Case 4

Recurrence of familial focal segmental glomerulosclerosis(FSGS) after renal transplantation

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[Background]

Recurrence of proteinuria or FSGS after renal transplantation was observed in approximately 30% of idiopathic FSGS patients, but its management remains controversial. While it is observed that familial FSGS recurs less frequently than idiopathic FSGS after renal transplantation, we report the case of recurrence of familial FSGS after renal transplantation.

[Case]

A 13-year-old boy was admitted to our hospital due to azotemia, proteinuria. At the age of 5 years, he visited local hospital due to abdominal pain and eyelid swelling, and was diagnosed as nephrotic syndrome. He was treated with steroid, but his symptom did not improve, and was referred to our hospital at his age of 5.4 years. Kidney biopsy showed grossly globally and segmentally sclerosing immune complex-associated glomerulonephritis. IF stainings revealed mesangial and paramesangial fine granular for C1q (1+), C4(1+), IgM(1+), IgG(1+). EM findings showed irregulary thickened glomerular basement membrane with diffuse mesangial and subendothelial, and scattered subepithelial somewhat fibrillar electron dense deposit.

He was treated with methylPD pulse and cyclophosphamide, but he didn't respond to those treatment and progressed to ESRD. He started hemodialysis at his age of 6 years and converted to peritoneal dialysis at his age of 7 years. He underwent cadaveric kidney transplantation at his age of 9 years. He was treated with immunosuppressant (mycophenolate, FK-506) and followed up regularly. Two months ago, he showed azotemia, proteinuria again and was adimitted. The second kidney biopsy showed relapsed chronic sclerosing glomerulonephritis with 80% of glomeruli being globally sclerotic, while 20% of glomeruli was visible focal scloerosis. He had been treated with immunosuppressant (mycophenolate, FK-506) continuously, and we added methylpulse therapy. But he didn't response to our treatment.

He had a younger brother who is 10 years old. His brother had asymptomatic proteinuria and microscopic hematuria from his age of 4 years. His brother underwent kidney biopsy due to

increased proteinuria at his age of 7 years. His renal biopsy revealed FSGS(26% of glomeruli was visible global sclerosis and the 11% of glomeruli was focally sclerotic). IF staining revealed focal mesangial staining of IgM(1+), IgG(1+), C1(1), C3(1+) and C4(1+). EM finding showed that glomerular basement membrane is relatively regular in thickness and contour with focal effacement of epithelial foot processes. Mesangial and subepithelial electron deposits were found. He had been treated with steroid and methylPD pulse like his elder brother, and he did neither respond. His proteinuria persists. He takes ACE inhibitor, and followed up regularly. In this case, no mutations associated with familial FSGS were revealed, but investigation of DNA analysis is doing now.

[Points of discussion]

1. Is it possible that this case is familial FSGS?
2. What is the proper management policy for recurrence of familial FSGS after renal transplantation?