Renal Manifestation of Pustulosis Palmaris et Plantaris

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Diffuse proliferative glomerulonephritis without IgA deposits (DPGN) in patients with pustulosis palmaris et plantaris (PPP) is described. This association has not been previously reported. Although PPP was resolved transiently after tonsillectomy, it was worsened coincidently at the time of exacerbation of DPGN. Improvement of proteinuria and PPP was observed after antiplatelet therapy. It was suggested that the renal manifestation and the development of PPP were associated.

Key words: Diffuse proliferative glomerulonephritis, Proteinuria, Hematuria, Focal infection, Tonsillectomy, Antiplatelet therapy

CASE REPORT

A 50-year-old female patient was admitted to the Tokai University Hospital because of microscopic hematuria and pustular skin lesions in the palms and soles (Fig. 1a). She was well until 39 yrs of age when she suffered from recurrent tonsillitis. At 40 yrs of age, she noticed pustular skin eruptions in her bilateral palms and soles, the skin lesions gradually worsened after recurrent tonsillitis. She was diagnosed as having pustulosis palmaris et plantaris (PPP) by a local dermatologist in April 1985. Since recurrent tonsillitis was frequently observed and tonsillar hypertrophy was grade 2 in Mackenzie’s classification, after a positive tonsillar provocation test on June 12, 1985, she underwent tonsillectomy in our hospital. At that time, she was found to have microscopic hematuria (3+) and slight proteinuria (+). Her renal function test was within the normal range. No urological abnormality was observed. Although microscopic hematuria persisted, her skin lesions were gradually improved after the tonsillectomy (Fig. 1b).

On January 22, 1986, she was admitted to the Tokai University Hospital for further examinations and treatment. In her past history, she had a gastric ulcer which was pointed out in February 1984. However, there was no fever, edema, macroscopic hematuria, polyuria, polydipsia, photosensitivity, Raynaud’s phenomenon, polyserositis or deafness before admission. No relevant family history of renal diseases, deafness, diabetes mellitus, vasculitis and/or collagen diseases was found.

On admission, her body temperature was 36.2°C, pulse 76/min with regular rhythm, and her blood pressure was 128/80 mmHg. White blood cell (WBC) count was 5,100/mm³ with 66% neutrophils, 30% lymphocytes, 4% monocytes and...
Fig. 1. Skin lesions in this patient with pustulosis palmaris et plantaris. upper (la), the skin lesion before tonsillectomy; lower (lb), the skin lesion two yrs after tonsillectomy.

1% eosinophils. Red blood cell (RBC) count was $4.26 \times 10^6$/mm$^3$, hemoglobin (Hb) 14.3 g/dl, hematocrit (Ht) 42.0% and platelet count 2.76 $\times 10^5$/mm$^3$. Blood urea nitrogen (BUN) was 13 mg, serum creatinine (s-Creat) 0.5 mg, uric acid 4.2 mg, glucose (FBG) 73 mg, total cholesterol (T.Chol) 213 mg and triglyceride 183 mg per 100 ml. Sodium was 140 mEq, potassium 3.4 mEq, chloride 106 mEq, glutamic oxalacetic transaminase (SGOT) 24 U, glutamic pyruvic transaminase (SGPT) 18 U, lactic dehydrogenase (LDH) 229 U, total cholesterol (T.Chol) 213 mg and triglyceride 183 mg per 100 ml. Sodium was 140 mEq, potassium 3.4 mEq, chloride 106 mEq, glutamic oxalacetic transaminase (SGOT) 24 U, glutamic pyruvic transaminase (SGPT) 18 U, lactic dehydrogenase (LDH) 229 U and alkaline phosphatase (ALP) 108 U/l. Serum protein was 7.1 g (albumin 4.5 g), serum IgG 1,053 mg, IgA 292 mg and IgM 137 mg per 100 ml. IgE was 23 IU/ml. C3 was 90 mg, C4 29 mg per 100 ml, and CH$_50$ was 33.6 U/ml. Clq-binding immune complex was less than 1.5 $\mu$g/ml.

$\beta$-2-microglobulin ($\beta$-2-MG) was 1.3 mg in serum and 64 $\mu$g in urine/l. Tissue plasminogen activator (t-PA) was 5.0 ng in plasma (normal range, less than 7.6), 6.8 ng in urine (normal range, less than 7.6), and D-dimer was 41 ng in plasma (normal range, less than 150) and 80 ng in urine (normal range, less than 150)/ml. Maximal aggregation of plasma was 8.5% by 1 $\mu$M of adenosine diphosphate (ADP), 16.2% by 2 $\mu$M of ADP, 33.9% by 5 $\mu$M of ADP and 66.9% by 2 $\mu$M/ml of collagen. Platelet adherence was 72.3%. Antinuclear antibody, RA test and cryoglobulin were negative. On urinalysis, urine contained 0.3 g of protein a day and 10–50/HPF of red cells in urinary sediments.

