ABSTRACT

Acute nephritic syndrome is a collection of clinical symptoms due to a sudden decrease in glomerular filtration rate (GFR) accompanied by water and salt retention, positive erythrocytes, erythrocytes cast and albumin in urinalysis. Information about nephritic syndrome is essential to know for accurate diagnoses and treatment. A 13-year-old adolescent with nephritic syndrome has been reported with rapid progressive glomerulonephritis (RPGN). He has been treated with corticosteroid, antibiotic, immunosuppressant, antihypertensives, and diuretic followed by improvement of clinical condition and laboratory of the patient. The biopsy was done for definitive diagnosis. The biopsy results showed a picture of IgA nephropathy (IgAN) and resolving stage of acute poststreptococcal glomerulonephritis (APSGN). Acute poststreptococcal glomerulonephritis (APSGN) was suspected due to finding of apparent hematuria, swollen and acute renal failure after streptococcal infection. To support the diagnosis, evidence of laboratory streptococcal infections and low levels of C3 complement are needed. Whereas IgAN was known by examining the IgA serum. Management of patients was including supportive and symptomatic therapy. Giving corticosteroids or cytotoxic agents is needed for RPGN therapy. The prognosis is generally good, with improvement of more than 90% of cases. Long-term observation is needed to observe the possibility of the disease being chronic. In this case, the symptoms and results of the laboratory examination were following the results of the biopsy, namely IgAN and resolving APSGN. Immediate and proper management provides an improvement in general and laboratory conditions.

KEYWORDS

acute poststreptococcal glomerulonephritis, IgA Nephropathy, Nephritic Syndrome

Introduction

Acute nephritic syndrome is a collection of clinical symptoms due to a sudden decrease in glomerular filtration rate (GFR) accompanied by water and salt retention, positive erythrocytes, erythrocytes cast and albumin in urinalysis. Although the common cause of the acute nephritic syndrome is APSGN [1,2] however, it is necessary to consider another differential diagnosis. Nephritic syndrome and nephrotic syndrome has similar symptoms, except the two significant differences symptoms such as hypertension and red blood cell cast. Glomerulonephritis is a significant cause of kidney disorders accounting for 10%-15% of cases of late-stage renal failure in the United States.[1] In primary glomerulonephritis, the disease is almost entirely confined to the kidneys (as in IgAN or APSGN) in secondary glomerulonephritis there is more widespread inflammation (such as in systemic lupus erythematosus (SLE) or systemic vasculitis).[1,2] This case report is made to provide sufficient information about the management of nephritic syndrome as described as the course of the disease is useful for the management of the disease immediately and precisely.

Case report

A 13-years-old male adolescent was hospitalized with chief complaints of swelling on the face and both extremities. The patient also complains of dark urine accompanied by fever without pain...
Figure 1: A. A glomerulus with moderate hypercellular appearance of mesangial cells and endocapillary cell proliferation. B. Glomerulus with mild hypercellular appearance. C. Glomerulus with attachment to the glomerular tuft.

Figure 2: Glomerulus with neutrophil infiltrates (400x magnification).

Figure 3: A. The tubular epithelium is relatively within normal limits (100x magnification); B. Some lumen tubules contain erythrocytes (400x magnification).

On urinating. Four days afterwards the patient complained of fever, cough, runny nose and pain when swallowing.

On physical examination, the patient was alert and in good orientation, blood pressure 160/110 mmHg, pulse 104 times/minute, respiratory rate 24 times/minute, temperature 37.30°C, body weight 60 kg, and height 155 cm, and with normal body mass index (BMI). No eyelid edema, no signs of anemia or jaundice. Tonsil enlargement was found, no pharyngeal hyperemia, normal jugular venous pressure (JVP). Normal heart sounds without murmurs, vesicular pulmonary sounds, no rhonchi nor wheezing. In the abdomen not found ascites, liver and spleen were not palpable, and all four limbs appear edema and warmth. The color of the urine looked like tea-colour.

