Nephrotic Syndrome in Two Cases with Sickle Cell Nephropathy

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Dear Sir,

Sickle cell anemia (SCA) is associated with various renal functional and morphological changes and these alterations can be considered together under the term of sickle cell nephropathy (SCN) [1, 2]. Proteinuria and glomerular lesions of SCN have not been established as well as renal tubular lesions in SCA [2]. The etiology of the glomerulopathy in SCA is uncertain. The first case report of SCA with nephrotic syndrome was published by Berman and Schreiner [3] in 1958. After that, mesangial proliferation, membranoproliferative glomerulonephritis (MPGN), and focal and segmental glomerulosclerosis were described in patients with SCA [4]. We present 2 cases of SCA-associated nephrotic syndrome who have no recent history of infection, toxin or drugs or other systemic diseases.

Case 1: An 18-year-old male SCA patient was admitted with a history of abdominal fullness and swollen ankles. His physical examination showed hepatomegaly, ascites and periorbital and pretibial edema. On his laboratory analysis, hematocrit (Hct) was 22%, serum total protein 6.2 g/dl, albumin 2.5 g/dl, cholesterol 206 mg/dl, HDL 46 mg/dl, triglycerides 267 mg/dl, AST 21 IU/l, ALT 12 IU/l, alkaline phosphatase 217 IU/l, BUN 50 mg/dl, creatinine 4.3 mg/dl. Daily proteinuria was 9.2 g. Antinuclear antibody (ANA) and rheumatoid factor (RF) were negative, and C3 level was normal. Hepatitis B virus (HBV) markers and hepatitis C virus (HCV) antibody were not detected. Renal biopsy revealed endo- and extracapillary proliferative glomerulonephritis. (GN) Hemosiderin granules were present in the tubular epithelium. On immunofluorescence microscopy (IF), capillary wall and mesangial deposits of IgM were identified.

Case 2: A 29-year-old male SCA patient presented malaise, fascial and ankle swelling, abdominal distension, and shortness of breath. His physical examination was normal except for anemia and pretibial edema. His proteinuria was 10.5 g/day. Laboratory investigation: Hct was 20%, serum total protein 7.2 g/dl, albumin 2.9 g/dl, cholesterol 226 mg/dl, HDL 63 mg/dl, LDL 130 mg/dl, triglycerides 163 mg/dl, AST 35 IU/l, ALT 19 IU/l, ALP 205 IU/l, BUN 45 mg/dl, creatinine 2.5 mg/dl, creatinine clearance 26 ml/min. ANA and RF were negative, C3 was found in normal values. HBV markers and HCV antibody were not detected.
in serum. Renal biopsy showed MPGN. IF revealed glomeruli basement membrane and interstitial capillary wall deposits of IgG, IgM and IgA.

SCA-associated glomerulopathy with immunocomplex deposition has been reported [5,6]. McCoy [7] and Bakir et al. [8] reported that in SCA-associated GN, immunocomplex deposition was uncommon. Iskandar et al. [4] reported 2 cases with SCA GN and found diffuse global capillary wall and mesangial deposits of IgG, IgA, IgM, C3 and C1q. Review of the literature has identified fewer cases of SCA-associated GN with demonstrated immunocomplex deposition. We did not measure antibodies to renal tubular antigens. HBV and HCV infection or other infections were not found. In the histories of our patients, drugs or toxin were not identified. We can say that SCA-associated GN of our patients is possibly immunocomplex-mediated.

References