



Large Renal Arteriovenous Malformation in a Patient With von Willebrand Disease

Charles Schlaepfer, Jonathan Weese, Daniel Picus, and Ramakrishna Venkatesh

We describe a rare case of a large renal arteriovenous malformation in a patient with von Willebrand disease. Initial attempts at technically challenging embolization failed requiring a nephrectomy. Extra-intestinal vascular malformations are rare in von Willebrand disease. However, there is more recent evidence of von Willebrand factor's regulatory role in angiogenesis and vascular malformations. *UROLOGY* 130: 211–212, 2019. © 2019 Elsevier Inc.

A 63-year-old white male with von Willebrand disease type 2b was consulted for atrial fibrillation with remote history of gross hematuria and cardiac failure. A large right renal arteriovenous malformation (AVM) (11 × 7 cm) was incidentally discovered at cardiac computed tomography imaging and confirmed by abdominal computed tomography angiography (Fig. 1). The patient lacked traditional risk factors for renal AVM, but aberrations of von Willebrand factor are implicated in endothelial cell and angiogenesis deregulation.¹⁻³ Decreased Von Willebrand factor is associated with gastrointestinal angiodysplasias and rarely with splenic, lung, and soft tissue AVMs.^{4,5} Associated renal

AVM has not been reported. Twenty-two and 20-mm AMPLATZER vascular plugs (AGA Medical, North Plymouth, MN) as well as 8,10,12,15, and 20 mm Nester embolization coils (Cook, Inc, Bloomington, IN) and Ruby packing coils (IPenumbra, Alameda, CA) failed to fully occlude the AVM. A 22-mm AMPLATZER vascular plug was placed in the renal vein to prevent migration of arterial embolization coils into inferior vena cava (Fig. 2 and Supplemental Video 1 and 2). Open nephrectomy and AVM excision succeeded with infusion of human factor VIII complex (Fig. 3). The patient was discharged without complications. His serum creatinine was 1.6 mg/dL at 6 months follow-up.

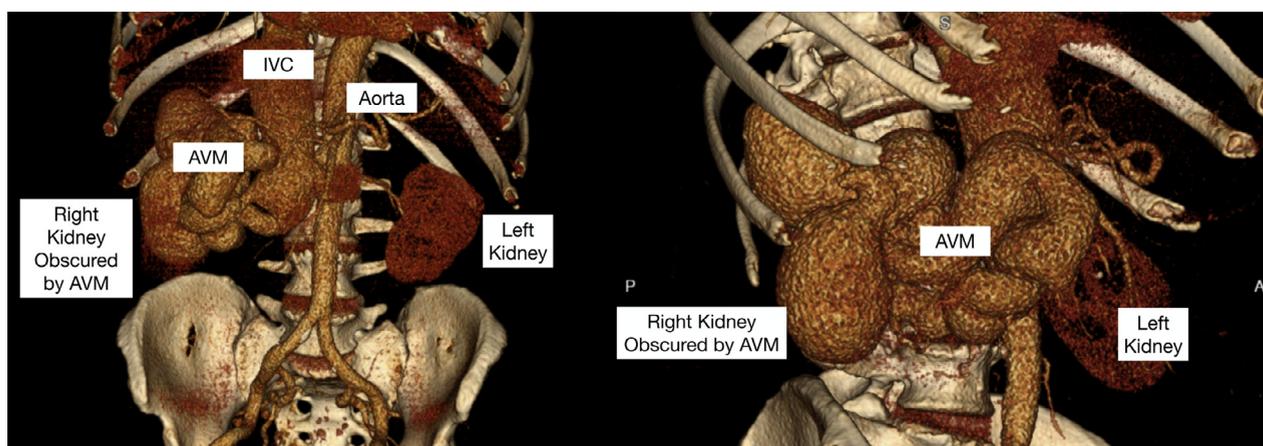


Figure 1. Three-dimensional reconstruction of arteriovenous malformation of right kidney in anterior-posterior (left panel) and oblique (right panel) views.

Declaration of Interests: CS: None; JW: None; RV: Consultant for Retrophin Inc.; DP: None.

Financial Disclosure: There are no financial disclosures for this manuscript.

From the Division of Urologic Surgery, Washington University in St. Louis School of Medicine, Saint Louis, MO; and the Division of Diagnostic Radiology, Interventional

Radiology Section, Washington University in St. Louis School of Medicine, Saint Louis, MO

Address correspondence to: Charles Schlaepfer, B.A., 4960 Childrens Place, Campus Box 8242, Saint Louis, MO 63110. E-mail: cschlaepfer@wustl.edu