Laboratory tests showed leukocyte or white blood cells (WBC) 5.9 x 10³ / ul; hemoglobin 11.85 g / dl; platelets 246,5 x 10³ / ul; blood urea nitrogen (BUN) 30 mg / dl; serum creatinine (SC) 3.3 mg / dl; aspartate aminotransferase (AST) 17.8 IU / l; alanine aminotransferase (ALT) 8.4 IU / l; sodium 135 mmol / l; potassium 3.7 mmol / l; albumin 1.94 g / dl; total cholesterol 466.7 mg / dl; high density lipoprotein (HDL) 47.31 mg / dl; low density lipoprotein (LDL) 351.4 mg / dl; triglycerides (TG) 230 mg / dl, from urinalysis showed turbidity (+); pH 5; specific gravity 1.020; leukocytes (+++) 500; protein (+4); nitrite (-); blood (+5); bacteria (++); many leukocyte cells / field of view; many erythrocytes / morphic / visual field, viral marker for hepatitis was negative, anti-streptolysin O (ASTO) negative results. On examination of protein Esbach showed proteinuria of 3.8 grams / liter / day. Abdominal ultrasound (USG) was done and found no abnormalities on examination. Throat swab confirmed Streptococcus viridans as the infectious agent. Complement C3 and C4 examination were normal.

Treatment given were furosemide 40 mg OD, pulse dose methylprednisolone 500 mg for three days, tapering down until dose of 4 mg BID, Irbesartan 150 mg OD, spironolactone 25 mg OD, Cefoperazon 1 gr OD intravenous. Patients continued treatment at the polyclinic using azathioprine 25 mg TID, cyclosporine A (Sandimmun Neoral®) 25 mg BID, irbesartan 150 mg OD, furosemide 40 mg OD, spironolactone 25 mg OD, methylprednisolone 4 mg BID, and amlodipine 5 mg OD.

After continued the treatment, improvement as no swelling, no tea-colored urine and no fever. On physical examination not found abnormalities of heart and lungs. No eyelid and extremities edema. Urine production in 24 hours is 1000 cc (0.92 cc / kgBW / hour).

Laboratory results were improved, WBC 7.5.10³ / ul; hemoglobin 14.9 g / dl, platelet 376.6.10³ / ul; BUN 9 mg / dl; SC 0.67 mg / dl; ALT 10.9 IU / l; Na 138 mmol / l; K 4.24 mmol / l; albumin 4.24 g / dl; total cholesterol 269.4 mg / dl; HDL 67 mg / dl; LDL 198 mg / dl; TG 123 mg / dl, from urinalysis showed turbidity (-); pH 6; BJI 1,019; leukocytes (-); protein (+1); nitrite (-); blood (-); bacteria (-); bilirubin (-); many leukocyte cells / field of view; many erythrocytes / morphic / visual field, viral marker for hepatitis was negative, anti-streptolysin O (ASTO) negative results. On examination of protein Esbach showed proteinuria of 2 grams / liter / day. Renal biopsy was performed and confirmed a picture of IgA Nephropathy, and resolving GNAPS.

Discussion

Rapid progressive glomerulonephritis is characterized by rapid deterioration of kidney function, usually a 50% reduction in GFR in less than 3 months.[2-4] Although various diseases can cause RPGN, all types of RPGN have the characteristics of glomerular injury and crescent form on renal biopsy. Severe damage
to glomerular basement membrane (GBM) causes plasma protein leakage through GBM. Among proteins, fibrin is thought to make the strongest contribution to crescent formation. The epithelial cells lining the Bowman capsule react to fibrin leakage and replicate. Infiltration of WBC such as monocytes and macrophages can also multiply. These cells multiply around and compress the glomerulus, forming a crescent-shaped scar that is easily visible on a light microscope.[2–4]

