Glomerular Disorder of Unknown Etiology in a Child with Unusual Course

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Abstract
A 3-year-old girl was presented with periorbital edema, hypertension, proteinuria, and hematuria. She recovered clinically after 9 days with normal urinalysis. During the follow-up, she developed recurrent episodes of nephrotic syndrome. The kidney biopsy revealed mild mesangial proliferation and a low dose of prednisolone could effectively control the disease.


Keywords ● Glomerulonephritis ● Nephrotic syndrome ● C3

Introduction
Sudden onset of edema, hematuria, hypertension, proteinuria and renal insufficiency is generally considered as acute glomerulonephritis (AGN). The characteristic features of nephrotic syndrome (NS) are heavy proteinuria, hypoalbuminemia, hyperlipidemia and edema. Complete normalization of urinalysis in AGN, during the first two weeks, is a rare event. To our knowledge, initial presentation of a glomerular disease as AGN with unknown etiology and rapid normalization of urinalysis followed by recurrences as NS has not been explained previously. So this patient is presented as an unusual form of glomerular disease with undetermined cause.

Case report
A 3-year-old girl was admitted to the division of pediatric nephrology of Nemazee hospital affiliated with Shiraz University of Medical Sciences, Shiraz, Iran, with tea color urine and periorbital edema for five days. Three weeks before admission she was treated with amoxicillin and diphenhydramine for her upper respiratory tract infection. In the second month of her life and for a period of 2 weeks, she was treated for thrombocytopenia and after that she had a normal platelet count. On admission she had no history of fever, skin rash, joint pain, or abdominal pain but had a history of allergic rhinitis (confirmed by skin test). Her physical examination revealed a blood pressure of 150/100 mmHg with body weight of 14 kg. She had bilateral pulmonary wheezing and pitting edema of the lower limbs. Her family history was negative for allergic disorders, but her parents had a history of renal stone.

Urinalysis showed: 2+ proteins, 2+ blood (many RBCs with no RBC cast). Erythrocyte sedimentation rate was 110 mm/hr; random urine protein/creatinine was 8 mg/mg. Other parameters such as CBC, platelet count, serial BUN, serum creatinine, TG, cholesterol, C4, electrolytes, and ultrasonography of the kidneys were normal at the time of admission. ASOT was 100 IU/ml, ANA was negative, C3 was 1.5 g/l (normal range: 0.7-1.2 g/l) and throat culture was negative.
Nine days after admission she was discharged with normal urinalysis and without receiving any medication. During two months follow-up period, she developed four relapses with 10-14 days intervals. Each relapse was associated with edema, proteinuria (2-3+) and microscopic hematuria, with normal BUN, serum creatinine and blood pressure. During each relapse and 3-4 days thereafter, and without receiving any specific treatment, proteinuria spontaneously disappeared.

Kidney biopsy was performed and of the total 20 glomeruli in the light microscopy, some were normal and some with mild to moderate mesangial cell proliferation and matrix expansion. Tubules, interstitium and vessels were normal (Fig 1). Immuno-fluorescent study of the kidney tissue revealed trace IgM in mesangium and 1+ C3 in tubular basement membrane and interstitium.

Electron microscopic study revealed seven glomeruli with patent capillary lumen containing RBCs, mild mesangial matrix expansion, cell proliferation, and focal fusion of foot processes without any electron dense deposit (Fig 2). C3 declined to 0.5g/l, so prednisolone therapy (15 mg/day) started with rapid response and gradual normalization of C3. During 36 months follow-up, two more relapses occurred by slowly tapering the dose of prednisolone. The relapses responded to temporarily increasing the dose of prednisolone to 15mg/day.

Discussion

This is a rare form of glomerular disease with unknown etiology. The patient was initially presented with hypertension, gross hematuria and edema which all are in favor of AGN rather than of idiopathic nephrotic syndrome. Dunn has reported two cases of AGN with initial normal urinalysis. Rapid normalization of urinalysis with hematuria and proteinuria, in less than 2 weeks, is not expected. Most patients with post streptococcal AGN have shown normal urinalysis within a period of a few months to one year.

In this patient, pathologic findings ruled out IgA-nephropathy, lupus nephritis and other secondary causes of AGN. Immune thrombocytopenic purpura and allergic rhinitis might be expected in the idiopathic nephrotic syndrome, but the initial presentation as AGN with C3 changes were against it. Deposition of IgM in mesangium may be suggestive of IgM-nephropathy, but the pathologic and clinical courses were not in favor of this disease. The recurrence of AGN is unusual, especially in the form of nephrotic syndrome, whereas the relapse rate of idiopathic nephrotic syndrome is as high as 60 to 75%. As reported earlier, in this patient relapses were significantly prevented with a starting low dose of prednisolone and by gradually increasing the interval.

In summary, this patient was initially presented with an acute glomerulonephritis of unknown etiology followed by frequent relapses of nephrotic syndrome which dramatically responded to low dose of prednisolone.

References


