cases of non-AL MGRS were presented with combination of light chain deposition disease and thrombotic microangiopathy. The majority of patients (80.4%) were treated with clone-specific agents. Non-IgM cases of MGRS were managed with bortezomib/melphalan/lenolidamidine–based regimen. In IgM-MGRS group (n=9) rituximab-based chemotherapy protocols were used. Autologous stem cell transplantation was performed in 13 cases. For 23 AL and 14 non-AL MGRS patients assessment of HR/RR was not available due to death before treatment initiation (n=6) or missed follow-up (n=31). HR was achieved in 74% and 80% of the treated in AL and non-AL MGRS groups, respectively (Fig.1b). RR was obtained in 42% of AL patients and in 62% of non-AL MGRS cases. Stable proteinuria without kidney function deterioration was registered in 8% of AL and in 14% of non-AL MGRS patients. The five-year cumulative renal survival did not differ significantly between the groups (Fig.1c). The cumulative renal survival in patients who did not achieve HR was significantly lower than in cases with complete HR, p=0.0014 (Fig. 1d).

Conclusions: MGRS is a clinically and morphologically heterogeneous nosological entity characterized by a poor renal prognosis, especially in the absence of clone-specific therapy. Treatment in MGRS should be carried out in a timely manner with the participation of hematologist and nephrologist in order to prevent loss of kidney function and increase life expectancy. No conflict of interest

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POLYCLONAL FREE LIGHT CHAINS IN PATIENTS WITH PRIMARY GLOMERULAR DISEASES

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Introduction: The mechanisms of kidney injury by monoclonal free light chains in multiple myeloma and other plasma cell dyscrasias are well described. The pathogenic role of polyclonal free light chains (pFLC) in primary glomerular diseases is not clearly understood. We hypothesized that increased serum level of pFLC could have special role in activation of interstitial inflammation and fibrosis formation and be associated with chronic kidney disease (CKD) progression in patients with glomerulopathies. The aim of this retrospective study was to clarify the association of pFLC kappa (pFLC-K) and lambda (pFLC-λ) assessed in serum by Freelite® with clinical and morphological parameters and CKD progression in primary glomerular diseases.

Methods: 60 patients with morphologically proven primary glomerular disease (IgA-nephropathy (n=24), minimal change disease (n=11), membranous nephropathy (n=11) and focal segmental glomerulosclerosis (n=14) were enrolled in this retrospective study. Serum pFLC-K and pFLC-λ levels were assessed in all cases at the time of kidney biopsy by Freelite® (normal range for pFLC-K 3.3-19.4 mg/l, normal range for pFLC-λ 5.7-26.3 mg/l) as well as estimated glomerular filtration rate (eGFR) by CKD-EPI and 24-hour proteinuria. Patients with abnormal k/λ ratio were excluded from analysis. Morphological findings (presented in Table 1) were semiquantitatively measured (0 - absent, 1 - mild, 2 - moderate, 3 - severe) according to currently accepted criteria. Data are presented as median and interquartile range (M (25%; 75%)) and mean and the standard error of mean (m±SEM) for semi-quantitative parameters or %. Correlation between parameters was assessed by Spearman’s coefficient. Progression of CKD was determined as decline of eGFR >25% from the initial level at the end of follow-up. Cox proportional hazards regression was used to estimate the association of pFLC with CKD progression. Differences were considered statistically significant at p<0.05. Median follow-up was 18 (3; 36) months.

Results: Clinical and morphological parameters are presented in Table 1. The levels of pFLC-K and pFLC-λ were 24.3 (16.2; 37.9) mg/l and 25.3 (19.6; 35.1) mg/l, respectively. 60% and 43% of patients had higher than normal pFLC-K and pFLC-λ, respectively. Both pFLC were elevated in 40 % of cases. FLC ratio was 0.92 (0.72; 1.12). Correlations between clinical and morphological parameters and pFLC are presented in Table 1. In univariate Cox regression analysis pFLC-k (Exp(B)=1.011; 95% CI: 1.001-1.024, p=0.023) and FLC ratio (Exp(B)=4.125; 95% CI: 1.381-12.320, p=0.011) were associated with CKD progression as well as % of sclerotic glomeruli (Exp(B)=1.024; 95% CI: 1.001-1.047, p=0.037), glomerular basement membrane segmental thickening (Exp(B)=2.486; 95% CI: 1.139-5.425, p=0.022), tubular atrophy (Exp(B)=2.311; 95% CI: 1.181-4.522, p=0.014) and interstitial fibrosis (Exp(B)=2.212; 95% CI: 1.158-4.227, p=0.016).

Conclusions: In primary glomerulopathies increased level of pFLC, both kappa and lambda, are associated with glomerular lesions, tubular atrophy, renal interstitial inflammation and fibrosis, and could be proposed as predictors of CKD progression. The mechanisms of kidney injury by pFLC requires further investigation.

No conflict of interest

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CRESCENTIC POSTINFECTIOUS GLOMERULONEPHRITIS IN AN ADULT PATIENT WITH JUVENILE NASOPHARYNGEAL ANGIOFIBROMA: A CASE REPORT

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Introduction: Crescentic glomerulonephritis is usually associated with an acute nephritic syndrome with rapidly declining renal function. Although prognosis is generally poor in most crescentic glomerulonephritis, postinfectious cases usually have a higher possibility of recovery. Juvenile nasopharyngeal angiofibroma (JNA) is a rare but benign, locally aggressive tumor affecting mostly young males. We present herein the case of a young male who with JNA and subsequently developed acute renal failure resolving after removal of the tumor.

Methods: A 28-year-old male presented at a tertiary hospital for an acute nephritic syndrome with rapidly declining renal function. The patient was initially managed as a case of acute glomerulonephritis and be associated with chronic kidney disease (CKD) progression. The mechanisms of kidney injury by pFLC requires further investigation. No conflict of interest

Conclusions: In primary glomerulopathies increased level of pFLC, both kappa and lambda, are associated with glomerular lesions, tubular atrophy, renal interstitial inflammation and fibrosis, and could be proposed as predictors of CKD progression. The mechanisms of kidney injury by pFLC requires further investigation.

No conflict of interest