Co-existing autosomal dominant polycystic kidney disease and nephrotic syndrome in a Nigerian patient with lupus nephritis

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Abstract

A little over 30 cases on co-existing nephrotic syndrome and autosomal dominant polycystic kidney disease (ADPKD) have been reported from different regions of the world since 1957. We present a case report on co-existence of nephrotic syndrome (secondary to lupus nephritis) with ADPKD in a 24-year-old woman from Nigeria. She was positive for anti-double stranded DNA. Renal histology showed International Society of Nephrology/Renal Pathology Society Class II lupus nephritis. The co-existence of nephrotic syndrome and ADPKD may have been overlooked in Africa in the past. There is a need to screen for nephrotic syndrome in patients with ADPKD among clinicians in the African setting.

Keywords: Africa, autosomal dominant polycystic kidney disease, lupus nephritis, nephrotic syndrome, renal biopsy

Introduction

Lupus nephritis is an autoimmune glomerulopathy which ranks high among the secondary causes of nephrotic syndrome besides amyloidosis, diabetes mellitus, preeclampsia, human immunodeficiency
Akinbodewa, et al.: Rare co-occurrence of ADPKD and lupus nephritis

Virus-associated nephropathy, and hepatitis-B virus,\(^1\) autosomal dominant polycystic kidney disease (ADPKD) is the most prevalent inheritable kidney disorder globally. It is usually associated with proteinuria < 1 g/day.\(^2\)

There are only about 32 case reports on the co-existence of nephrotic syndrome and ADPKD globally since 1957 to date but none from Africa;\(^3\) hence, our interest to report this case.

**Case Report**

A 24-year-old female patient presented to the renal outpatient in September 2014 on account of body swelling and frothy urine while being managed for periodontal gingival cellulitis. She had facial puffiness, jaw swelling, and massive lower limb edema and ascites. The blood pressure was 120/80 mmHg. In the 3rd week of admission, she was observed to develop hyperpigmented malar rash, gum hypertrophy, and bleeding with oliguria and rising serum creatinine levels. Her mother died of a kidney-related illness of unspecified cause.

At admission, her serum creatinine was 167.3 umol/l with an estimated glomerular filtration rate (eGFR) of 42 ml/min/1.73m\(^2\) (Modification of Diet in Renal Disease equation). Her hemogram showed leukopenia and anemia of 1.7 × 10\(^9\)/L (normal range, 4–11 × 10\(^9\)/L) and 18% (normal range, 33–36% by Kidney Disease Outcome Quality Initiative), respectively; the erythrocyte sedimentation rate (ESR) was 133 mm in the 1st h (Westergren).

She had reduced level of high-density lipoprotein cholesterol of 0.8 mmol/L (reference range: 1.2–2.0 mmol/L female) and elevated triglyceride of 5.8 mmol/L (reference range: 0.1–1.7 mmol/L), severe hypoalbuminemia (17.2 g/L), and heavy proteinuria (2.7 g/day) which prompted suspicion of nephrotic syndrome. Spun urine sample showed 15–20 pus cells/hpf, numerous dysmorphic red blood cells, granular casts, and epithelial cell casts. The urine culture was sterile.

Renal scan [Figures 1 and 2] showed enlarged kidneys with multiple cysts of varying sizes (2.7–31.4 mm). Other intra-abdominal organs were normal.

Serology showed a strong positive homogenous pattern for the antinuclear factor while the anti-double stranded DNA titer was elevated to 30 IU/ml (normal < 4 IU/ml) by ELISA was demonstrated. Renal tissue histology [Figure 3] on light microscopy showed marked mesangial cell increase, some of which contain apoptotic debris; significant features of acute tubular necrosis with simplification of tubular epithelial cells, some of
which are cystically dilated containing amorphous eosinophilic material; and interstitial fibrosis with tubular atrophy in about 10–20% of the section examined. There was no increase in the mesangial matrix and thickening or duplication of the glomerular basement membrane. Features were in keeping with lupus glomerulonephritis (International Society of Nephrology/Renal Pathology Society [ISN/RPS] Class II).

She underwent a series of dialysis sessions when she became oliguric and uremic during admission and received intravenous methyl prednisolone (1 g daily for 3 days) followed up with tapered doses of oral prednisolone and mycophenolate mofetil (Myfortic) 720 mg bid. After 6 months of treatment, proteinuria, serum creatinine, and ESR reduced to 0.2 g/day, 94 umol/l and 44 mm/h, respectively. Her eGFR rose to 82 ml/min/1.73 m²

At 10 months, she remains stable on medications with proteinuria of 0.07 mg/day and ESR of 16 mm in 1 h (Westergren) and hemoglobin concentration of 12 g/dl.

**Discussion**

It is unusual to find nephrotic syndrome and ADPKD co-existing. To date, the association between these disorders remains unexplained and is generally considered to be rare and at best, coincidental.

Our electronic literature search indicates that there has not been any previous case report in Africa; most reported cases have originated from the United States, Europe, and Asia since the first documented description of nephrotic range proteinuria in ADPKD by Dalgaard in 1957.[9]

The first two cases of lupus nephritis induced nephrotic syndrome co-existing with ADPKD were reported by Wan et al. in 2009 in a 49-year-old female with WHO Class IV lupus nephritis and Park et al. in 2012 in a 48-year-old woman with WHO Class IV lupus nephritis, respectively.[4-10]

In our index, a patient from Ilesha, South West of Nigeria, who is also female, the age at presentation was much younger at 24 years while the histology grade was ISN/RPS II lupus nephritis on light microscopy. Facilities for immunofluorescence study and electron microscopy of the renal tissue were not readily available in our environment. Our report also appears to be the third case of nephrotic syndrome secondary to lupus nephritis co-existing with ADPKD to be reported.

It is remarkable that there may be an increasing incidence report of lupus nephritis induced nephrotic syndrome in patients with co-existing ADPKD in recent times in contrast to earlier histological patterns.[9] Since Wan et al. in 2009, eight cases have been reported (including this report). Among them, three renal histology (37.5%) showed lupus nephritis while others showed each of idiopathic membranous glomerulonephritis, diffuse proliferative glomerulonephritis, amyloidosis, mesangiproliferative glomerulonephritis, and focal segmental glomerulosclerosis.[4-10]

Our patient recovered fully after a 6 months course of mycophenolate mofetil and corticosteroid hence, indicating a need for the exclusion of co-existing glomerulonephritis in patients with ADPKD.

One may assume that the report on the co-existence of nephrotic syndrome and ADPKD in Africa is low because it may have been overlooked by our clinicians. There is therefore a need to lay emphasis on routine investigations for identification of nephrotic range proteinuria and other features of nephrotic syndrome among patients who present to our clinics with a diagnosis of autosomal polycystic kidney disease.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**References**


