorrhaging. Only a handful of cases have been described in the literature. Case reports have shown retinopathy presenting as vitreal, intraretinal, and subretinal hemorrhages with one case revealing retinal neovascularization. This case demonstrates the importance of clinicians considering von Willebrand disease as a cause of retinal hemorrhaging especially when patients present with an unremarkable medical history and/or controlled systemic conditions. As many patients are asymptomatic, appropriate testing is vital for diagnosis.

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FIGURE 1
Optos widefield photo OD showing scattered pinpoint hemorrhaging temporally

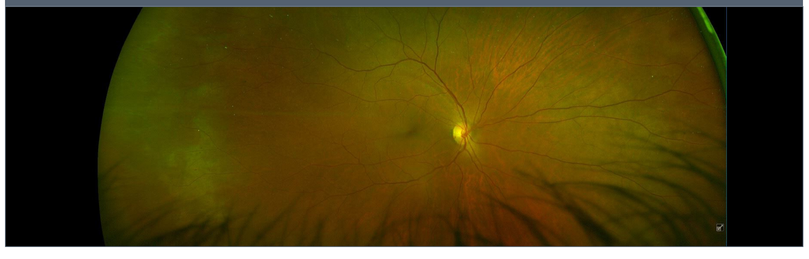


FIGURE 2
Optos widefield red free photo OD showing scattered pinpoint hemorrhaging temporally

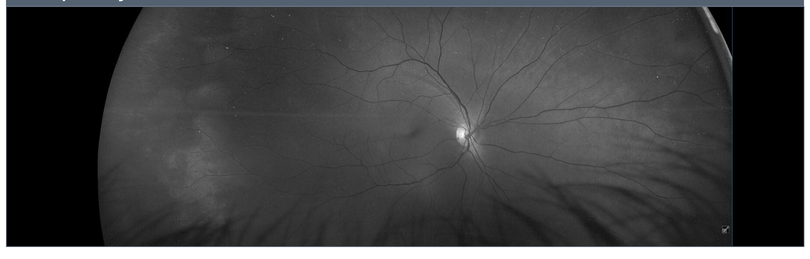


FIGURE 3
Optos widefield photo OS showing scattered pinpoint hemorrhaging temporally

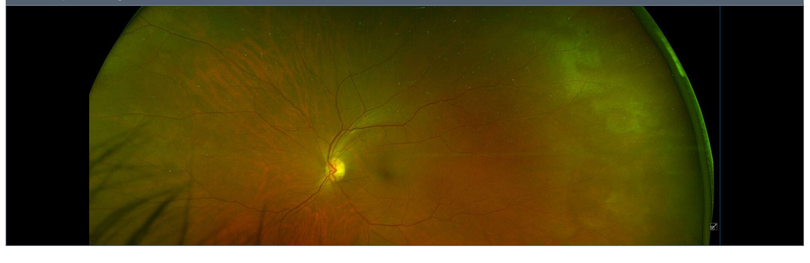


FIGURE 4
Optos widefield red free photo OS showing scattered pinpoint hemorrhaging temporally

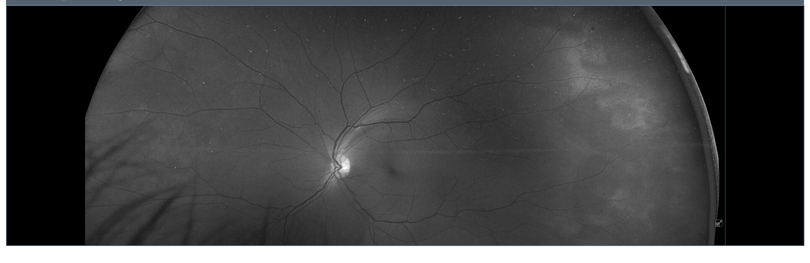


FIGURE 5
FA OD showing micro aneurysms in the temporal periphery with no obvious neovascularization

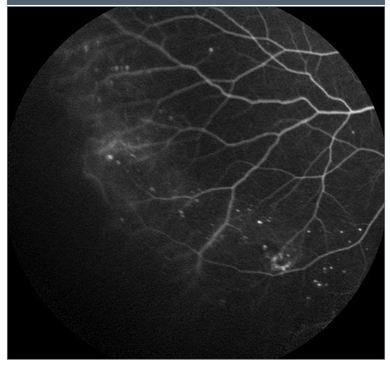
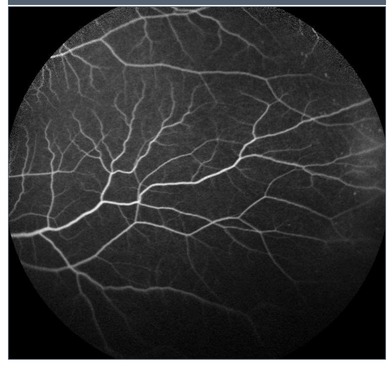


FIGURE 6
FA OS showing micro aneurysms in the temporal periphery with no obvious neovascularization



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