MORPHOLOGY AND CLINICAL PRESENTATION OF PAUCI-IMMUNE SMALL VESSELS VASCULITIDES IN KINDEY

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Aim: Pauci-immune small vessels vasculitides in kidney, could be divided into ANCA associated vasculitides (AAV) and non-ANCA associated vasculitides (NAAV). Our aim was to examine and compare clinical and pathohistological characteristics of small vessel vasculitides in kidneys of patients with AAV and NAAV.

Methods: In this study we performed a retrospective analysis of pathohistological kidney biopsies, diagnosed between 2000-2020, at Institute for Pathology "Dr Đorđe Joannović" Faculty of Medicine University of Belgrade. Overall, 157 kidney biopsies with pauci-immune small vessels vasulitides were examined (79 patients with AAV and 78 patients with NAAV). Clinical data collected from available medical records, and pathohistological parameters were examined and compared between the AAV and NAAV patients.

Results: Table 1 summarized all investigated clinical and pathohistological parameters. ANCA positive patients were significantly older (55.5±17.6 years) than ANCA negative (45±18.1 years) patients, and they also had significantly higher frequency of glomeruli involved by crescent formations (≈40%) compared to patients without ANCA (≈30%), as illustrated in *Table 1*. Moreover, ANCA positive patients had slightly higher frequency of chronic lesions such as interstitial fibrosis, tubular atrophy and glomerular sclerosis. Laboratory parameters were high in both groups, without significant difference. However, mean CRP values were higher in ANCA positive patients. Clinically assessed BVAS score was similar in both investigated groups. The morphology of heterogenious presentation is illustrated on Figures 1 and 2.

Conclusion: Patients with ANCA positive renal vasculitis are significantly older and have higher frequency of chronic glomerular and tubulointerstitial lesion, while laboratory and clinical parameters are not significantly different between ANCA positive and ANCA negative patients.

Table 1. Clinical and pathohistological parameteres in patients with pauci-immune small vessels vasculitides in kidney.

		ANCA -	ANCA +	p
Gender n (%)	male	33 (48,5%)	35 (51,5%)	0,695
	female	46 (51,7%)	43 (48,3%)	
Age (years)		45±18,1	55,5±17,6	**<0,001**
Total crescents (%)		29,39	39,59	**0,013**
Cellular crescents (%)		7,19	9,05	0,331
Fibrocellular crescents (%)		21,9	28,06	0,136
Fibrous crescents (%)		1,11	2,49	0,159
Fibrinoid GBM necrosis (%)		30,08	33,85	0,357
Segmental glomerulosclerosis (%)		5,92	9,35	0,080
Global glomerulosclerosis (%)		7,57	7,42	0,948
Tubular atrophy (%)	+	74 (94,9)	74 (96,1)	0,712
	-	4 (5,1)	3 (3,9)	
Interstitial inflammation (%)	+	71 (49,3%)	73 (50,7%)	0,360
	-	7 (63,6%)	4 (36,4%)	
Interstitial fibrosis n(%)	+	72 (49%)	75 (51%)	0,099
	-	5 (83,5%)	1 (16,7%)	
CRP (mg/l)		29±41,96	77±65,96	0,124
Creatinine (µmol/l)		499,8±421,5	492,9±330,2	0,917
BVAS score		11,78±4,67	12,60±4,21	0,292

