Chapter 12

FOCAL SEGMENTAL GLOMERULOSCLEROSİS (FSGS)

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Focal segmental glomerulosclerosis (FSGS) is one of the most common glomerulonephritis (GNP) worldwide. It may be primary (idiopathic) or secondary to diverse etiologies. Renal biopsy is the gold standard method for the diagnosis of glomerular diseases such as FSGS. A circulating permeability factor has long been implicated in primary FSGS. The combination of glomerulomegaly and mechanical stretch from glomerular hyperfiltration may play important pathogenic roles in the development of what has been described as secondary FSGS. FSGS refers to a histological pattern of injury recognized on kidney biopsy that is characterized by sclerotic (fibrotic) lesions in glomeruli that are focal (less than 50% of all glomeruli affected on light microscopy) and segmental (less than 50% of the glomerular tuft affected). This pathological pattern has been further classified by the Columbia group according to specific pathological light microscopic findings (tip lesion, cellular, collapsing, perihilar, and not otherwise specified) which might have diagnostic and prognostic utility (Table 1) (1-3).

Especially, alterations of podocyte cytoarchitecture identified by electron microscopy, define the pathogenetic importance of podocyte injury. The distribution of the disease varies worldwide. FSGS represents 20-46.7% of cases in adults. Primary (idiopathic) FSGS is usually a progressive disorder with <5% spontaneous remission and a 50% ESRD rate over a period of 5–8 years from the time of biopsy in patients that are either unresponsive to treatment or not treated. Nephrotic-range proteinuria with or without other features of the nephrotic syndrome is the classic pattern of presentation of primary (idiopathic) FSGS and is seen in 75%–90% of children and 50%–60% of adults. Secondary forms of FSGS typically have a more chronic presentation. The recurrence of FSGS, following transplantation develops in approximately 30% (4-9).

Primary (idiopathic) FSGS is potentially treatable and curable in optimal type with duration of immunosuppresive therapy. Effective therapies slow the pace of FSGS to renal insufficiency in secondary FSGS (10, 11).

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Finally, infusion of allogeneic mesenchymal stem cells used successfully to stabilize kidney function in children with recurrent FSGS and to prevent renal dysfunction. We need more large studies about mesenchymal stem cell treatment (116).

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