Several hyaline casts were also observed in the urine. Creatinine clearance was 101 ml/min and PSP test for 15 min was 57% (total amount for 120 min, 86%).

An electrocardiogram, chest x-ray film and audiogram were normal. Abdominal x-ray film revealed gas bubbles in the stomach on the right side. Partial visceral inversion was observed under her diaphragm. No peculiar gas accumulation was observed. Gastric endoscopic findings showed atrophic gastritis without gastric or duodenal ulcer. Examination for ocular lesions showed no arteriosclerotic changes.

Open renal biopsy was performed without any complications on January 24, 1986. Renal biopsy specimens contained 72 glomeruli per section. In sections stained with hematoxylin and eosin (H-E) and periodic acid Schiff (PAS), all glomeruli showed slight to moderate diffuse mesangial expansion with segmental mesangial cell proliferation (Fig. 2). Glomerular lobulation, tram track sign, wire-loop lesion or foam cells were not observed in such glomeruli. The percentages of glomerular adhesion to Bowman’s capsule, glomerular sclerosis and crescent formation were 16.7%, 10.4%, and 4.2%, respectively. Thickening of glomerular capillary walls or that of intrarenal extraglomerular arterioles was not observed in the renal tissue. Inflammatory changes of intrarenal arterioles, i.e. vasculitis, were not observed. Focal tubular atrophy was observed. Immunofluorescent studies revealed diffuse granular and/or interrupted linear deposition of IgM and properdin in glomerular capillary walls and mesangial areas (Fig. 3). Less intense deposition of IgA, C3, C4 and fibrinogen was also observed in
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Fig. 2. Light microscopic findings in the glomeruli (PAS, ×400).

Fig. 3. Immunofluorescent staining findings of IgM in the glomeruli (×400).

Fig. 4. Electron microscopic findings (×3,000).

The patient was treated with an antiplatelet drug (Ticlopidine, 200 mg/day) because of persistent microscopic hematuria. During the therapy, microscopic hematuria and pustular skin lesions were gradually improved. In June 1986, she noticed such glomeruli. No deposition of IgG, IgE, C1q or C5 was observed. Electron dense deposits were observed in the glomerular capillary walls, mesangium and subendothelial cells (Fig. 4). However, splitting, lamellation or irregular attenuation of glomerular basement membrane (GBM) or the subepithelial hump was not observed.

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worsening of the pustular skin lesions on her soles. At that time, she had no fever, osteoarthropathic symptoms or preceding infections such as pharyngitis, although in the past she had suffered from upper respiratory tract infections. Her skin lesions were soon improved without any dermatological treatment such as ointment or lotion. Six wks after the worsening of the skin lesions, exacerbation of microscopic hematuria was observed in her urinalysis (Fig. 5). These urinary abnormalities were gradually improved after dipyridamole administration (300 mg/day). In May 1988, she again noticed worsening of pustular skin lesions in the palms and soles after acute pharyngitis. Slight worsening of urinary findings was again observed eight wks after the worsening of the skin lesions. However, no decline in renal function or occurrence of the nephrotic syndrome was noted during that episode.

DISCUSSION

A 50-year-old female patient with pustulosis palmaris et plantaris (PPP) associated with chronic diffuse proliferative glomerulonephritis without IgA deposits (DPGN), who showed improvement of proteinuria and PPP after tonsillectomy and antiplatelet therapy, is reported. This association has not been previously described.

PPP is a localized pustular eruption involving the palms and soles which was originally described by Barber in 1927 (1). Although the pathogenesis of this disease remains unknown (2, 5–10), it was suggested that anti-stratum corneum (anti-SC) might play a role in the pustular formation in the skin (5). It was also suggested that a focal infection such as tonsillitis is a major factor in the occurrence and persistence of PPP (2, 9), because PPP was frequently improved after tonsillectomy (2). In the present case, PPP and proteinuria were significantly improved after tonsillectomy, although PPP was again worsened at the time of exacerbation of DPGN. On the other hand, it was suggested that the pathogenesis of glomerulonephritis is partly associated with viral or bacterial focal infections (11–13). We have previously reported that exacerbation of IgA nephropathy is occasionally observed after upper respiratory tract infections (14). Tomino et al reported that some viral substances in the pharyngeal washings were associated with eluted antibodies in the glomeruli obtained from patients with IgA nephropathy (15, 16). Furthermore, the deposition of IgM and properdin was observed in the glomeruli in this patient. It is feasible that such a deposition of IgM and properdin is suggested to have a role in the pathogenesis of PPP and/or DPGN. The present results suggest that there might be a common pathogenetic cause in both renal and skin manifestations in this patient. There is also the possibility that the tonsils were not the sole focus of focal infection, but it is premature to conclude whether the renal or skin lesions were one of the focal infections. It can be suggested that DPGN and the development of PPP were associated. Careful observations are warranted to determine if the infectious focus is actually deleterious to renal lesions.

REFERENCES

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