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CASE REPORT

Thirty years old lady with nephrotic syndrome: a case of biopsy proven lupus nephritis in Tanzania

FRANCIS FREDRICK[1,2*], PASCHAL J. RUGGAJO[2,3],GYAVIIRA MAKANGA[3], CHARLES K. SHIJA[3], MIKAEL AMDEMARIAM[3], BELSON RUGWIZAGONGA[4] and JAMES N. KITINYA[4]

[1]*Department of paediatrics and Child Health, School of Medicine, Muhimbili University of Health and Allied Sciences, P. O. Box 65001, Dar es Salaam, Tanzania*

[2]*Department of Internal Medicine, Muhimbili National Hospital, P. O. Box 65000, Dar es Salaam, Tanzania*

[3]*Department of Internal Medicine, School of Medicine, Muhimbili University of Health and Allied Sciences, P. O. Box 65001, Dar es Salaam, Tanzania*

[4]*Department of Pathology, School of Medicine, Muhimbili University of Health and Allied Sciences, P. O. Box 65001, Dar es Salaam, Tanzania*

Abstract: We describe a case of a 30 years old female patient who presented with nephrotic syndrome and impaired renal function diagnosed to have systemic lupus erythematosus (SLE). This is the first biopsy proven lupus nephritis in Tanzania. SLE is common among females and is reported be more common among Africans as compared to other races. This patient presented with nephrotic syndrome, pleural effusion and pericardial effusion which depicts the multisystem effects of SLE. This patient was treated with cyclophosphamide in combination with steroid as induction therapy and attained remission after a month of treatment. Systemic lupus erythematosus should be considered in patients with nephrotic syndrome and these patients should have renal biopsy to determine renal involvement.

Keywords: Nephrotic syndrome, lupus nephritis, systemic lupus erythermatosus, Tanzania

Introduction

Lupus nephritis is a common presentation of systemic lupus erythermatosus (SLE). It may present in a spectrum of manifestations including nephrotic syndrome which is estimated to be present in 45-65% of patients with lupus nephritis (Cameron, 1999). The diagnosis of lupus nephritis is usually considered in patient with SLE presenting with features indicating renal involvement, proteinuria, haematuria, red blood cell casts and elevated serum creatinine (Sigdel *et al*., 2013). This report describes a patient who presented with nephrotic syndrome to renal unit at Muhimbili National Hospital in Dar es Salaam, Tanzania. MNH is the national referral hospital which is also serving as a teaching hospital for Muhimbili University of Health and Allied Sciences (MUHAS).

Case report

A 30 years old lactating female was admitted in the medical ward at MNH with history of joint pain and generalized body swelling for 6 months, skin lesions and breast swelling for one month. Joint pain was involving multiple big joints; there was no history of joint swelling or morning stiffness. Body swelling started on the face and was worse in the morning subsiding towards the evening. Swelling also involved abdomen and the lower limbs. There was history of frothy urine. However, no history of haematuria or reduced urine output was given. The patient reported history of dyspnoea especially on lying flat and worse on exertion three weeks after generalized body swelling.

One month prior to admission, the patient developed itching generalized hyper-pigmented skin lesions which did not affect the face. The patient also gave history of swelling on the right breast which started on the nipple and progressed to involve the entire breast, six days

* Correspondence: Francis Fredrick; E-mail: fredrick.francis78@gmail.com

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before admission. The swelling was painful and was accompanied with fever. No history of trauma to the breast was reported prior to onset of swelling.

On physical examination she was wasted with light and easily pluck-able sparse hair with patchy alopecia. She had some palmar pallor with pedal oedema and no lymphadenopathy. Respiratory rate was 36 breaths/ minute and trachea was central. She had reduced tactile vocal fremitus bilaterally in the infra-mammary and infra-scapular positions and reduced breath sounds consistent with bilateral pleural effusion. Pulse rate was 112 beats/minute, blood pressure was 140/90 mmHg, apex beat with difficult to locate and heart sounds were muffled, these features were consistent with pericardial effusion. Patient had hyper-pigmented lesions on the abdominal skin, ascites and hepatomegaly of 12 cm span. The right breast was tender reddish with a pus discharging ulcer, the left breast was normal.

Urinalysis revealed proteinuria +3, twenty four hours urinary protein excretion was 41 g, serum albumin was 15 g/L, and serum cholesterol was 6.24 mmol/L. Serum creatinine was 192 µmol/L, serum urea was 43 mmol/L, glomerular filtration rate was estimated to be 34 ml/min. serum electrolytes revealed hyperkalaemia (6.9 mmol/L) and hyponatremia (128 mmol/L). Haemoglobin level was 7.8 g/dL with mean corpuscular volume and mean corpuscular haemoglobin concentration of 78.6 fL and 25 pg. respectively. Platelet count was 490 x 10^3/ µL. HIV, hepatitis B and C screening were negative. Anti-nuclear antibody test (ANA) and anti-double stranded DNA (anti-dsDNA) were both positive. Abdominal ultrasound revealed normal sized kidneys, grade one echogenicity and mild loss of cortical-medullary differentiation. Chest x-ray revealed bilateral pleural effusion, electrocardiogram showed low voltage and echocardiogram revealed pericardial effusion. The diagnoses of lupus nephritis and breast abscess were made based on the clinical features.

Renal biopsy was performed after stabilizing the patient and light microscopy slide with Haematoxylin and Eosin, Periodic acid Schiff and Silver stains were prepared. No immunofluorescence slides were prepared as the hospital does not have facility for immunohistochemistry. Light microscopy revealed 13 glomeruli with diffuse endocapillary proliferation, fibro-cellular crescents in three glomeruli, wire loops in capillary tufts and mesangial matrix expansion (Figures 1 and 2) which are consistent with lupus nephritis class IV-S.

