e-ISSN: 3025-6224

Scientific Journal of Pediatrics (SJPed)

Journal website: https://phlox.or.id/index.php/sjped

Nephrotic Syndrome in Pediatrics: A Case Report

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ARTICLE INFO

Keywords:
Glomerular
Kidney biopsy
Nephrotic syndrome
Neoplasia
Pediatric

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All authors have reviewed and approved the final version of the manuscript.

https://doi.org/10.59345/sjped.v1i2.46

A B S T R A C T

Introduction: The prevalence of childhood NS worldwide is approximately 16 cases per 100,000 children, with an incidence of two to seven per 100,000 children. Primary causes of NS include minimal change disease, focal segmental glomerulosclerosis, membranous nephropathy, genetic disorders, and secondary diseases associated with infections, drugs, and neoplasia; however, it can also be idiopathic. **Case presentation:** A 14-year-old male toddler weighing 39 kg with a 1-week history of swelling around the eyes and both legs and generalized body swelling. On physical examination, swelling was found in both lower legs (pitting type). Laboratory investigations showed protein in urine and reduced serum albumin (2.0 g/dL) with elevated lipid levels. Although a kidney biopsy could not be performed due to the economic problem of the family, a diagnosis of idiopathic nephrotic syndrome (NS) was made based on clinical and laboratory findings. **Conclusion:** Nephrotic syndrome in children includes dietary adjustments, administration of diuretics to reduce edema, and administration of immunosuppressants such as steroids to induce remission.

1. Introduction

One of the most common childhood kidney diseases is nephrotic syndrome (NS). The prevalence of childhood NS worldwide is approximately 16 cases per 100,000 children, with an incidence of two to seven per 100,000 children. Primary causes of NS include minimal change disease, focal segmental glomerulosclerosis, membranous nephropathy, genetic disorders, and secondary diseases associated with infections, drugs, and neoplasia; however, it can also be idiopathic. NS can affect children of any age, from infancy to adolescence, and predominantly occurs in those aged 1–6 years. Pediatric nephrotic syndrome, also known as nephrosis, is defined by the presence of nephrotic-range proteinuria, edema, hyperlipidemia, and hypoalbuminemia. Nephrotic-range proteinuria in adults is characterized by protein excretion of 3.5 g or more per day. However, because of the great range of body sizes in children, the pediatric definition of nephrotic-range proteinuria is more cumbersome.[1-4]

Nephrotic-range proteinuria in children is protein excretion of more than 40 mg/m^2/hr. Because 24-hour urine collections are potentially unreliable and burdensome, especially in young children, many pediatric nephrologists instead rely on a single, first-morning urine sample to quantify protein excretion by the ratio of protein to creatinine. The use of a first-morning urine sample eliminates the contribution of potentially nonpathological orthostatic proteinuria, which might otherwise falsely elevate the protein level in a urine sample collected while a patient is active during the day. A urine protein/creatinine value of more than 2-3 mg/mg indicates nephrotic range

proteinuria and correlates with results from 24-hour urine collection.[5-7]

Nephrotic syndrome is a constellation of clinical findings that is the result of massive renal losses of protein. Thus, nephrotic syndrome is not a disease itself but the manifestation of many different glomerular diseases. These diseases might be acute and transient, such as postinfectious glomerulonephritis, or chronic and progressive, such as focal segmental glomerulosclerosis (FSGS). Still, other diseases might be relapsing and remitting, such as minimal change nephrotic syndrome (MCNS). The glomerular diseases that cause nephrotic syndrome generally can be divided into primary and secondary etiologies. Primary nephrotic syndrome, also known as idiopathic nephrotic syndrome (INS), is associated with glomerular diseases intrinsic to the kidney and not related to systemic causes. The subcategories of INS are based on histological descriptions, but clinical-pathological correlations have been made. The disorder can affect people of all ages. In most children, the first sign of nephrotic syndrome is facial swelling. Adults usually present with dependent edema. The nephrotic syndrome could affect adults and children of both genders and any race. Also, it could occur in a typical form or with nephritic syndrome. Glomerular inflammation leads to hematuria and impaired renal function.[8-10]

2. Case Presentation

A 14-year-old male weighing 39 kg presented with a history of swelling over the face, which initially started around the peri-orbital (which is more during the morning) and gradually progressed to the face, which decreased by evening. The child had decreased urine output (oligouria). The baby was spontaneous vaginal delivery and weighed 1,90 kg after birth. On examination, a pitting type of oedema was present over the lower limbs, and swelling over the face was present. Based on these clinical presentations, nephrotic syndrome was suspected, and specific laboratory testing was performed to establish the diagnosis. The urine dipstick indicated proteinuria and haematuria. Blood testing showed a significant hypoalbuminaemia of 2.0 g/dL (reference interval 3.5-5.5 g/dL), indicating nephrotic syndrome (NS). The urine creatinine level was 0.3mg/dl (reference interval 0,6-1,1mg/dl). The lipid levels were markedly increased, as outlined in Table 1.