Submitted: March 25, 2019, accepted (with revisions): April 18, 2019

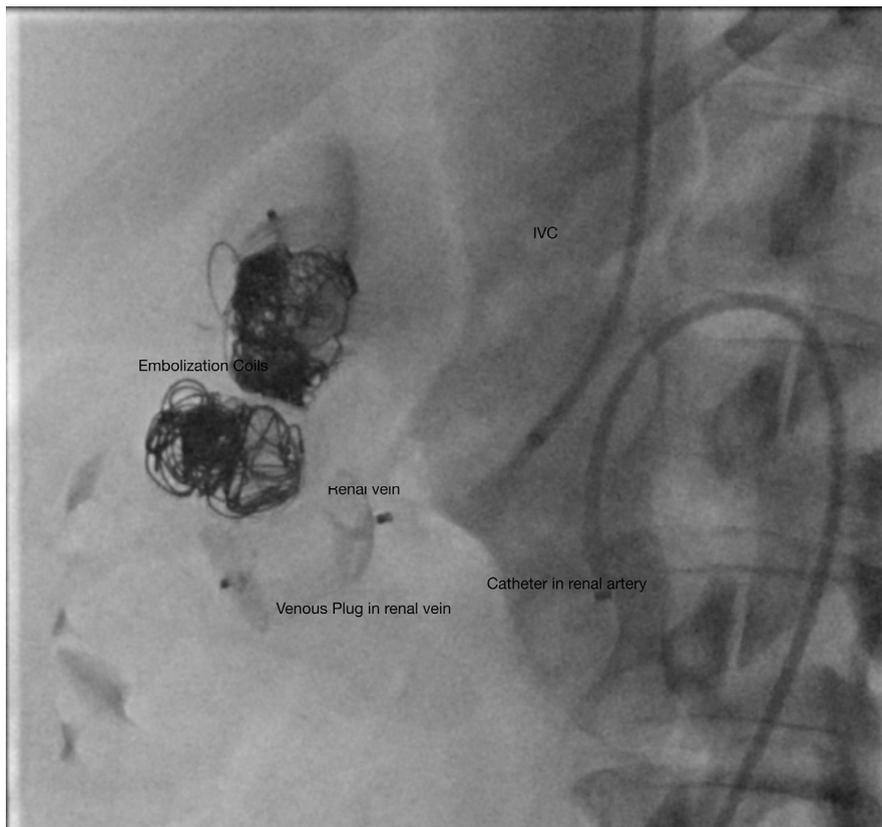


Figure 2. Embolization coils and vascular plugs in the right main renal artery and vein, respectively.

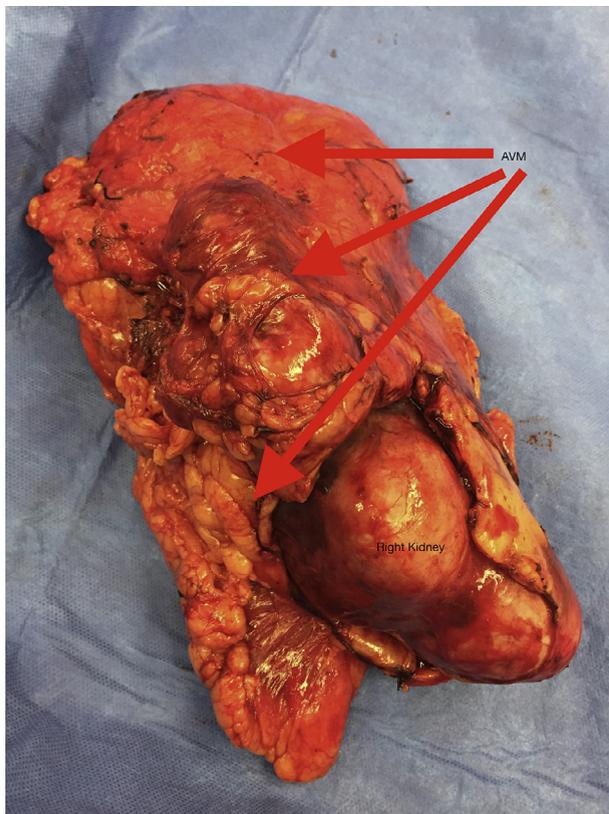


Figure 3. Final specimen of right kidney and arteriovenous malformation.

SUPPLEMENTARY MATERIALS

Supplementary material associated with this article can be found in the online version at <https://doi.org/10.1016/j.urology.2019.04.024>.

References

1. Randi AM, Smith KE, Castaman G. von Willebrand factor regulation of blood vessel formation. *Blood*. 2018;132:132–140.
2. van Vulpen LFD, Keeling D, Makris M. von Willebrand disease and extra-intestinal angiodysplasia. *Haemophilia*. 2017;23:e354–e355.
3. Starke RD, Ferraro F, Paschalaki KE, et al. Endothelial von Willebrand factor regulates angiogenesis. *Blood*. 2011;117:1071–1080.
4. Chey WD, Hasler WL, Bockenstedt PL. Angiodysplasia and von Willebrand's disease type IIB treated with estrogen/progesterone therapy. *Am J Hematol*. 1992;41:276–279.
5. Franchini M, Mannucci PM. Gastrointestinal angiodysplasia and bleeding in von Willebrand disease. *Thromb Haemost*. 2014;112:427–431.