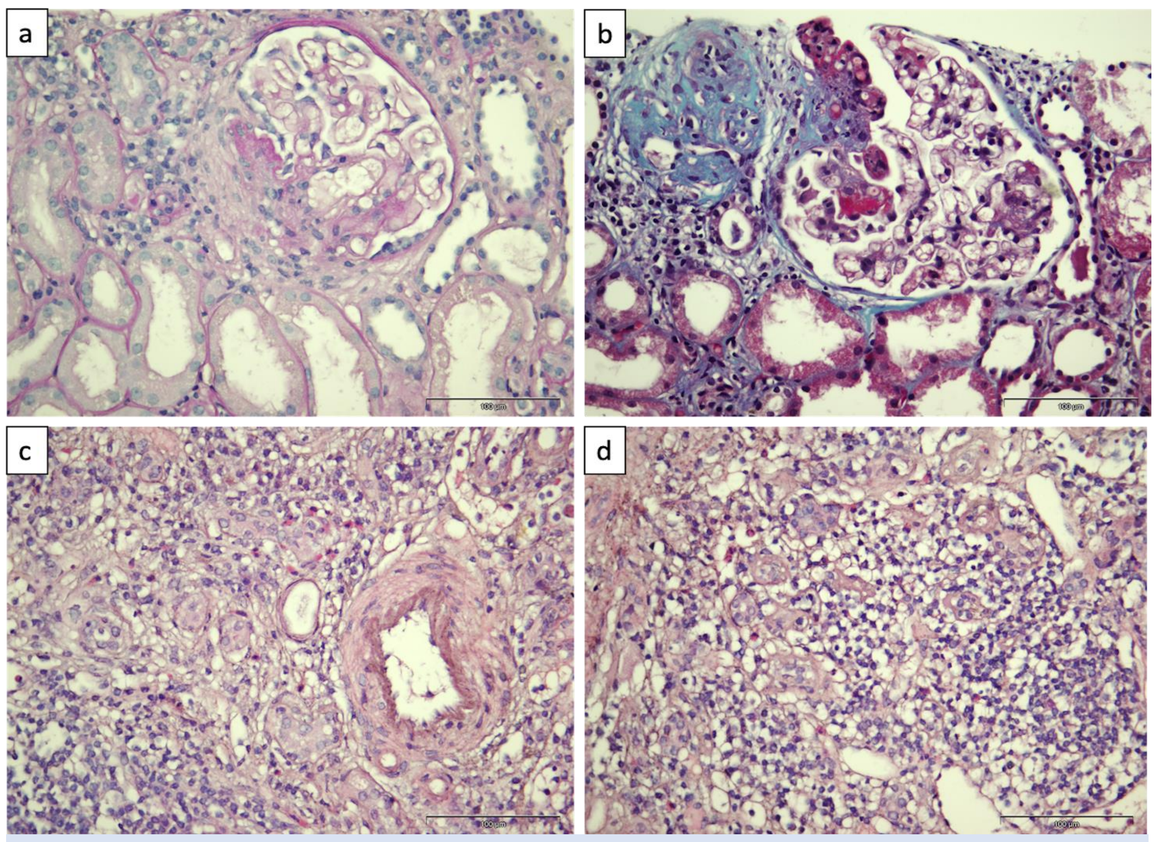

Figure 1. Interesting case of EGPA (Churg-Strauss syndrome). A) segmental fibrinoid necrosis of glomerular basement membrane (GBM) clearly visible on HE and B) MTS stained slides; C) tubulointerstitial compartment was extensively affected with diffuse infiltration of mononuclear inflammatory cells with focally prominent eosinophil-rich accumulation; eosinophils were found in a form of granulomatous eosinophil-rich inflammatory process surrounding blood vessels and D) occupying interstitial space between tubuli.

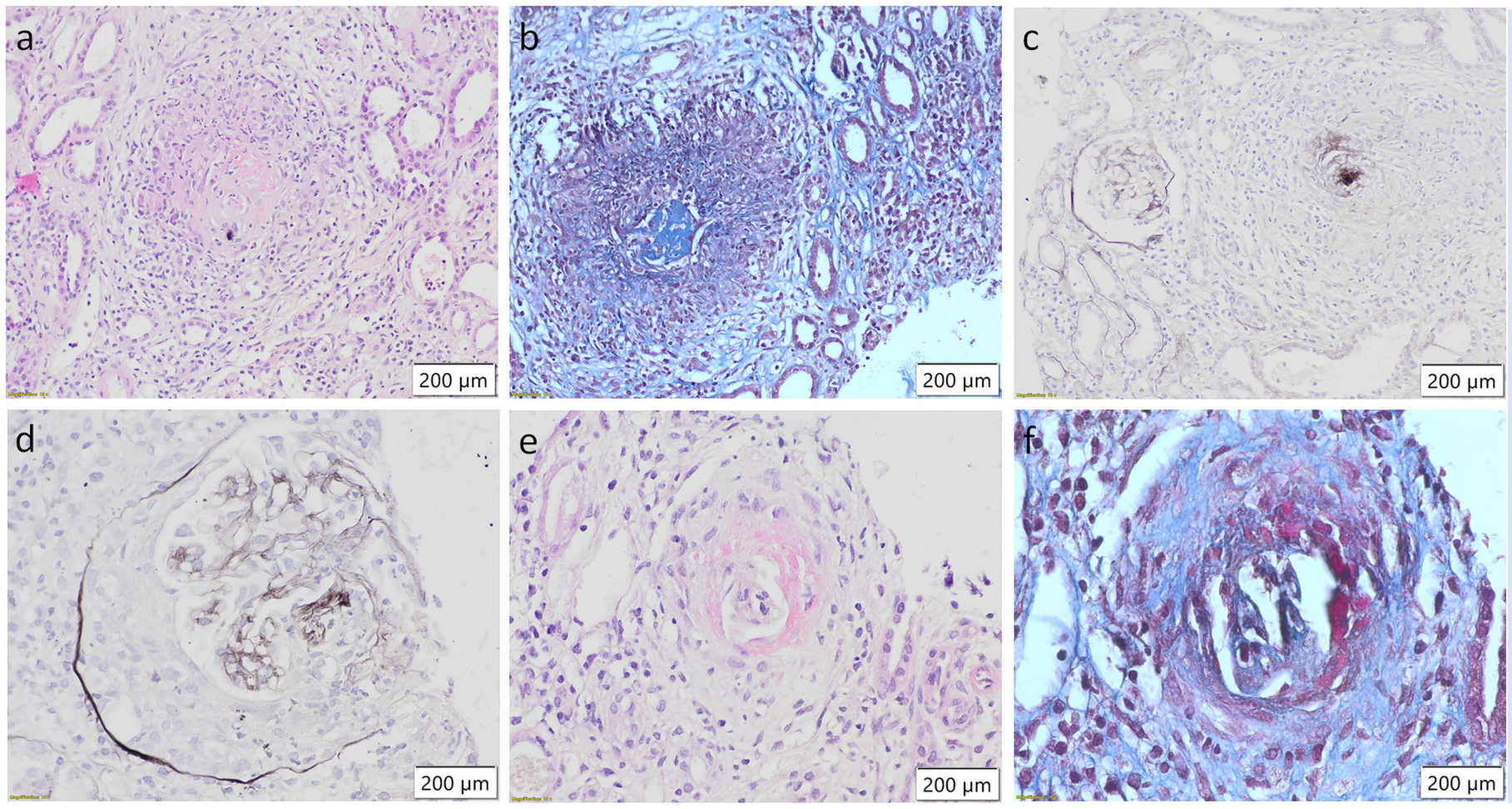

Figure 2. Interesting case of GPA (Wegener granulomathosis). A) fibrinoid necrosis with granulomatous inflammation, visible on HE slide; B) the same area stained with MTS and with C) methenamine silver stain; D) fibrinoid necrosis of GBM with crescent formation and partial distruption of Bowman capsule; E) fibrinoid necrosis of medium sized blood vessles on HE and F) MTS staining.