

## Case Report

# Lupus Podocytopathy: An Unusual Entity Requiring Inclusion in the Conventional Classification of Lupus Nephritis

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## Keywords

- Systemic lupus erythematosus (SLE)
- Lupus podocytopathy

## Abstract

Lupus podocytopathy, which is characterized by diffuse foot process effacement without peripheral capillary wall immune deposits and glomerular proliferation, has been described in SLE patients with proteinuria in case reports and small series. We present the case of a 38 year old female with systemic lupus erythematosus and significant proteinuria who had normal glomeruli on light microscopy. Immune fluorescence showed minimal mesangial deposits and electron microscopy revealed podocytopathy. Lupus podocytopathy is a relatively unusual subclass of Lupus Nephritis which should be considered for inclusion in the classification of SLE. It responds well to steroids and physicians and pathologists should keep a high index of suspicion to pick up such cases.

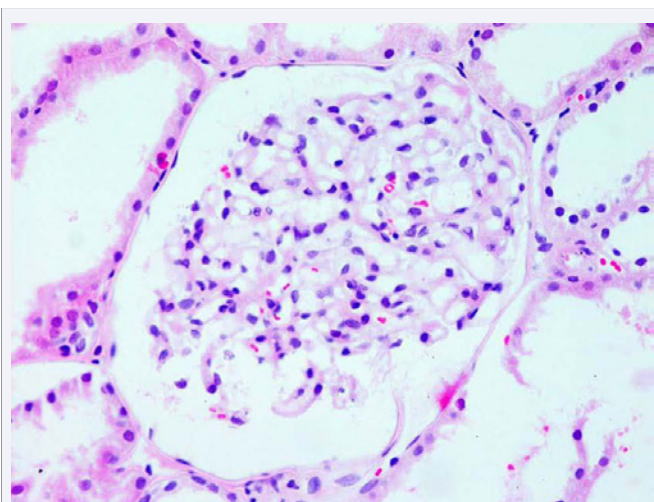
## INTRODUCTION

Proteinuria is a ubiquitous finding in lupus nephritis (LN), and is usually associated with immune complex deposition in the glomerular capillary wall. It is also frequently accompanied by endocapillary proliferation or necrosis. A subset of systemic lupus erythematosus (SLE) patients with significant proteinuria have been found to have either normal glomerulus or mild mesangial proliferation [1]. They have diffuse foot process effacement on electron microscopy (EM) without immune deposits in the capillary walls, suggestive of podocytopathy. Very few patients with lupus podocytopathy have been presented so far, that too in case reports or small series [2].

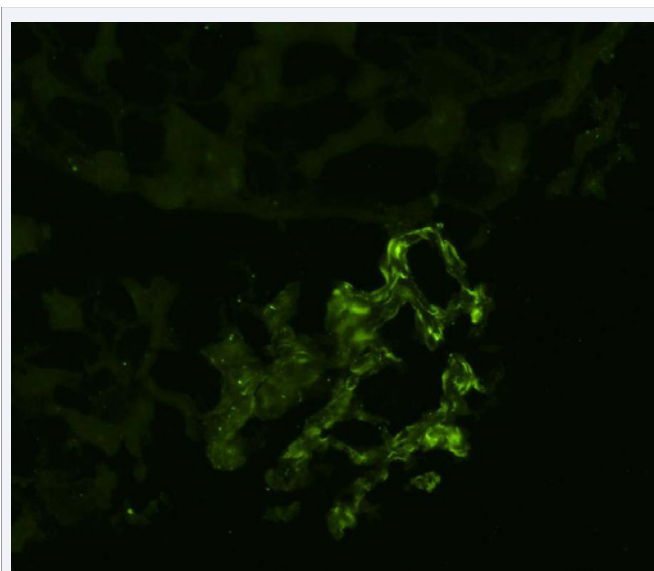
## CASE REPORT

A 38 year old lady presented with photosensitive hypopigmented plaques over bilateral malar region of face and trunk since last 5 years. She had also been complaining of hair loss and hard palate ulcers. Patient did not take any treatment in the past. She denied any frothy urine, hematuria, use of Non Steroidal Anti Inflammatory Drugs or tuberculosis in the past. She was found to be hypertensive during her evaluation on admission. She had hypopigmented plaques over the face bilaterally, abdomen and upper back. Rest systemic examination did not reveal any significant finding. Initial investigations revealed Hb- 11.6gm/dl, TLC- 5100/cumm, platelets- 125000/cumm. Urine routine

