Case Report

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

Dian Samudra1,*, Widodo2, Nunuk Mardiana2, Artaria Tjempakasari2, Pranawa2, Aditiawardana2, Anny Setijo Rahaju2

ABSTRACT

Introduction: Nephrotic syndrome is a clinical syndrome of heavy proteinuria and hypoalbuminemia or hypoproteinemimia. 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 hypoproteinemimia, 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.6 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, diovan, 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 24th 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...
Perihilar adhesion.

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increased permeability of serum protein through the damaged basement

proteinuria, hypoalbuminemia, and peripheral edema which caused by

Nephrotic syndrome is a clinical syndrome that includes massive

diovan 1x80mg, simvastatin 1x20mg, osteocal 1x1.

The patient was discharged with methylprednisolone 16mg - 16mg - 20 bpm, spo2 99% free air, axillary temperature 36 c, drinking 1200 clear. Consciousness comos mentis, blood pressure 103/67 mmHg, pulse 87 times/minute, regular, adequate contents, respiratory rate 20 bpm, spo2 99% free air, axillary temperature 36 c, drinking 1200 ml/24 hours, infusion 1500cc/24 hours, urine output 1900 ml/24 hours. The patient was discharged with methylprednisolone 16mg - 16mg - 8mg tapering down gradually, furosemide 1x20mg, myfortic 2x360mg, duvane 1x80mg, simvastatin 1x20mg, osteocal 1x1.

Clinical course

After ninth day of treatment, there is no complaints of pain, urine is clear. Consciousness comos mentis, blood pressure 103/67 mmHg, pulse 87 times/minute, regular, adequate contents, respiratory rate 20 BPM, SpO2 99% free air, axillary temperature 36 C, drinking 1200 ml/24 hours, infusion 1500cc/24 hours, urine output 1900 ml/24 hours. The patient was discharged with methylprednisolone 16mg - 16mg - 8mg tapering down gradually, furosemide 1x20mg, myfortic 2x360mg, duvane 1x80mg, simvastatin 1x20mg, osteocal 1x1.

DISCUSSION

Nephrotic syndrome is a clinical syndrome that includes massive proteinuria, hypoalbuminemia, and peripheral edema which caused by increased permeability of serum protein through the damaged basement membrane in the renal glomerulus. 6-14 This case showed a patient with swollen face (especially on the cheek). Previously of renal tubule, erythrocytes were shown on the lumen of renal tubule, lymphocyte inflammatory cell, the swollen was on his whole body since October 2020, but already diminished and only remained on his face area. Patient was hospitalized for a week and was diagnosed with Nephrotic Syndrome. On physical examination, there were visible edema on both of his palpebra and cheeks. On laboratory findings, there were proteinuria +4, pcr >50 g/gC, and esbach protein excretion 1.5g/24 hours. In adults, early kidney biopsy is fundamental to assess not only the type but also the degree of disease activity and also prognosis and response to treatment often depend on the severity of histological lesions and their reversibility. 1 Indications of renal biopsy are nephrotic syndrome, urine abnormalities (hemiauria and proteinuria), AKI and CKD. Nephrotic syndrome is the most frequent indication for renal biopsy in adults. The presence of glomerulosclerosis, arteriosclerosis, and interstitial fibrosis are all suggestive of an irreversible process that is less likely to respond to treatment; conversely, the presence of active lesions potentially indicates good responsiveness to corticosteroid treatment. Patients with proteinuria ≥ 1 g/day would deserve renal biopsy for clarifying the nature of the underlying nephropathy and should periodically be followed if such levels persist over time. 12,13 On laboratory findings, there were hemiauria with RBC 44.5/uL on urine sedimentation and proteinuria +4 on urinalysis. Also, there was proteinuria >1g/day which was measured on several visits: 1g/day on January 11th 2021 and increased 1.5g/day on March 6th 2021.

The renal biopsy result (March 22nd 2021) was cell proliferation and mesangial matrix were shown on glomerulus, adhesion, sclerosis. Focal Segmental Glomerulosclerosis. Histological feature: A Case Report

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).

Segmantal sclerosis

Perihilar adhesion.

HE, 400x; B. Masson Trichome,400x; C. PAS, 400x.

Figure 2: Proposed classification of FSGS.

Table 1:

| Disease                                | Frequency of disease (%) as a cause of nephrotic syndrome |
|----------------------------------------|----------------------------------------------------------|
|                                        | 1960s and 1970s                                          |
|                                        | Patients <60 years | Patients >60 years | 1990s to the present |
| Focal segmental glomerulosclerosis     | 15               | 2                 | 35                  |
| Membranous glomerular disease          | 40               | 39                | 33                  |
| Minimal change glomerular disease      | 20               | 20                | 15                  |
| Membranoproliferative glomerular disease (for example, IgA) | 7               | 0                 | 14                  |
| Other glomerular disease               | 18               | 39                | 3                   |

examination of IgG, IgM, and IgA is negative, C3 and Clq also negative. The conclusion is Focal Segmental Glomerulosclerosis (Figure 1).

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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

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