SIGNIFICANCE STATEMENT

Anti-brush border antibody (ABBA) disease is a rare disorder associated with acute kidney injury and tubulointerstitial damage in which circulating antibodies reactive with kidney proximal tubular brush border are found. IgG-containing deposits are often found in the tubular basement membranes as well as segmentally in the glomerular subepithelial space. This paper presents a detailed case series of 10 such patients, all above the age of 65, and identifies the target of the circulating ABBA autoantibodies as LDL receptor-related protein 2 (LRP2), also known as megalin. Detection of anti-LRP2 antibodies in elderly patients with unexplained acute kidney injury may aid in diagnosis and may lead to the consideration of immunosuppressive therapy to forestall further renal deterioration.