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Case Report

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

Résumé

Une peu plus de trente cas de la coexistence du le syndrome néphrotique et maladie polyktose rénale autosomique dominante (ADPKD) a été reporté dans divers regions du monde depuis 1957. Nous presentons un cas de la coexistence du syndrome néphrotique (secondaire à néphrite lupique) avec l'ADPKD sur une femme de 24 ans au Nigeria. Elle était positif pour ADN double brin anti. L'histologie rénale a presenté ce cas a la Société Internationale de Néphrologie/Société Rénale de Pathologie Classe II néphrite lupique. La coexistence du syndrome néphrotique et l'ADPKD peut avoir été négligé en Afrique dans le passé. Il existe un besoin pour cribler le syndrome néphrotique chez les patients du ADPKD parmi le Cliniciens dans le contexte Africain.

Mots-clés: Afrique, maladie polyktose rénale autosomique dominante, néphrite lupique, le syndrome néphrotique, biopsie rénale

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Introduction

Lupus nephritis is an autoimmune glomerulopathy which ranks high among the secondary causes of nephrotic syndrome besides amyloidosis, diabetes mellitus, preeclampsia, human immunodeficiency

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;

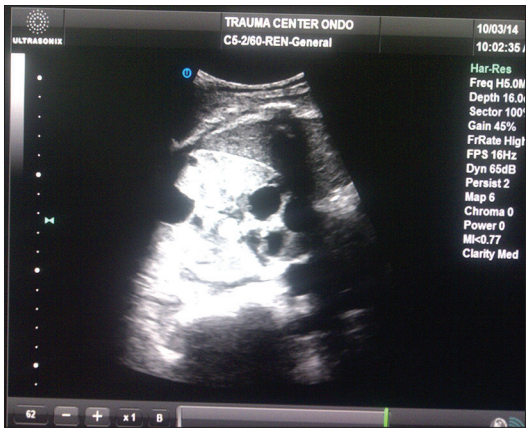

Figure 1: Two-dimensional ultrasound picture showing multiple cysts in the kidney. Only a minute portion of the lower pole was spared of renal tissue for kidney biopsy by ultrasound guidance

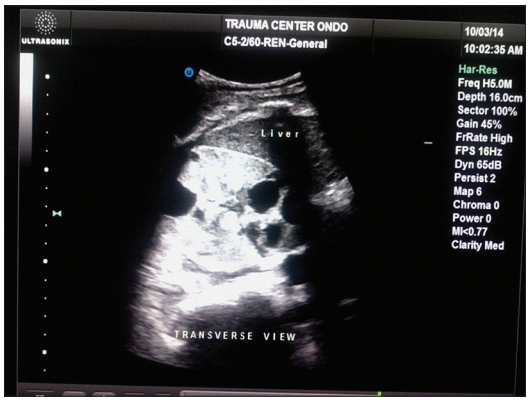

Figure 2: Two-dimensional ultrasonic transverse view of the right kidney demonstrating multiple renal cysts. The liver appears to be devoid of cysts

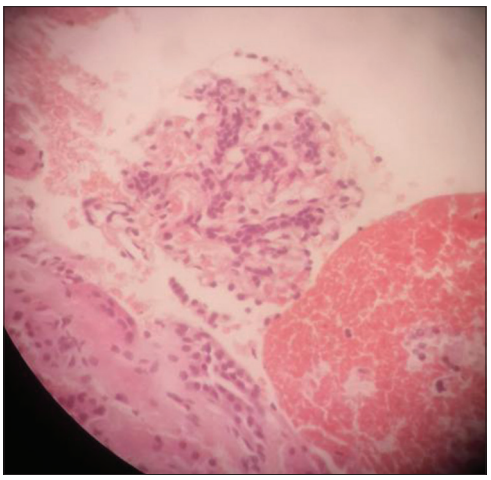

Figure 3: Renal histology. Photomicrograph showing two glomeruli, one totally sclerotic. There is marked mesangial cell increase and apoptotic debris with simplification of tubular epithelial cells, some of which are cystically dilated containing amorphous eosinophilic material. There are interstitial fibrosis and tubular atrophy in about 10–20% of the section examined

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 m^2

At 10 months, she remains stable on medications with proteinuria of 0.07 mg/day and ESR of 16 mm in 1st 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.[3]

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 V lupus nephritis and Park *et al.* in 2012 in a 48-year-old woman with WHO Class IV lupus nephritis, respectively.[4,5] 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.[6] 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, mesangioproliferative 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.

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Conflicts of interest
There are no conflicts of interest.

References

1. Nephrotic syndrome. The Merck Manuals Online Medical Library. Available from: www.merckmanuals.com/professional/genitourinary_disorders/glomerular_disorders/overview_of_nephrotic_syndrome.html?qt=Nephrotic Syndrome in Adults&alt=sh. Updated March 2013. [Last accessed on 2013 Dec 4].
2. Chapman AB, Johnson AM, Gabow PA, Schrier RW. Overt proteinuria and microalbuminuria in autosomal dominant polycystic kidney disease. J Am Soc Nephrol 1994;5:1349-54.
3. Dalgaard OZ. Bilateral polycystic disease of the kidneys; a follow-up of two hundred and eighty-four patients and their families. Acta Med Scand Suppl 1957;328:1-255.
4. Wan RK, Kipgen D, Morris S, Rodger RSC. A rare cause of nephrotic syndrome in autosomal dominant polycystic kidney disease. NDT Plus 2009;2:136-38.
5. Park JI, Lee H, An JH, An JN, Chin HJ, Kim S. Laparoscopic biopsy-proven lupus nephritis in autosomal dominant polycystic kidney disease. Kidn Res Clin Pract 2012;31:192-5.
6. Visciano B, Renata A. Pietro D, Rossano R, Mancini A, Pasquale Zamboil P, Cianciaruso B, *et al*. Nephrotic syndrome and autosomal dominant polycystic kidney disease. Clin Kidney J 2012;0: 1–4.
7. Peces R, Martínez-Ara J, Peces C, Picazo M, Cuesta-Lopez E, Vega C, *et al*. Nephrotic syndrome and idiopathic membranous nephropathy associated with autosomal-

dominant polycystic kidney disease. Scientific- World Journal 2011;11:1041-7.
8. D'Cruz S, Singh R, Mohan H, Kaur R, Minz RW, Kapoor V, *et al*. Autosomal dominant polycystic kidney disease with diffuse proliferative glomerulonephritis–an unusual association: a case report and review of the literature. J Med Case Rep 2010;4:125.
9. Yenigun EC, Dede F, Ozkayar NV, Turgut D, Piskinpasa SV, Ozturk R, *et al*. Coexistence of Autosomal Dominant Polycystic Kidney Disease and Amyloidosis in a Patient With Nephrotic-range Proteinuria. IJKD 2014;8:243-5.
10. Savaj S, Parvin M, Savoj J. Massive proteinuria and autosomal dominant polycystic kidney disease: A rare coincidence. Iran J Kidney Dis 2012;6:73-6.