Nephrotic Syndrome with Focal Segmental Glomerulosclerosis
Histological Feature: A Case Report

Dian Samudra1,*, Widodo2, Nunuk Mardiana2, Artaria Tjempakasari2, Pranawa2, Aditiawardana2, Anny Setijo Rahaju3

ABSTRACT

Introduction: Nephrotic syndrome is a clinical syndrome of heavy proteinuria and hypoalbuminemia or hydroproteinemia. Renal biopsy is fundamental to assess not only the type but also the degree of disease activity. The overall prognosis and response to treatment often depend on the severity of histological lesions and their reversibility.1,2 Case Presentation: An eighteen years old man with nephrotic syndrome and planned for a kidney biopsy. This case showed a patient with swollen face (especially on the cheek). On Biopsy results showing the glomerulus proliferation of cells and mesangial matrix, adhesions in (50%) glomerulus, focal sclerosis in some glomeruli, erythrocyte cells visible in the urinary space and thickening of the basement membrane in some glomeruli, partially atrophic tubules, visible erythrocytes in the tubular lumen. Conclusion: An eighteen years old man with nephrotic syndrome with cushing syndrome and hypokalemia who had a renal biopsy. The histological feature from the renal biopsy was focal segmental glomerulosclerosis. The underlying cause of FSGS is still unclear. Assessing the diagnosis and etiology become important to direct the subsequent clinical approach and therapy

Key words: Nephrotic syndrome, Focal segmental glomerulosclerosis, Renal biopsy.

INTRODUCTION

Nephrotic syndrome is a clinical syndrome of heavy proteinuria and hypoalbuminemia or hydroproteinemia, caused by increased permeability of serum protein which through the damaged basement membrane in the renal glomerulus. The definition of nephrotic syndrome includes massive proteinuria (3-3.5 g/day, or spot urine protein-creatinine ratio of 300-350 mg/mmol, or ≥3+ dipstick), hypoalbuminemia (serum albumin < 2.5 g/dl), and peripheral edema.1,2 In adults incidence of nephrotic syndrome is three per 100,000 persons.3 One of the primary glomerular diseases that can cause nephrotic syndrome is focal segmental glomerulosclerosis (FSGS).4 FSGS is a histological pattern of glomerular lesion that includes several completely different clinicopathological diseases that share injury within the podocyte as a primary pathophysiological feature.5 Early kidney biopsy in adults is critical to properly categorize the disease and direct the subsequent clinical approach.4 Renal biopsy is fundamental to assess not only the type but also the degree of disease activity. The overall prognosis and response to treatment often depend on the severity of histological lesions and their reversibility.2

CASE REPORT

Clinical feature

An eighteen years old man with nephrotic syndrome and planned for a kidney biopsy. This case showed a patient with swollen face (especially on the cheek). Previously, the swollen was on his whole body since October 2020 started from lower extremities, went up to stomach, upper extremities, but already diminished and only remained on his face area urinating a lot, yellow and foamy. At first, patient had medical treatment at hospital, he was hospitalized for a week and was diagnosed with nephrotic syndrome. During medication he received furosemide, osteocal, myfortic, methylprednisolone, diovon, simvastatin, albumin and calcium lactate. Patient was drunk alcoholic beverages for the last 6 years almost every weekend and claims to have stopped drinking since before the symptoms of the disease appeared. On physical examination, there were visible edema on both of his palpebra and cheeks.

Laboratory findings

On laboratory findings, there were proteinuria +4, PCR >50 g/gCr, Esbach protein excretion 1.5g/24 hours, negative from hepatitis C virus and HIV, but the hepatitis B virus test was equivocal and from the peripheral blood smear showed of polycythemia vera. The elucidation of lesions of glomeruli mandates that a variety of histochemical stains be used and that tissue sections be cut thinner than for other tissues. In paraffin-embedded sections, hematoxylin and eosin stain does not ordinarily allow for distinction of extracellular matrix from cytoplasm in a clear or convincing manner. Periodic acid-Schiff (PAS), periodic acid-methenamine silver (Jones), and Masson's trichrome stains all provide excellent definition of extracellular material. On March 22nd 2021, biopsy results showing the glomerulus proliferation of cells and mesangial matrix, adhesions in (50%) glomerulus, focal sclerosis in some glomeruli, erythrocyte cells visible in the urinary space and thickening of the basement membrane in some glomeruli, partially atrophic tubules, visible erythrocytes in the tubular lumen. The interstitial cells show lymphocytic infiltration of inflammatory cells. On immunofluorescence...
Focal Segmental Glomerulosclerosis (FSGS) is a disease characterized by thickening of the basement membrane of the glomeruli, which can lead to decreased filtration of blood and increased permeability of serum protein through the damaged basement. Nephrotic syndrome is a clinical syndrome that includes massive proteinuria, hypoalbuminemia, and peripheral edema which caused by the damage to the glomeruli.

