ABSTRACT

Sickle cell disease (SCD), a genetically inherited disease, often presents with disabling acute complications which can occasionally be fatal. Improvement in the management of SCD has allowed many patients to survive into adulthood, which nevertheless is marked by occurrence of end-organ damage including kidney dysfunction. This study determined the prevalence and pattern of CKD in adult SCD patients and also investigated the histopathological changes in sickle cell nephropathy. It also assessed the relationship between kidney function, renal histopathology and haematologic parameters.

The study prospectively screened 70 patients with SCD for the presence of proteinuria (microalbuminuria or overt proteinuria), glomerular filtration rate (GFR) <60ml/min and tubular dysfunction and renal biopsy was performed in those with indications. Data was analysed using descriptive and inferential statistics in an SPSS package (version 16).

The age of the patients ranged between 18 and 56 years (Mean±SD; 27.5±8.9years) with a female preponderance, M: F ratio (1:1.3). Of the 70 patients screened, 25 (35.7%) had CKD as defined by GFR <60ml/min and/or proteinuria for 3 months. CKD was common among HbSS (42.1%) than HbSC (7.7%) subjects. The markers of kidney dysfunction found were 23 (32.9%) patients had GFR <60ml/min and 5 (7.1%) had hyperfiltration GFR >120ml/min; overt proteinuria was found in 4 (5.7%) while microalbuminuria was found in 12 (17.1%). GFR correlated positively with haematocrit (r=0.472; p<0.0001) and BMI (r=0.518; p<0.0001) while microalbuminuria correlated negatively with GFR (r =-0.255; p=0.04).
Tubular function were assessed by fractional excretion of sodium (FE_{Na}), fractional excretion of potassium (FE_{K}), specific gravity and pH; mean (±SD) of (FE_{Na}), (FE_{K}), specific gravity and pH were 6.5 (±2.2), 31.1 (±10.1), 1.0 (±0.01) and 6.3 (±0.5) respectively. All recruited patients had markedly elevated FE_{K} while 98.6% had elevated FE_{Na}. Hyposthenuria was present in 85.7%. GFR correlated negatively with percentage sickle cell count(r=-0.616, p<0.0001), FE_{K}(r=-0.448, p<0.0001) and FE_{Na}(r=-0.336; p=0.004). Percentage sickle cell count correlated most significantly than the other parameters measured and may be an independent risk factor for CKD in SCD patients.

Of the 25 patients with CKD, 23 underwent renal biopsy out of which 22 were successive and the remaining 2 declined. Glomerulosclerosis was demonstrated in 9 (39.1%) of the patients, 6(69.6%) had matrix expansion while 15 (65.2%) had mesangial hypercellularity. Tubular thickening was found in 7(30.4%) patients while tubular atrophy was demonstrated in 14(60.9%) of the patients. Interstitial fibrosis was demonstrated in 14(60.9%) patients while interstitial cellular infiltration with inflammatory cells was demonstrated in 2(8.7%) of the patients. Medial arteriolar thickening was seen in 4 (17.4%) patients while intimal fibrosis was found in 7 (30.4%) of the patients.

Pattern of histological diagnosis were mesangioproliferative glomerulonephritis in 11 (50%) patients, 6 (27.3%) had minimal change disease, 3 (13.6%) had focal segmental glomerulosclerosis while interstitial nephritis was diagnosed in 2 (9.1%) patients.

In conclusion, CKD is common among SCD patients. It is more prevalent in homozygous SS than heterozygous SC patients. Kidney disease was characterized by a preponderance of tubular dysfunction and mesangioproliferative glomerulonephritis.