Case 7

Full-House Nephropathy in a Girl with Nephrotic Syndrome

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[Introduction]

Full-house nephropathy is a poorly defined disease entity characterized by full-house immunofluorescence pattern in renal pathology without clinical or serological lupus parameters. Full-house immunofluorescence deposits are associated most commonly with lupus nephritis and less commonly with posthepatic cirrhosis, diabetic nephropathy and membranous nephropathy. There have been several reported cases of full-house nephropathy. Some of them became seropositive or manifested extrarenal lupus symptoms, and others did not. Here we report another pediatric case of full-house nephropathy presented with nephrotic syndrome.

[Clinical case]

An eight-year-old girl was admitted for generalized edema and 3 kg of weight gain in 2 weeks. One year before admission, isolated proteinuria was detected in school urine screening and were confirmed twice in a private clinic but was not further evaluated. At admission, the vital signs were stable with blood pressure of 110/77 mmHg. Her height and weight were 133 cm and 28.4 kg. The laboratory findings were as following: Hb 13.4 g/dL, Hct 39.5%, WBC 6,300/uL (P 55%, L 36%, M 6%), PLT 178k/uL, BUN/Cr 11/0.5 mg/dL, TP/Alb 4.0/2.3 g/dL, cholesterol 304 mg/dL, Ca/P 8.1/5.7 mg/dL, PT/aPTT 10.6/27.9 sec, fibrinogen 362 mg/dL, ASO (-), HBs

Ag/Ab (-/-), C3/C4 110/16.2 mg/dL, CH50 3.4 U/mL, C1q 5.6 mg/dL, C2 1.8 mg/dL, C5 16 mg/dL, IgG/IgA/IgM 311/80.6/196 mg/dL, IgE 23.6 IU/mL, ANA (+, golgi), ANCA (-), cryoglobulin (-), anti-ds DNA (-), anti-RNP (-), anti-Sm (-), anti-SSA (-), anti-SSB (-), anti-cardiolipin IgG/IgM (-/-), lupus anticoagulant (-), anti-GBM (-), Urine prot (3+), occult blood (2+), RBC 3-5/HPF, Spot urine prot/cr 9.5 mg/mg, 24 hr urine protein 206.8 mg/m^2/hr (5.1g/24hr), Ccr 87.2 mL/min/1.73m^2, urine protein electrophoresis: selective glomerular proteinuria. Renal ultrasonography : normal.

She was initially treated with calcort (2.2 mg/kg/day) under the diagnosis of nephrotic syndrome. But early renal biopsy was done on the 9th HD because of prior long-standing proteinuria and no early response to steroid. Renal pathology showed mixed membranous and membranoproliferative glomerulonephritis and full-house immunofluorescence deposits, which suggested lupus nephritis WHO class III. Methylprednisolone (30 mg/kg) was tried for 6 alternate days. Edema, proteinuria (spot urine protein/cr 2.7) and hypoalbuminemia (2.6 g/dL) improved slightly. She was discharged on daily calcort and lenipril (0.2 mg/kg) on the 30th HD. After being transfered to SNUH, azathioprine (2 mg/kg) was added. During 6 months of follow-up, her proteinuria has improved very slowly and her serologies and clinical symptoms for lupus are still negative.

[Pathologic findings]

In light microscopy, 18 glomeruli were examined which showed diffuse thickening of capillary loops and segmental double contour formation. Segmental mesangial cell proliferation with increase of mesangial matrix was also seen. Segmental glomerulosclerosis was found in 4 glomeruli. Tubules, interstium, and blood vessels were unremarkable.

Immunofluorescent staining was done in 4 glomeruli which revealed dominant mesangial IgG,

IgM, and C1q deposits, mild mesangial IgA and C4 deposits, minimal mesangial C3 and fibrinogen deposits.

In summary, the biopsy specimen showed mixed membranous and membranoproliferative glomerulonephritis and full house pattern of immunofluorescence, which suggested lupus nephritis WHO class III.

[Point of discussion]

1. Is the diagnosis, full-house nephropathy correct?

2. Full-house nephropathy will develop to lupus nephritis?

3. What is the significance of low CH50 and C1q

4. What is the most effective immunosuppressive agents?