

Patients with Steroid Sensitive Nephrotic Syndrome: A Case Series

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ABSTRACT

Nephrotic syndrome (NS) is one of the most common kidney diseases found in childhood. The prevalence of nephrotic syndrome worldwide is approximately 16 cases per 100,000 children with an incidence of two to seven per 100,000 children. Nephrotic syndrome may affect adults and children of both genders and any race. Nephrotic syndrome in children can be classified into three groups; secondary, congenital, infantile, and idiopathic. In first case, a 3-year-old male patient was presented with complaints of fever, cough for 6 days and swelling all over the body, decreased urine output, oral intake for 2 days, edema of face, body and tiredness for 3 days. Laboratory investigations showed elevated ESR, ferritin and lipid profile. Urine routine examination showed elevated urine albumin levels (+++). USG abdomen and pelvis conveyed right basal pleural effusion. In second case, a 4-year-old male patient was presented with complaints of facial puffiness, fever, breathing difficulty, abdominal distension and pain but no edema. He had history of right inguinal hernia and atrial septal defect. Cholesterol level was found to be elevated. Urine routine examination showed elevated urine albumin levels (+++). USG abdomen and

pelvis showed mild ascites and right pleural effusion. Presenting third case, a 12-year-old female patient was presented with complaints of edema and decreased urine output. Patient had history of recurrent episodes of edema and decreased urine output for 11 months. Laboratory investigations showed elevated total cholesterol, declined LFT test and RFT test was found to be normal. Urine routine examination showed elevated urine albumin levels (+++), pus cells, RBC, granular cast and bacteria present. USG abdomen and pelvis indicate acute glomerulo nephritis and acute to moderate ascites. Here case series with 3 cases showed the sequence of nephrotic syndrome which is treated with corticosteroid with its side effects and cumulative dose of steroid in children.

KEYWORDS: Nephrotic syndrome, Hypocholesteremia, Hypoproteinuria, Hyponatremia, pediatric

INTRODUCTION

One of the most frequent childhood kidney diseases is nephrotic syndrome (NS) ⁽¹⁾. Nephrotic syndrome is a chronic disorder characterized by leakage of proteins in urine resulting hypovolemia, hyper coagulation and infection, this may be due to alterations of pam selectivity at the glomerular

capillary wall. In nephrotic syndrome, proteinuria is exceeding 1000mg/m²/day or spot urinary protein to creatinine ratio exceeding 2mg/mg⁽²⁾. Main causes of NS include genetic disorders, nephropathy, drugs and secondary diseases associated with infections. It could affect adults and children of both genders and any race. Nephrotic syndrome in children can be classified into three groups; secondary, congenital, infantile, and idiopathic. Secondary nephrotic syndrome is associated with inflammatory diseases like IgA nephropathy, glomerulonephritis, lupus nephritis. Congenital and infantile nephrotic syndrome are occurring before the age of one year and are mostly related to infections like syphilis, toxoplasmosis or with gene mutation coding for podocytes proteins and are steroid resistant. Idiopathic (primary) nephrotic syndrome is the most common form of NS in children reporting more than 90% of cases between 1 and 10 years of age and 50% after 10 years of age ⁽³⁾. Children with long lasting nephrotic syndrome may be increased risk for ischemic cardiovascular events due to hyperlipidemia ⁽⁴⁾. On the basis of clinical examinations, nephrotic syndrome was suspected and specific laboratory tests was performed to confirm the diagnosis. The urine examination indicated proteinuria, albuminuria and monitoring the lipid levels, urine output, blood pressure and renal function tests. Renal biopsy is another option to diagnose kidney disease. NS isn't curable, but medical management helps to relieve nephrotic symptoms and prevent further damage to the kidneys. Supportive treatment includes intravenous albumin, corticosteroids, low salt intake, high caloric and protein diet.

CASE PRESENTATION

CASE: 1

A 3-year-old male patient was presented with complaints of fever, cough for 6 days and swelling over the body, decreased urine output and oral intake for 2 days. He also had edema of face, body and tiredness for 3

days. On physical examination he has conscious, afebrile, tiredness and retractions present with bilateral rhonchi positive. Hematological investigations was found to be normal except ESR (78 mm/hr) and lymphocytes (15 %). LFT showed total protein (3.3 g/dL) and serum albumin (1.4 g/dL) was declined. RFT results revealed declined sodium (118 mmol/L) and creatinine levels (0.4 mg/dL). Ferritin (351 ng/ml) was seemed to be increased in this condition. Lipid profile revealed the total cholesterol (252 mg/dL) was elevated. Urine routine examination showed elevated urine albumin levels (+++). USG abdomen and pelvis conveyed minimal ascites and right basal pleural effusion. He was started with IV antibiotics (INJ. MONOCEF 650 mg IV BD), Nebulization (LEVOSALBUTAMOL 0.63mg), Proton Pump Inhibitors (INJ. RANTAC 15mg IV Q8H), Corticosteroids (T. PREDNISOLONE 10 mg BD), Calcium supplement and Multivitamin syrup and infusion normal saline. The treatment for nephrotic syndrome was started with T. PREDNISOLONE 10 mg BD and continued as syrup for 6 weeks. After this regimen, the dose of Syrup PREDNISOLONE was doubled and given once daily. On review, he was presented with puffy face, then the treatment was continued for 6 weeks with alternate day.

