Disseminated microinfarctions of the right kidney requiring nephrectomy: an unusual complication of acquired hemophilia A

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We report a rare case of acquired hemophilia A (AHA), a severe bleeding diathesis caused by autoantibodies inactivating coagulation factor VIII (FVIII).¹,² A 50-year-old woman was admitted to the Urology Department due to macroscopic hematuria and intense lumbar pain. The patient underwent ureterorenoscopy to reduce renal pelvis enlargement, followed by laparotomy because of right ureteral stenosis. Catheterization of the right kidney with a double-J catheter was performed with ureteral anastomosis and retroperitoneal drainage (figure 1A and 1B). Histopathological examination showed chronic ureteritis with fibrosis. Due to persistent lumbar pain, progressive drain bleeding, and transfusion-dependent anemia, the patient was referred for computed tomography (CT), which revealed a large retroperitoneal hematoma (size, 18 × 5 cm) and right-sided hydronephrosis grade 2. Another laparotomy was performed, and 1500 ml of blood was evacuated. However, the patient’s condition did not improve, and she still required regular blood transfusions. Three days after the laparotomy, the pain and swelling worsened and a repeated CT showed a hematoma surrounding the right kidney and moving it forward, lack of contrast exertion, as well as edematous and ischemic lesions along with disseminated microinfarctions (figure 1C and 1D). The left kidney was unchanged. Nephroureterectomy was performed. Histopathological examination confirmed multiple necrotic and hemorrhagic kidney lesions (figure 1E–1H). Owing to prolonged activated partial thromboplastin
time, coagulation tests were scheduled and revealed significant coagulation FVIII deficiency (7%) and the presence of FVIII inhibitor (2 Bethesda units [BU]). Treatment with recombinant activated factor VII (rVIIa) (100 µg/kg every 3 hours) and prednisone (1 mg/kg/d) was initiated. Despite continued rVIIa treatment, extensive soft tissue bleeds occurred, successfully treated with a sequential alternate administration of rVIIa and activated prothrombin complex concentrate.

The patient was discharged home on low-dose prednisone but was readmitted after 5 weeks due to joint and muscle bleeding in the calf and hand, treated with rVIIa. Apart from prednisone, she was treated with rituximab and eventually with the Budapest protocol (cyclophosphamide, methyprednisolone, FVIII concentrate). As a result, her bleeding symptoms resolved but she did not achieve remission (maximal FVIII activity was 12%, and the inhibitor titer increased to 13 BU). Renal function parameters returned to normal.

No underlying condition was found that could explain the development of FVIII autoantibodies. Therefore, AHA was considered idiopathic.

Gross hematuria is a very unusual presentation of AHA, with single cases reported in the literature. None of those cases required nephrectomy or report similar histopathologic lesions as described above. Our report emphasizes the need to consider acquired bleeding disorders in the differential diagnosis of hematuria, even in the presence of renal pathology, especially if other bleeding symptoms occur or abnormalities in a coagulation screening test are discovered. Urinary tract bleeding in AHA can lead to significant morbidity.

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