showed proteinuria with no active sediments. 24 hour urinary protein excretion was 2.08gm. She had normal renal function (urea- 18mg/dl, creatinine- 0.5mg/dl). Serum sodium- 141meq/Lt, potassium- 4.5meq/Lt. Liver function tests were normal with serum albumin being 3.5gm/dl. Her total cholesterol was 123mg/dl and triglycerides were 345mg/dl. Serology was positive for ANA (speckled pattern on IF with 1:160 titres) and anti-double-stranded DNA with low C3. SSA 3+, SSB 3+ and U1SnRNP 1+ were also positive. Serology was negative for rheumatoid factor, anti-proteinase 3 and anti-myeloperoxidase antibodies to cyclic citrullinated peptide. C Reactive Protein < 0.6mg/dl. Also serology for hepatitis B virus, hepatitis C virus and HIV was negative. Ultrasound abdomen showed left kidney- 12x4cm, right kidney- 10.5 x 3.5cm, with raised echotexture bilaterally. Kidney biopsy revealed unremarkable glomerular basement membrane and tubules with no significant interstitial inflammation on light microscopy (Figure 1). Immune Fluorescence (IF) showed mild mesangial staining for IgG and IgM. EM showed thickened glomerular basement membranes with extensive foot process effacement (Figure 2). Patient was managed with prednisolone 60 mg once a day for 12 weeks, which was then gradually tapered down over a period of next 3 to 4 months. Also she was started on angiotensin receptor blockers and statins. She showed improvement in the proteinuria after initiating therapy. 24 hour urinary protein was 1000mg at 4 weeks and 640 mg at 12 weeks.



**Figure 1** Normal Glomerular basement membrane, endothelial cells and interstitium on LM (40X).



**Figure 2** Immunofluorescence image of glomerulus showing mild mesangial staining for IgM.

She was also advised hydroxychloroquine 200mg once day for her cutaneous manifestations.

## DISCUSSION

The clinical presentation of nephrotic syndrome in a patient with SLE, with either absence of subendothelial or subepithelial immune deposits or confined to mesangium and foot process effacement on EM is suggestive of lupus podocytopathy. The current classification of lupus nephritis, revised by the International Society of Nephrology/Renal Pathology Society

(ISN/RPS) in 2003, does not recognize lupus podocytopathy as a distinct entity [3]. A series of 7 SLE patients who presenting with the nephrotic syndrome having diffuse podocyte effacement on renal biopsy was reported by Dube et al. [4]. The statistical chance of idiopathic minimal change disease and SLE occurring in the same individual is less than 1 in 10000 [5]. Similar series of cases have been reported the prevalence of this entity as 2 in 132 and 7 in 470 [5,6]. Various theories have been proposed for the pathogenesis of the mesangial glomerulopathy of lupus (World Health Organization class II) in patients with the nephrotic syndrome. Most common hypothesis suggests that a product of aberrant T cell function may be responsible for glomerular epithelial cell injury resulting in diffuse podocyte effacement [7]. There is activation of Th2 subset which overproduces IL-6, IL-10, IL-13, and tumor necrosis factor- $\alpha$  whereas Th1 subset under produces IL-2, interferon- $\gamma$  and transforming growth factor- $\beta$ . Some authors favour a circulating glomerular toxin hypothesis, where IL-13 has been the suggested toxin on the basis of overexpression in rats, apparently inducing minimal change picture on EM [8]. Our case provides support for the addition of a class or subclass of lupus podocytopathy to the histological classification of lupus nephritis. Podocytopathy in SLE responds to treatment with oral corticosteroids, and the response can be monitored by a decline in the magnitude of proteinuria. Clinicians and renal pathologists should be aware of this entity as a cause of nephrotic range proteinuria in patients with SLE.

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