Rapidly Progressive Glomerulonephritis Undetected by Routine School Urinalysis: A Case Report

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A twelve year old girl with rapidly progressive glomerulonephritis (RPGN) is reported. First symptoms of renal failure developed in August 1987 just five months after a routine school urinalysis which detected no abnormalities. The patient developed end stage renal failure within 2 months of onset. Continuous ambulatory peritoneal dialysis (CAPD) was introduced to manage the renal failure, and open renal biopsy was performed simultaneously in order to elucidate the underlying renal disease in this patient. Microscopic examination of the biopsy specimen demonstrated marked formation of crescents in about 90% of the glomeruli, and some of the glomeruli were already sclerotic. Granular depositions of IgG (trace), IgA (1 +), IgM (3 +), properdin (3 +), C1q (3 +) and C3 (5 +) were observed by immunofluorescence staining. A definite diagnosis of rapidly progressive glomerulonephritis was made as the cause of renal failure in this patient. It was suggested that open renal biopsy is useful in diagnosing the underlying kidney disease of end stage renal failure in cases where the clinical course is relatively short.

(Key Words: RPGN, renal biopsy, CAPD, routine urinalysis, acute renal failure, pediatrics)

INTRODUCTION

Rapidly progressive glomerulonephritis is known to take an extremely acute clinical course, and often results in renal failure (4). The main pathological feature of this nephritis is remarkable crescent formation in the glomeruli (1, 12, 17). Therefore, this disease has also been referred to as malignant (3), acute necrotizing (12), extracapillary proliferative (1), and idiopathic crescentic glomerulonephritis (3). The disease is predominantly seen in the third to sixth decade, with a median age ranging from 39 to 58 years (5, 11). Occurrence of this disease in childhood is known to be very rare (1, 10). In this report we present a case of a young girl with rapidly progressive glomerulonephritis which was not detected by urinalysis at school. She developed end stage renal failure after a very short clinical course.

We performed an open renal biopsy to define the underlying renal disease in this patient, and continuous ambulatory peritoneal dialysis (CAPD) was successfully introduced to manage her renal failure.

CASE REPORT

A twelve year old girl was admitted to the Tokai University Hospital with complaints of appetite loss, systemic edema and worsening dyspnea on September 2, 1987. Just five months before her admission, a routine school examination including urinalysis, chest X-ray and a physical examination revealed no abnormalities. The main vital signs on admission were a body temperature of 37.4°C, heart rate of 112/min. and blood pressure of 150/108 mm Hg. Physical examination revealed the following: the palpebral conjunctivas were markedly anemic. Distinct gallop rhythm and ejection...
murmurs (Levine III/VI) were audible. Wet rales were detected in the bilateral lungs. On the abdomen, the liver edge was palpable about 1.5 cm just below the right costal margin. Peri orbital and bilateral pretibial edema was marked. Laboratory studies demonstrated the following data: proteinuria; 0.42g/day, urinary sediment: 10–20 red blood cells/high power field, and 5–10 hyaline casts/high power field, erythrocyte sedimentation rate: 77mm/hr, hemoglobin: 5.1g/dl, hematocrit: 15.6%, urea nitrogen: 156 mg/dl, creatinine: 17.4 mg/dl, sodium: 158 mEq/l, potassium: 7.1 mEq/l, chloride: 110 mEq/l, IgG: 1240 mg/dl, IgA: 146 mg/dl, IgM: 303 mg/dl, C3: 55 mg/dl, C4: 30 mg/dl, CH50: 40 mg/dl, antinuclear antibody: (1 +) diffuse type, antigranular base ment membrane antibody: negative, and anti-DNA antibody: negative. The clinical diagnosis was acute renal failure (ARF), and rapidly progressive glomerulonephritis was suspected as the major cause of this ARF.

Open renal biopsy was performed on October 1 for a pathological diagnosis. The biopsy specimen revealed marked cellular or fibrous crescents in about 90% of the glomeruli. Glomerular tufts had collapsed, and some of the glomeruli were completely sclerotic. Urinary tubules were atrophic and mononuclear cell infiltration was observed in the interstitium (Fig. 1). Granular, partly linear, deposition of IgG(trace), IgA(1 +), IgM(3 +), proper dine(3 +), Clq(3 +) and C3(3 +) was observed by immunofluorescence (Fig. 2). However, no electron dense deposits were detected by electron microscopy (Fig. 3).

