P378

P378 -Fibrillary glomerulonephritis: A case series (a single centre experience)

Dr Zay Htet[1]
[1]*Norfolk and Norwich University Hospital NHS, Norwich, United Kingdom*

Introduction: Fibrillary glomerulonephritis (FGN) is a rare glomerular disease, accounting for 0.5-1% of native renal biopsies1. Although it is mostly a primary or idiopathic disorder, more recent studies observed some underlying associations, such as malignancy, infection, monoclonal gammopathy, and autoimmune disease2. Patients with FGN typically present with proteinuria, haematuria, and renal impairment. Randomly arranged non-branching Congo-red negative fibrillary deposits in the mesangium and glomerular basement membrane (GBM) are considered pathognomonic features on a biopsy. No evidence-based therapeutic options are available currently and that prognosis is usually guarded, with nearly half of the patients progress to end-stage renal disease (ESRD) within 2 to 4 years.
Aims: We report our experience of 4 patients with FGN, with their clinicopathologic characteristics, treatment, and outcome. This would aid to improve our understanding of the nature of this disease, and to acknowledge the challenges in treatment options and poor prognosis.
Methods: Our centre renal histology database was used to retrospectively identify patients diagnosed with FGN between 2014 and 2018.
Results: A total of 317 native kidney biopsies were performed over a 5-year period, and 4 cases of FGN (1%) were identified. All patients presented with nephrotic syndrome, or nephrotic range proteinuria, renal dysfunction, haematuria, and hypertension. All patients had stage 3 acute kidney injury at presentation (serum creatinine ranged from 360-650 µmol/L). Concomitant monoclonal gammopathy was present in one patient. All patients had mesangial and/or membranoproliferative pattern with glomerulosclerosis on light microscopy, and two had active cellular crescents. Positive IgG and C3 with negative Congo red stain, were observed in all biopsies. One patient had both clinical and histological appearance of thrombotic microangiopathy, which later found to have fibrillary deposition, though this was inconclusive due to poor sample quality. Deposition of randomly arranged fibrils in the mesangium and GBM, ranged 12.6-23 nm in diameter, were seen in three patients. Three patients received immunosuppressive therapy with an oral corticosteroid, and two of them had intravenous cyclophosphamide in combination for 3 months. One patient was treated as an atypical haemolytic-uraemic syndrome with plasma exchange and eculizumab prior to a biopsy. None of the patients achieved partial or complete remission and all progressed to ESRD within 3 months of diagnosis. Three patients were established on maintenance haemodialysis, and one received a kidney transplant.
Conclusion: Fibrillary GN is a rare diagnostic entity associated with poor renal outcomes. The degree of proteinuria and renal impairment at presentation, and the extent of glomerulosclerosis on a biopsy appear to dictate clinical response and prognosis. No evidence-based treatment recommendations are available currently, and treatment decisions for all our patients were made based on light microscopy findings. High quality randomized controlled clinical trials are urgently warranted to determine the optimal disease-specific treatment strategy.