Rapidly Progressive Glomerulonephritis in Association with Henoch-Schönlein Purpura in a Patient with Advanced Liver Cirrhosis

M.	Makoto	Ogawa ^a
Y.	Yasuhiko	Makino ^a
S.	Shiro	Ueda ^a
M.	Masao	Ohto ^a
B.	Bunshiro	Akikusa ^b

^a1st Department of Internal Medicine and ^b2nd Department of Pathology, Chiba University School of Medicine, Chiba, Japan

Makoto Ogawa, MD, First Department of Internal Medicine, Chiba University School of Medicine, 1-8-1 Inohana Chuouku No. 260, Chiba (Japan)

Dear Sir,

Secondary IgA nephropathy is a common complication of liver cirrhosis (LC) [1, 2]. However, it seldom takes a rapidly progressive course [1]. Herewith we report a case of LC with Henoch-Schönlein purpura (HPS) and crescentic glomerulonephritis with mesangial IgA deposits.

A 56-year-old man with LC was admitted in December 1993. He had been followed up since 1989, when the diagnosis of LC was confirmed by biopsy. He was positive for hepatitis C virus (HCV) antibody. His urinalysis revealed mild haematuria, but serum creatinine concentration was normal. 1 week before admission, he suddenly developed massive haematuria and purpura in his leg following acute respiratory tract infection.

Physical examination on admission revealed icteric conjunctivae, abdominal distension, and non-tender purpuric macules of his trunk and extremities (fig. 1). The laboratory examination revealed haemoglobin 95 g/l, white blood cells $6.5 \times 10^9/l$, platelets $88 \times 10^9/l$, bilirubin $36 \mu\text{mol/l}$ (2.1 mg/dl), alanine aminotransferase 88 IU/l, serum creatinine $344 \mu\text{mol/l}$ (3.9 mg/dl), serum albumin 26 g/l, and prothrombin time 14.4 s. Urinalysis showed 150 red blood cells per high-power field, and the urine protein level was 2.6 g/day. Anti-HCV antibody was positive, but serological examination was negative for hepatitis B surface antigen, hepatitis B core antigen antibody, antiglomerular basement membrane antibody, antimitochondrial antibody, anti-DNA antibody, and antineutrophil cytoplasmic antibody. The serum C3 level was 0.35 (normal 0.5-

Fig. 1. HSP of the trunk. The abdomen was distended due to ascites.

tion showed that half of the glomeruli had cellular or fibrocellular crescents with mesangial change (fig. 2). Granular deposits of IgA and C3 in the mesangial area were also confirmed. The vascular change was minimal.

Cutaneous vasculitis in LC has often been associated with HSP [3, 4] and mixed cryoglobulinaemia [5, 6]. In this case, leuco-clastic vasculitis with IgA deposits seems compatible with HSP, and cryoglobulin testing was negative. The glomerular changes might

have existed before the onset of HSP, but cellular crescents are rare in LC [1]. We think, therefore, that the renal changes in

1.3) g/l. Ultrasonography revealed massive ascites and an atrophic liver with an irregular surface. Skin biopsy revealed leucocytoclastic vasculitis with perivascular IgA deposits suggestive of HSP. The patient received plasmapheresis after intravenous methylprednisolone therapy ($0.5 \text{ g} \times 3 \text{ days}$), but the serum creatinine concentration increased to $637 \mu\text{mol/l}$ (6.0 mg/dl). His condition deteriorated with an increase of the total bilirubin level and a prolongation of the prothrombin time. On the 22nd hospital day, he died of hepatorenal failure. Postmortem examination confirmed LC, but no malignancy was found. Microscopic examina-

5%¹⁰*f

1/8

Fig. 2. Photomicrograph of a typical glomerulus showing cellular crescents and mesangial sclerosis.

this case occurred, at least in part, with the cutaneous lesions. Aggarwal et al. [4] already reported a case of LC with acute renal failure and HSP with IgA deposits. Such a deposition has been associated with an impaired clearance of IgA immune complex in LC [4, 7]. This mechanism seems to have worked in our case as well. The preceding upper respiratory tract infection or HCV might have contributed to the formation of immune complex.

The prognosis of rapidly progressive glomerulonephritis has been improved after plasmapheresis and intravenous methyl-prednisolone therapy were widely introduced [8]. However, such treatment was ineffective in this case. Hepatic function seems to have a decisive influence on the prognosis of HSP-associated rapidly progressive glomerulonephritis in patients with LC.

References

- Newell GC: Cirrhotic glomerulonephritis: Incidence, morphology, clinical features, and pathogenesis. *Am J Kidney Dis* 1987;9:183-190.
- Strom EH, Dumuller Gudat F, Mihatsch M: Hepatitis C virus plays no role in the pathogenesis of immunoglobulin A nephropathy in liver cirrhosis. *Nephron* 1994;67:370.
- Robeva R, Krusteva R, Belovezhkov N: Benign monoclonal gammopathy in a female patient with Schönlein-Henoch glomerulonephritis and liver cirrhosis. *Vutr Boles* 1990;29:105-109.
- Aggarwal M, Manske CL, Lynch PJ, Paller MS: Henoch-Schönlein vasculitis as a manifestation of IgA-associated disease in cirrhosis. *Am J Kidney Dis* 1992;20:400-402.
- Popp JW, Harriest TJ, Dienstang JL, Bhan AK, Wands JR, LaMont T, Mihm MC Jr: Cutaneous vasculitis associated with acute and chronic hepatitis. *Arch Intern Med* 1981; 141: 623-629.
- Misiani R, Bellavita P, Fenilli D, Borelli G, Marchesi D, Massazza M, Vendramin G, Comotti B, Tanzi E, Scudeller G, Zanetti A: Hepatitis C virus infection in patients with essential mixed cryoglobulinemia. *Ann Intern Med* 1992;117:573-577.
- van de Wiele A, Valentijn RM, Schuurman HJ, Daha MR, Hene RJ, Kater L: Circulating IgA immune complexes and skin IgA deposits in liver disease in relation to liver histopathology. *DigDisSci* 1987;33:679-684.
- Levy JB, Winearls CG: Rapidly progressive glomerulonephritis: What should be first-line therapy? *Nephron* 1994;67:402-405.

366

Ogawa/Makino/Ueda/Ohto/Akikusa

Rapidly Progressive HSPN in Liver Cirrhosis