The clinical course is depicted in Fig. 4. Hemodialysis had been performed for 14 times after admission and before CAPD induction. With this therapy, her clinical condition greatly improved and CAPD was introduced as maintenance dialysis therapy on October 1, 1987 with simultaneous open renal biopsy. The clinical course after CAPD induction was favorable. Her edema and cardiomegaly also improved, and all subjective symptoms had almost disappeared by about 4 weeks after CAPD induction.

DISCUSSION

Pediatric rapidly progressive glomerulonephritis (RPGN) is generally thought to be rare (1, 10). However the South West Pediatric Nephrology Study Group reported 50 cases of

Fig. 1 Glomerular tufts had collapsed with marked cellular crescent formation. (PAS × 400)
Fig. 2 Deposits of IgM and C3 in the glomeruli were demonstrated by immunofluorescence. (×400)

Fig. 3 Electron microscopy showed oppressed capillary loops due to crescent formation, and wrinkling of the basement membrane was revealed. No electron dense deposits were observed.
E.K.  12yo, female

Fig. 4  Clinical course.

pediatric RPGN in 1985 (15). They classified these cases into nine categories (1) nonspecified immune complex disease, (2) systemic lupus erythematosus, (3) idiopathic (no evidence of immune complex), (4) poststreptococcal glomerulonephritis, (5) IgA nephropathy, (6) Henoch-Schönlein purpura, (7) vasculitis, (8) possible antilglomerular basement membrane disease, and (9) dense deposit disease.

Our case seems to correspond to idiopathic type on the basis of the pathological findings such as no linear pattern IgG and no evidence of the existence of dense deposits in electron microscopy.

We have already reported the usefulness of open renal biopsy for diagnosis of the underlying renal disease in a patient with moderate stage renal failure (13). We performed open renal biopsy on this patient with rapidly progressive end stage renal failure, and found that the underlying disease was idiopathic RPGN with remarkable crescent formation which seemed to be resistant to any curative therapy. We think that renal biopsy for patients with end stage renal failure is not always meaningless because patients with RPGN, which has a rather short clinical course, appear to have a number of residual glomeruli in the kidney. If a sufficient amount of kidney specimen can be obtained, accurate diagnosis of the underlying renal disease may be feasible and thus lead to the selection of the appropriate type of treatment.

Poor linear body growth has been reported in children undergoing hemodialysis, and this growth retardation is thought to be due mainly to nutritional restrictions and renal osteodystrophy (6, 16). On the other hand, improved nutrition is considered to be an important benefit of continuous ambulatory peritoneal dialysis. Therefore, we suspected that children managed with CAPD might have better growth than those managed with hemodialysis. Our patient has been almost free from any nutritional restrictions, and has shown 2.7 cm of linear growth with a satisfactory nutritional state during the 10 months since introduction of CAPD. This growth rate was similar to that of healthy children of her age.

Generally, RPGN takes a very acute clinical course and the mortality of this disease is extremely high (1, 4). Even in our case, her condition on emergency admission with dyspnea and systemic edema was life threatening. We performed emergency hemodialysis to save her life. This serial hemodialysis resulted in steady
improvement of her uremic symptoms and we selected continuous ambulatory peritoneal dialysis for maintenance therapy of the renal failure. Transient increases in body weight, CTR, BUN and serum creatinine were seen after the start of CAPD, but these signs with improved with stabilization of CAPD ultrafiltration.

In the therapy of rapidly progressive glomerulonephritis, as with many other diseases, early detection of the disorder and early initiation of the therapy are the most important points in achieving a satisfactory outcome. In this case we did not attempt aggressive curative therapy for RPGN to improve renal function after the introduction of CAPD because histological destruction of the glomeruli was so severe that we thought these changes were irreversible no matter what the therapy. If our case could have been detected 2 or 3 months earlier and a precise diagnosis could have been made with open renal biopsy or other diagnostic means, some treatment such as steroid (7), anticoagulant (2), antiplatelet (9), immunosuppressive (14) or plasma exchange (8) therapy might be effective for improving renal function, and maintenance dialysis might have been avoided. However, 2 months before admission, the patient was almost free from subjective symptoms and detection of the disease at that point seemed to be very difficult. The only means to prevent patients with rapidly progressive glomerulonephritis such as our patient from falling into end stage renal failure is to perform frequent urinary screenings every 2 or 3 months. As a means of preventing end stage renal failure, further improvements in the screening system (frequency and/or procedure) should be considered.

REFERENCES