CLINICAL IMAGE

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

Hubert Burdziak[1], Joanna Zdziarska[2], Karol Burdziak[3], Monika Tomaka[4]

1 Urology Department, St. Father Pio Hospital in Przemyśl, Przemyśl, Poland
2 Hematology Department, The University Hospital in Krakow, Kraków, Poland
3 1st Faculty of Medicine, Medical University of Warsaw, Warsaw, Poland
4 2nd Faculty of Medicine, Medical University of Lublin, Lublin, Poland

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. Histopathologic examination confirmed multiple necrotic and hemorrhagic kidney lesions (FIGURE 1E–1H). Owing to prolonged activated partial thromboplastin

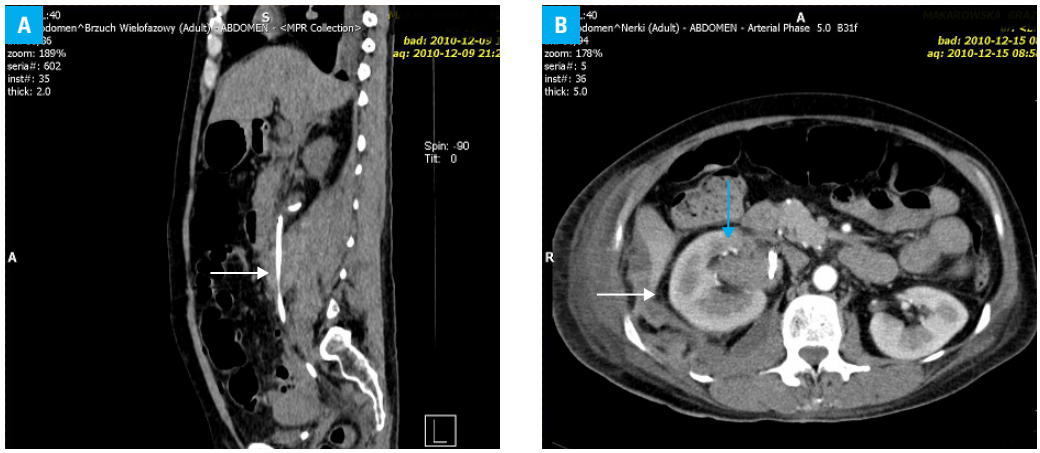

FIGURE 1 A – computed tomography scan showing a double-J catheter inside the right ureter (arrow) and renal pelvis; hematoma of the retroperitoneal space; B – computed tomography scan showing right kidney dislocation, fresh hematoma (size, 28 × 30 × 53 mm) surrounding the right ureter, disseminated microinfarctions in right kidney parenchyma, hematoma around renal fascia (white arrow), and narrowed intrarenal arteries (blue arrow)