The pathogenesis of GNAPS is not known with certainty. The latent period between streptococcal infection and glomerular abnormalities shows that immunological processes play an important role in the mechanism of disease. Allegedly the excessive response of the host immune system to the antigen stimulus with excessive antibody production causes the formation of antigen-antibody (Ag-Ab) complexes which later cross the GBM. Here occurs the activation of the complement system that releases substances that will attract neutrophils. Lysosomal enzymes released by neutrophils are responsive factors to damage the glomerulus.[1-5]

The etiology and pathogenesis of IgAN are still not fully understood, but basic evidence suggests this disease is an immune-mediated disease in which the IgA-immune complex settles in the glomerulus. IgAN is described as an IgA-IgG immune complex disease with a multi-hit pathogenic process. In particular, galactose deficient (Gd)-IgA1 is produced in large quantities in patients with IgAN (Hit 1) detected in the circulation by autoantibodies (Hit 2). The result is the formation of immune pathogenic immune complexes (Hit 3), some of which eventually settles in glomerular mesangium and cause kidney injury (Hit 4).[6-8]

Acute poststreptococcal glomerulonephritis suspected when clinical symptoms are seen in the form of hematuria that arises suddenly, swollen and acute kidney failure after streptococcal infection. Characteristic signs of glomerulonephritis in urinalysis, evidence of laboratory streptococcal infections and low levels of complement C3 support the evidence for diagnosis. Patient with suspicion of IgAN need to fulfilled the following of four clinical markers, that is: 1). More than 5 erythrocyte cells were found in urine sediment, 2) Persistent proteinuria of more than 0.3 g / day, 3). Serum IgA more than 315 mg / dL, 4). Serum IgA / C3 ratio is more than 3.01.[6-8]

In APSGN, renal biopsy is considered if severe kidney function is impaired especially if the etiology is unclear (develops into kidney failure or nephrotic syndrome), there is no evidence of streptococcal infection, no decrease in complement levels, long-lasting improvement with persistent hypertension, azotemia, gross hematuria after 3 weeks, low C3 levels after 6 weeks, proteinuria that persists after 6 months and hematuria that persists after 12 months.[9-11]

In this case, the decline in kidney function, hematuria, and proteinuria occurred within 2 weeks. Patients also experience hypertension and edema, these characteristics are in accordance with the RPGN. In this case, renal impairment begins with an infection in the upper airway, as evidenced by the presence of Streptococcus viridans from the throat swab. Unfortunately the patient could not be tested for serum IgA on cost grounds. Biopsy is performed on patients with consideration to find out a definitive diagnosis, there is no decrease in complement, prolonged improvement and the presence of proteinuria and hematuria after 6 months of treatment.

Renal biopsy showed that >50% glomeruli with mild-moderate hypersellularity in the form of increased global mesangial cell cellularity, as well as endocapillary cell prolifera-

tion. Some glomeruli show focal epithelial proliferation. One glomerulus shows a neutrophil infiltrate, endocapillary impression. Some glomerules also show the attachment of the tuft glomerulus to Bowman’s capsule. At some tubular lumen erythrocytes appear. In another focus, mononuclear cells infiltrates in interstitial tissue appear. The conclusion of the biopsy results was the picture can be found in IgAN and found APSGN resolving.

Conclusion
A 13-year-old male adolescent with nephritic syndrome has been reported with RPGN. Treated with corticosteroid, antibiotic, immunosuppressant, antihypertensives and diuretic, followed by improvement of clinical condition and laboratory of the patient. Kidney biopsy was performed for diagnostic and therapeutic considerations. The biopsy results showed a picture of IgA nephropathy and resolving stage of GNAPS. The prognosis is generally good, with improvement more than 90% of cases. Long-term observation is needed to observe the possibility of the disease being chronic.

Conflict of Interest
There are no conflicts of interest to declare by any of the authors of this report.

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References