CASE: 2

A 4-year-old male patient was presented with complaints of facial puffiness, fever, breathing difficulty, abdominal distension and pain but no edema. He had history of right inguinal hernia and atrial septal defect (ASD), which was spontaneously closed. Physical examination revealed he has conscious, oriented and febrile. Hematological investigations, liver function test and renal function test was found to be normal. Cholesterol level was found to be elevated (326mg/dL). Urine routine examination showed elevated urine albumin levels (+++). USG abdomen and pelvis depicted bilateral echogenic kidneys, mild ascites and right pleural effusion. He was

started with IV antibiotics (INJ. CEFTRIAZONE+SULBACTAM 650 mg IV BD), corticosteroids (SYP. PREDNISOLONE 10ml-10ml-5ml for 2 weeks), Proton Pump Inhibitors (INJ. PANTOPRAZOLE 40mg IV BD). During follow up, first relapse had occurred which lasts for 6-month duration and was treated with T. PREDNISOLONE 20mg. As such, 4 relapse were occurred within a year.

CASE: 3

A 12-year-old female patient was presented with complaints of edema and decreased urine output. She had history of recurrent episodes of edema and decreased urine output for 11 months. She was earlier diagnosed with nephrotic syndrome at age of 11-month, first relapse occurred at the age of 4, second relapse at the age of 11 and currently with third relapse. Hematological investigations were found to be normal, except ESR (105 mm/hr). LFT showed total protein (3.9 g/dL), serum globulin (2.4 g/dL) and serum albumin (1.5 g/dL) were declined. RFT was found to be normal. Lipid profile revealed the total cholesterol (1073 mg/dL) was elevated. Urine routine examination showed elevated urine albumin levels (+++), puscells (20-25/hpf), RBC (3-4/hpf), granular cast and bacteria present. USG abdomen and pelvis conveyed acute glomerulonephritis and acute to moderate ascites. He was started with IV antibiotics (INJ. CEFUROXIME 1.5g mg IV BD), Corticosteroids (T. PREDNISOLONE 20 mg 1½-0-1½), statins (T. ATORVASATAIN 10mg HS for 1 month). After 6 weeks, the dose of T. PREDNISOLONE was tapered. After 4 months patient was presented with no symptoms.

DISCUSSION

Nephrotic syndrome is an important chronic disease in children. The defect in glomerular filtration results severe edema and proteinuria⁽⁵⁾. This is a case series of steroid sensitive nephrotic syndrome in childhood age. Two patients were male and one was

female. These three cases have similar features of nephrotic syndrome like swelling all over the body. One of the patients (case: 3) was diagnosed as Congenital nephrotic syndrome, it is a rare hereditary kidney disease that begins at birth or within the first three months of life⁽⁶⁾. In three cases, diagnosis were made by physical examination and laboratory parameters (edema, proteinuria, and hyperlipidemia). More than 80 percentage of nephrotic syndrome show minimal change disease (MCD) and the remaining shows focal segmental glomerulosclerosis (FSGS). Individuals with nephrotic syndrome are at risk for life threatening infections and thromboembolic condition. Urine albumin was positive in three cases, which showed the condition of proteinuria. In lipid profile, cholesterol is highly elevated in three patients. The chief complications of nephrotic syndrome is infection, hypertension, hyperlipidemia, features of corticosteroid toxicity and behavioral disorders⁽⁷⁾. Persistent hyperlipidaemia and prolonged steroid therapy are recognized in nephrotic syndrome. Corticosteroid therapy efficiently play a role in remission of proteinuria⁽⁸⁾. For those children who responds to steroids will have one or more relapses and half will have frequently relapsing or steroid dependent NS⁽⁹⁾.

CONCLUSION

Nephrotic syndrome is the defect in glomerular filtration results severe edema and proteinuria. Diagnosis of nephrotic syndrome depends on certain criteria. Heavy proteinuria, hypoalbuminemia, edema (especially lower extremities) and hyperlipidemia. Corticosteroid is the first line drug of choice for nephrotic syndrome. There are number of other therapeutic options which include Alkylating agents (Cyclophosphamide), Calcineurin inhibitors (Cyclosporine), Mycophenolate mofetil, Rituximab and Emerging drugs (Abatacept, Galactose, etc). The vital factor which determines the nephrotic syndrome is steroid responsiveness. But the side effect of

steroids can be reversible after the cessation of steroid therapy. Prolonged use of steroid may cause growth failure which can be managed by alternate regimen. Counselling to patient guardian about the treatment of nephrotic syndrome is important. This review discussed the treatment option and remission with minimal medication side effects.

Declaration by Authors

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