55-year-old man presented with flaccid paraparesis. Immunologic and serologic tests were unrevealing. Nuclear magnetic resonance revealed brain and spinal ischemic lesions suggestive of isolated central nervous system vasculitis. Treatment with cyclophosphamide and steroids was initiated, resulting in clinical improvement. However, 1 month later, the patient acutely developed altered mental status associated with respiratory distress. Laboratory testing showed acute renal failure and elevated serum lactate dehydrogenase levels. Renal biopsy showed an interstitium expanded by a large atypical CD20⁺ cell proliferation. These cells had finely granular chromatin and visible nucleoli (see panels A–E; stains: [A,C,E] hematoxylin and eosin, [B,D,F] peroxidase; original magnification: [A] ×100, [B] ×40, [C-F] ×400), and were frequently observed in the peritubular capillary lumens, as demonstrated with endothelial CD34 staining (see panel F). Therefore, the diagnosis of intravascular large B-cell lymphoma was made. A bone marrow biopsy demonstrated lymphoma infiltration, but atypical cells were not found in peripheral blood smear or bone marrow aspirate. The patient was treated with cyclophosphamide, hydroxydaunorubicin, vincristine, and prednisone and achieved remission.

Intravascular large B-cell lymphoma is a rare disease that may affect any tissue or organ, with no alteration of the cells seen in a peripheral blood smear and difficulty in diagnosis due to the lack of an obvious tumor.
Intravascular large B-cell lymphoma in a kidney biopsy

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