After establishing a diagnosis, optimal supportive treatment, including captopril per oral, prednisolone per oral., intravenous albumin, furosemide, low salt intake, and high caloric and protein diet, were given along with ceftriaxone. The urine output and blood pressure were monitored. Successful control of peripheral oedema with the administration of albumin and diuresis with furosemide was seen. Periorbital oedema and leg swelling were reduced, and there was a concomitant increase in serum protein levels. The lipid levels also gradually decreased in the course of time without any medication.

3. Discussion

A patient comes with complaints of swelling. In this patient, the swelling starts from the eyelids and continues until there is edema all over the body. This shows that the swelling in this patient leads to kidney disorders. To help establish the diagnosis, supporting examinations are needed in the form of complete blood laboratory tests, complete blood chemistry, and urine. From the laboratory examination results, albumin 2 g/dl, ureum 28 mg/dl, creatinine 0.3 mg/dl, total cholesterol 509 mg/dl, and urine protein was positive +3. From the results of anamnesis, physical examination, and laboratory examination, this patient had anasarka edema, hypoalbuminemia, hypercholesterolemia, and massive proteinuria. The hallmark of nephrotic syndrome is massive proteinuria, which causes a decrease in circulating albumin levels. The hallmark of nephrotic syndrome is massive proteinuria, which causes a decrease in circulating albumin levels.

Table 1. Laboratory parameters.

Blood serum		
Haemoglobin	17,0 g/dL	13-18 g/dL
Erythrocyte	6,1 jt/ul	4,4-5,5 jt/uL
Hematocrit	47%	40-45 %
Leukocyte	12.000/ul	5000-1000/uL
Platelets	329.000/ul	150.000-300.000/uL
Ureum	28 mg/dL	20-40 mg/dL
Creatinin	0,3 mg/dL	0.6-1,1 mg/dL
Urinalysis		
Color	Brown	Clear
PH	7	5,5-8,5
Glucose	Negative	Negative
Ketone	Negative	Negative
Bilirubin	Negative	Negative
Urobilinogen	positive +1	Negative
Blood	positive +2	Negative
Protein	positive +3	Negative
Leukocyte	Negative	Negative
Nitrite	Negative	Negative
Serum electrolytes		
Serum sodium	131 mmol/L	136-145 mmol/L
Serum chloride	98 mmol/L	98-107 mmol/L
Blood serum		
Serum potassium	4,5 mmol/L	3,4-4,7 mmol/L
Serum lipid profile		
Total cholesterol	509 mg/dL	<170 mg/dL
LDL cholesterol	494 mg/dL	<110 mg/dL
HDL cholesterol	33 mg/dL	
Triglycerides	596 mg/dL	<75 mg/dL
Others		
Albumin	2 g/dL	3,5-5,5 g/dL

The initial events that result in proteinuria are still unknown. However, strong evidence suggests that nephrotic syndrome has, at least in part, an immune pathogenesis.[11-13]

The classic explanation for edema formation is a decrease in plasma oncotic pressure as a result of low serum albumin levels, leading to extravasation of plasma water into the interstitial spaces. The resulting contraction in plasma volume (PV) causes stimulation of the reninangiotensin-aldosterone axis and secretion of anti-diuretic hormone. The resultant retention of sodium and water by the renal tubules contributes to the expansion and maintenance of the edema. A more recent theory of edema formation posits that massive proteinuria causes tubular-interstitial inflammation, local release of vasoconstrictors, and inhibition of vasodilation. This causes a decrease in glomerular filtration rate and retention of sodium and water. Almost all levels of lipids (cholesterol, triglycerides) and serum lipoproteins are elevated in nephrotic syndrome. This can be explained by explanations, among others, namely the existence of a hypothermic

condition that stimulates overall protein synthesis in the liver, including lipoproteins. In addition, fat catabolism decreases because there is a decrease in plasma lipoprotein lipase levels, the main enzyme system that extracts fat from the plasma. Treatment of nephrotic syndrome in patients includes dietary adjustments, administration of diuretics to reduce edema, and administration of immunosuppressants such as steroids to induce remission.We have presented a case of idiopathic Nefrotic syndrom successfully managed with a corticosteroid, furosemide, captopril, and ceftriakson. The mainstay of treatment for NS is corticosteroids (steroids) with protocols largely based on seminal studies from the International Study of Kidney Disease in Children and the Arbeitsgemeinschaft für Pädiatrische Nephrologie.[14,15]

4. Conclusion

Nephrotic syndrome is one of the clinical features of gromelurar disease characterized by massive proteinuria, edema hypoalbuminemia, and hypercholesterolemia. Investigations that can be carried out to confirm the diagnosis of nephrotic syndrome are blood tests, urinalysis, and blood chemistry. Treatment of nephrotic syndrome in children includes dietary adjustments, administration of diuretics to reduce edema, and administration of immunosuppressants such as steroids to induce remission.

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