SIGNIFICANCE STATEMENT

Fibrillary glomerulonephritis (FGN) is a primary glomerular disease with a poor prognosis. Currently FGN poses substantial diagnostic challenges, in part because there are no specific histological biomarkers. This manuscript describes the discovery, using proteomics, of a new potential biomarker, DNAJB9 (DnaJ Heat Shock Protein Family [Hsp40] Member B9). We demonstrate that DNAJB9 is present in overabundance in FGN glomeruli, but not in glomeruli from patients with other glomerular diseases or from healthy subjects. DNAJB9 is potentially a useful diagnostic marker for FGN. Study of its function may provide important clues to the underlying pathogenesis of this disease.