### DISCUSSION

Nephrotic syndrome is a clinical syndrome that includes massive proteinuria, hypoalbuminemia, and peripheral edema which caused by the damage to the glomeruli. The clinical course of this patient is characterized by a history of alcohol consumption and renal insufficiency. The patient was diagnosed with nephrotic syndrome and treated with high-dose oral corticosteroids. The biopsy results were consistent with Focal Segmental Glomerulosclerosis (FSGS).

#### Table 1:

<table>
<thead>
<tr>
<th>Disease</th>
<th>Frequency of disease (%) as a cause of nephrotic syndrome</th>
<th>1960s and 1970s Patients &lt;60 years</th>
<th>Patients &gt;60 years</th>
<th>1990s to the present</th>
</tr>
</thead>
<tbody>
<tr>
<td>Focal segmental glomerulosclerosis</td>
<td></td>
<td>15</td>
<td>2</td>
<td>35</td>
</tr>
<tr>
<td>Membranous glomerular disease</td>
<td></td>
<td>40</td>
<td>39</td>
<td>33</td>
</tr>
<tr>
<td>Minimal change glomerular disease</td>
<td></td>
<td>20</td>
<td>20</td>
<td>15</td>
</tr>
<tr>
<td>Membranoproliferative glomerular disease (for example, IgA)</td>
<td></td>
<td>7</td>
<td>0</td>
<td>14</td>
</tr>
<tr>
<td>Other glomerular disease</td>
<td></td>
<td>18</td>
<td>39</td>
<td>3</td>
</tr>
</tbody>
</table>

#### Figure 1: Kidney biopsy findings.

Biopsy results in patients with nephrotic syndrome who have received steroid therapy. Histopathological feature with several stains found indicates that kidney has Focal Segmental Glomerulo Sclerosis (FSGS).

#### Figure 2: Proposed classification of FSGS.

According to the etiology, FSGS lesion has been classified into primary, genetic, and secondary forms that include maladaptive, virus-associated, and medication-induced FSGS.

Patient had a history of alcohol (arak) consumption since the last 6 years, almost every weekend, as much as approximately 1 big bottle (1-1.5L). The probability of alcohol consumption induced nephrotic syndrome can be excluded since there are no significant association between high alcohol consumption and risk for developing proteinuria. History of drug or traditional herbal drink consumption was denied. Distinguishing between the different forms is crucial since management must be tailored according to the underlying etiology.

From laboratory findings, patient test resulted negative from hepatitis C virus and HIV, but the hepatitis B virus test was equivocal, hence the test need to be repeated. We can also exclude autoimmune from suspected etiologies since the ANA test, C3, and C4 test results were within normal limit. The ASTO test was also negative; therefore, we can rule out post-streptococcal glomerulonephritis infection.

Adult patients with FSGS should receive the necessary supportive treatment as advised for all patients with persistent proteinuria, including the use of RAS blockade, optimal blood pressure control, and dietary salt restriction. Primary FSGS may respond to immunosuppressive treatment. On the other hand, maladaptive and secondary forms often present with sub-nephrotic proteinuria and progressive kidney insufficiency, do not respond to immunosuppression, and the treatment focuses in resolving the underlying cause.

During medication he received furosemide, osteocal, myfortic, methylprednisolone, diovan, simvastatin, albumin and calcium lactate. KDIGO recommend that high-dose oral corticosteroids be used as...
the first-line immunosuppressive treatment for primary FSGS. The recommended therapy is Prednisone 1mg/kg (maximum 80mg), continued by high dose corticosteroid therapy for at least 4 weeks and until complete remission is achieved, or maximum 16 weeks, whichever is earlier. When complete remission, continue for 4 weeks or 2 weeks after disappearance of proteinuria and reduce by 5 mg every 1-2 weeks to complete total duration of 6 months. And if partial remission within 8-12 weeks, then continue until 16 weeks, make sure whether further proteinuria or complete remission may occur and reduce by 5 mg every 1-2 weeks to complete total duration of 6 months. Calcineurin-inhibitor might also be used as treatment option for adults who may not tolerate prolonged high-dose corticosteroids well. Patients who have secondary FSGS due to an underlying disease process should be managed as required for the primary medical condition. There is no evidence or a priori rationale justifying the use of corticosteroids or other immunosuppressive drugs in this population, and the potential for harm of such treatment is clear. 2,5

SUMMARY

An eighteen years old man with nephrotic syndrome with Cushing syndrome and hypokalemia who had a renal biopsy. The histological feature from the renal biopsy was focal segmental glomerulosclerosis. The underlying cause of FSGS is still unclear. Assessing the diagnosis and etiology become important to direct the subsequent clinical approach and therapy

REFERENCES


GRAPHICAL ABSTRACT