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Hidden face of lupus nephritis exposed: Isolated tubulointerstitial lupus nephritis

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Systemic lupus erythematosus (SLE) is a common multisystemic immunological disorder with protean manifestations. It commonly involves the kidney, which is associated with significant morbidity and mortality. In the kidney, it typically affects glomeruli and blood vessels with secondary involvement of tubulointerstitial component. Isolated or predominant tubulointerstitial nephritis is exceedingly rare. It is essential to recognize this rare presentation of lupus nephritis, as it has significantly better outcome.

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Systemic lupus erythematosus (SLE) is a common multisystemic immunological disorder with protean manifestations (1). It may involve any organ of the body, but has predilection for certain organs and systems including the joints, skin, serosal membranes and the kidney. Kidney involvement in SLE is fairly common and is of considerable importance to the practicing nephrologists. This complication is a serious disease with guarded prognosis if not treated promptly and judiciously. Its treatment is potentially toxic, prolonged and difficult to plan and carry out (2).

Within individual organs or systems, SLE often involves more than one tissue components in variable combinations, resulting in the heterogeneity of the pathological lesions and the clinical presentations (1). Involvement of the kidney best illustrates this phenomenon. The disease preferentially targets the glomeruli and the blood vessels, but also frequently involves the tubulointerstitial component (1, 2). Isolated or predominant involvement of any single tissue element is very rare (3-6). The case report by Ali et al. (7) in this issue of J Nephropathology highlights several important points regarding this rare presentation of lupus nephritis (LN). Indeed, most literature and the classification systems focus on the glomerular involvement in LN (8). However, from long-term prognostic point of view, it is the tubulointerstitial involvement that is more important (2). The extent and the chronicity of the process are important considerations in this regard. In the present case, the changes most probably represent an acute injury rather than the chronic scarring process, which explains the reversibility of the process with treatment.

The authors are right in pointing out that this case will raise the awareness of this extremely rare complication of LN, especially in patients with known SLE who present with unusual urinary abnormalities. It is of utmost importance to accurately diagnose this entity, as the majority of previously reported cases of this entity have behaved in a benign short-term manner (3-6). Occasional cases however have relapsed or required dialysis (9). However, as with all previously reported cases, this case also reports only two year follow-up. The authors have provided immunohistochemical evidence of localization of the immune reactants in the tubular basement membranes (TBMs) and peritubular capillary basement membranes (PTCBMs), but the lack of electron microscopic (EM) study is a weak point, as this would have confidently excluded the glomerular localization of the immune deposits and conclusively demonstrated their presence in the TBMs and PTCBMs. The authors have not tested for the presence of anti-TBM antibodies either, most probably due to non-availability of the test. There is also lack of serological data on followup, which could have markedly strengthened the causal relationship of SLE with tubulointerstitial nephritis (TIN); the other causes of which were excluded from history, clinical examination and laboratory findings. Concomitant connective tissue disorders were also excluded by negative serology. The authors also briefly discuss the pathogenesis of this rare phenomenon, which is still speculative at best. Despite the above shortcomings, the authors deserve commendation for bringing such an important topic for the readership of J Nephropathology.

Since the disease is very rare, its natural history and the appropriate treatment approaches are not well recorded. Use of cytotoxic therapy is controversial (3). It is imperative to establish an international registry to accurately document the disease findings and its follow up for better understanding the long-term outcome.

Conflict of interest

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