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Clinicopathologic Features and Long-Term Prognosis of Lupus Podocytopathy

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Objectives : Lupus podocytopathy (LP) is a distinct subtype of lupus nephritis (LN). Despite being reported since 1980, long-term follow-up outcomes for LP patients remain unclear. This study aims to describe LP prevalence, clinicopathologic features, treatment responses and prognosis.

Methods : A retrospective study on 2489 LN patients at the First Affiliated Hospital of Sun Yat-sen University (January 1996 to September 2023). LP is defined in patients with Class I or II histology, nephrotic-range proteinuria or nephrotic syndrome, and diffuse foot process effacement, but without peripheral capillary wall immune deposits.

Results : We identified 26 LP cases (1.04%), including 20 minimal change disease (76.92%), 5 mesangial proliferative glomerulonephritis (19.23%), and 1 focal segmental glomerulosclerosis (3.85%). Median time from symptom onset to LP diagnosis was 1.6 months. Clinical symptoms included fever (19.23%), rash (34.6%), joint pain (26.9%), and edema (80.8%). Laboratory findings revealed all patients had nephrotic proteinuria or hypoalbuminemia. Pathologically, all patients exhibited moderate to extensive foot process effacement and varying degrees of mesangial and matrix cell proliferation, 1 showed leukocyte infiltration, and 4 showed Bowman's capsule adhesions. Regarding treatment, all patients received steroids, with 8 and 4 patients additionally received immunosuppressants and hydroxychloroquine respectively. Median follow-up time was 8.5 years, with a range of 7.2 to 13.6 years. The 6-month complete remission rate was 96.1%. One patient progressed to end-stage renal disease (ESRD) and initiated hemodialysis 8 years after renal biopsy, while two patients succumbed to kidney disease and sudden cardiac death at 8 and 13 years, respectively.

Conclusions : LP represents a specific subtype of LN with distinctive pathological features. These patients exhibit a short time from onset to diagnosis and severe symptom at onset. LP patients had favorable responses to immunosuppressive therapy, but some were still risking of long-term progression to ESRD or death.