Correspondence to:
Hubert Burdziak, MD,
Urology Department, St. Father Pio Hospital in Przemyśl, ul. Monte Cassino 18; 37-700 Przemyśl, Poland,
phone: +48 16 677 51 39,
email: hubert.burdziak@gmail.com
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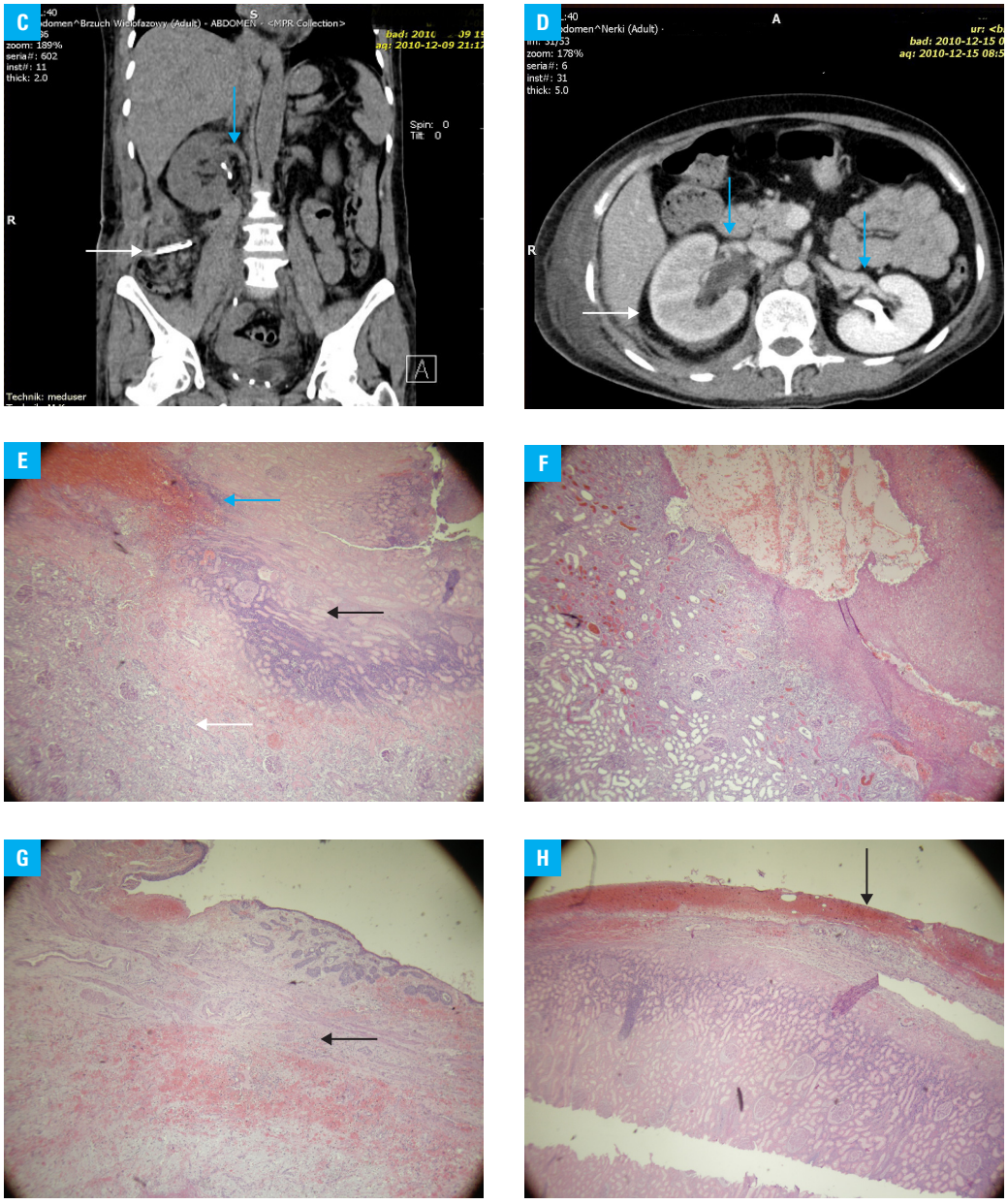

FIGURE 1 **C** – computed tomography scan showing a double-J catheter inside the right renal pelvis (blue arrow) and the bladder; hematoma and the drain of the retroperitoneal space (white arrow); **D** – narrowed intrarenal arteries of the right kidney and intrarenal vein sinuses of the right renal vein compared with the unchanged blood vessels of the left kidney (blue arrows), also seen in FIGURE 1B; segmental, wedge-shaped subcapsular, relatively hypodense zones after intravenous contrast administration (white arrow); **E** – hemorrhagic zone of the kidney cortex (blue arrow), the necrotic zone of the kidney cortex (black arrow), and normal kidney cortex (white arrow); **F** – necrotic zone of the kidney cortex; **G** – hemorrhagic zone of the renal pelvis wall (arrow); **H** – hemorrhagic surface of the fibrous capsule of the kidney (arrow)

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, methylprednisolone, 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.[3-5] 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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