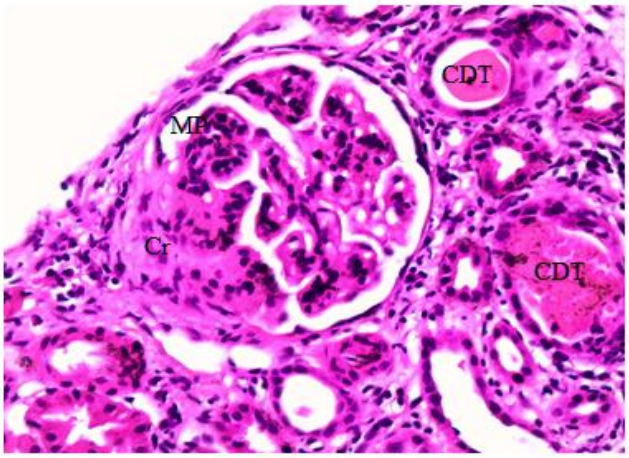

Figure 1: Glomerular lesion with fibro-cellular crescent (Cr) and mesangial proliferation (MP), CDT indicates cast in the tubules (Haematoxylin and Eosin stain; magnification x 400)

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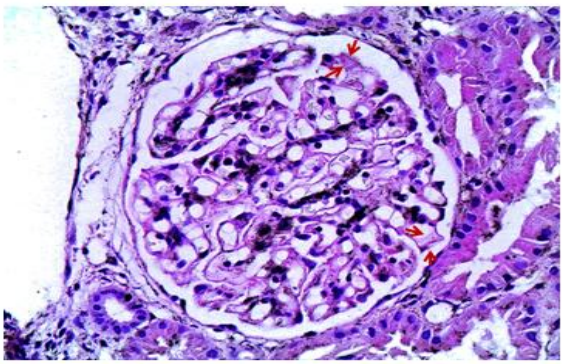

Figure 2: Double contouring (wire-loops) of glomerular basement membrane (arrows) (Periodic acid-Schiff (PAS) stain; magnification x 400)

Incision and drainage was performed for the breast abscess and the patient was also given ampicillin and cloxacillin for two weeks. She was given parenteral furosemide for oedema and hyperkalaemia and potassium was lowered to 5.4 after 5 days of diuretic treatment. The patient was given cyclophosphamide (500 mg) monthly injection for six months together with prednisolone (40 mg) as induction course for lupus nephritis and her laboratory normalized after two months of treatment. She was also given atorvastatin for hypercholesterolemia. Her laboratory results after one month of treatment revealed neither protein nor red blood cells, serum albumin was 40 g/L, serum cholesterol was 5.9mmol/L and serum creatinine was 52mol/L.

Discussion

Systemic lupus erythermatosus is a common connective disorder which has been described in East Africa by Tiffin et al., (2014). This condition has been ignored in resource limited countries including sub-Sahara Africa (McGill & Oyoo, 2002), this may be attributed to lack of rheumatologists in most of these countries. This is an autoimmune condition in which immune complexes play important role in causing tissue injury involving multiple organs and systems (Tsokos, 2011). Affected tissues in SLE show marked inflammation with deposition of antibodies and complements. The antibodies are autoantibodies directed against double stranded DNA of affected cells (Rahman & Isenberg, 2008). Female are more affected with SLE and Africans are reported to be more commonly than other ethnic groups (Carey *et al*., 2008; Kole & Ghosh, 2009; Dhakal *et al*., 2011).

Musculoskeletal and dermatological manifestations in the form of arthritis and discoid rashes are common presentation of SLE (Dhakal *et al*., 2011; Carey *et al*., 2008). These features were also reported by our patient. However the typical malar rashes of SLE were not reported. The patient also presented with pleural and pericardial effusion which are part of multi-organ involvement in patients with SLE (Gill *et al*., 2003). The diagnoses of SLE is usually made clinically with presence of at least four of the 11 American College of Rheumatology Classification criteria; malar rash, discoid rash, photosensitivity, oral ulcers, arthritis, serositis (pleural effusion, pericardial effusion), renal disorder, neurological disorder, haematological disorder, immunologic disorder and antinuclear antibodies (Gill *et al*., 2003). Our patient fulfilled the above criteria and the diagnosis of SLE with lupus nephritis was made.

Management of lupus nephritis requires a biopsy to histologically classify the condition based on the extent, activity and chronicity. The extent of renal involvement is also assessed and provides a baseline subsequent follow (Seshan & Jennete, 2009). Lupus nephritis is histologically

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classified into six classes based on the International Society of Nephrology/ Renal Pathology Society classification (Markowitz & D'Agati, 2007). The six classes have been well described by Seshan& Jennete (2009), Bihl *et al*. (2006) and Weening *et al*. (2004). Our patient was classified as lupus nephritis class IV-S which signify segmental involvement of >50% of the glomeruli examined histologically.

Cyclophosphamide monthly injection for 6 months combined with prednisolone was given to our patient as remission induction therapy and the patient was in remission one month after starting treatment. This treatment has been shown to be effective in treatment of lupus nephritis class III and IV. After completing induction therapy the patient is usually given maintenance therapy with mycophenolate mofetil or azathioprine combined with glucocorticoid (Mok *et al*., 2003; Houssiau, 2004; Waldman & Appel, 2006).

This report describes the case of histologically proven lupus nephritis in a patient presenting with nephrotic syndrome at MNH. It is therefore important to evaluate patients presenting with nephrotic syndrome for SLE serologically and then histologically for those with SLE to determine the extent of disease and appropriate treatment.

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Competing interests

Authors declare that there are no competing interests.

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