



ANCA Associated Vasculitis With Renal Involvement - Clinical Presentation and Long-Term Outcome

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Abstract

Background/Aim: Renal involvement is one of the most frequently manifestation of the antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis and the cause of end-stage renal disease (ESRD). The aim of this study was to analyse the types of ANCA-associated vasculitis (AAV) at University Clinical Centre of the Republic of Srpska regarding clinical presentation including long term patient's survival and renal outcome.

Methods: This study was retrospective analysis of 27 patient with diagnosis of the AAV. The diagnosis of AAV was made at University Clinical Centre of the Republic of Srpska, Banja Luka between 2007 and 2023. The clinical presentation and patients' outcomes were analysed.

Results: There were 27 patient, 14 females and 13 males, median age 57.8 years, median creatinine level at presentation 391 $\mu\text{mol/L}$. Microscopic polyangiitis (MPA) was diagnosed in 17 patients, granulomatosis with polyangiitis (GPA) in 6 and renal-limited vasculitis (RLV) in 4 patients. Patients' follow-up was from 8 months to 16 years. Cumulative survival for all patients at 1-, 3- and 5-years was 96.3 %, 92 % and 88.2 %, respectively. Cumulative renal survival for all patients at 1-, 3- and 5- years was 81.5 %, 72 % and 53.3 %. At the end of follow-up, 16 patients (59.3 %) were in complete or partial remission, 7 patients (25.9 %) were on regular dialysis and 4 (14.8 %) patients died. Relapse occurred in four patients (14.8 %). Median age of survival patients was 55.69 years and those with fatal outcome 69.75 years ($p = 0.012$). Initial higher serum creatinine level and > 50 % crescent in kidney biopsy were bad prognostic factors for ESRD.

Conclusion: AAV with renal involvement is still a severe disease. Especially if it is not detected in time and promptly treated. Mortality and renal survival in cohort are comparable to similar reports in the literature.

Key words: Anti-neutrophil cytoplasmic antibody-associated vasculitis; Long term adverse effects; Outcome; Clinical presentation.

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Introduction

Vasculitis is a clinicopathological entity characterised by inflammation and necrosis of blood vessels. Antineutrophil cytoplasmic autoantibodies (ANCA) are associated with a group of

necrotising small vessel vasculitides that have a paucity (from Latin *pauci* = few) of deposition of immunoglobulins and complement.^{1, 2} Medium vessels may occasionally be involved.

The nomenclature established in 1994 by the Chapel Hill International Consensus Conference and revised in 2012 defines ANCA-associated vasculitis (AAV) as necrotising vasculitis with few or no immune deposits, predominantly affecting small vessels (ie capillaries, venules, arterioles and small arteries).³⁻⁵ AAV is associated with ANCA specific for myeloperoxidase (MPO-ANCA) or proteinase 3 (PR3-ANCA). The typical renal biopsy feature in AAV is pauci-immune necrotising glomerulonephritis (GN) with crescents formation.

On the basis of pathologic and clinical features, AAV is subdivided into microscopic polyangiitis (MPA), granulomatosis with polyangiitis (GPA) (previous Wegener granulomatosis), eosinophilic granulomatosis with polyangiitis (EGPA) (previous Churg–Strauss syndrome) and renal-limited vasculitis (RLV). GPA is a necrotising vasculitis of the small and medium-sized vessels associated with granulomatous inflammation of upper and lower respiratory tract and the frequent finding of GN. In active disease, 90 % of cases are PR3-ANCA positive. MPA is a necrotising angiitis involving capillaries, venules and arterioles of one or more organs either simultaneously or at different times, including the kidneys, lungs, skin, spleen, liver, heart and muscle.^{2, 6} Typically, MPO-ANCA are positive. The symptoms of the upper respiratory tract are usually less often and milder compared to GPA and there is no granulomatous inflammation. EGPA is characterised by vasculitis, granulomatous inflammation of the respiratory system, eosinophilia and asthma. Kidney involvement is relatively rare (10-25 % of cases). RLV is characterised with pauci-immune necrotising GN alone with no evidence for systemic vasculitis. There is no respiratory disease or any other organ involvement.

The kidney is the most commonly affected vital organ in AAV and patient's outcomes are largely determined by the severity of renal disease at diagnosis and its response to treatment.⁷ As mentioned before, AAV can be limited to the kidney only, or may involve the upper respiratory tract, lungs, skin and other organs in various combinations.^{8, 9} At least 50 % of patients with ANCA-associated GN have pulmonary disease and 10 % of them have pulmonary haemorrhages.⁷ Rapidly progressive GN is the most frequent clinical manifestation of ANCA-associated GN. Renal involvement is defined by the presence of

haematuria (30 red blood cells/high-power field) with or without red blood cell casts and elevated creatinine level attributable to the disease, or histological evidence of pauci-immune necrotising GN, while proteinuria is usually moderate (2–3 g/day).¹⁰

The annual incidence of renal vasculitis in Europe is 10 to 20 per million per year. GPA is more common in northern Europe and MPA in southern Europe.⁸ There is an increased incidence with age, peaking in those aged 65 to 74 years.¹¹

Currently, it is not clear whether there is any difference between GPA, MPA, EGPA and RLV with regard to the response to treatment.^{12, 13} Untreated, systemic vasculitis is associated with high mortality. The introduction of corticosteroids, cyclophosphamide and azathioprine led to a marked improvement in survival to 84 % and 76 % at 1 and 5 years, respectively. Predictors of death include age > 65 years, creatinine level at presentation, extent and severity of disease at diagnosis, pulmonary haemorrhage and treatment-related infection.^{6, 10, 14-17} The presence of pulmonary haemorrhage is the most important determinant of patient survival and the serum creatinine level is the most important prognostic marker for long-term renal outcome.¹⁸ Histopathologic features of chronic renal scarring (less than 20 % of normal glomeruli at kidney biopsy, severe interstitial fibrosis and tubular atrophy) have consistently been associated with poor renal outcomes.

The aim of this study was to analyse the types of AAV at University Clinical Centre of the Republic of Srpska regarding clinical presentation including long term patient's survival and renal outcome.

Methods

University Clinical Centre of the Republic of Srpska provides nephrology services for 1.2 million people. Between April 2007 and December 2023 at Department of Nephrology 27 patients were newly diagnosed with ANCA-associated vasculitis with the evidence of renal involvement according Chapel Hill International Consensus Conference.^{4, 5, 10} There were 17 (63 %) patients with MPA, 6 (22.2 %) with GPA and 4 (14.8 %) had RLV. There were no patients with EGPA.

Treatment

Treatment protocol included:

1. Induction

A. Cyclophosphamide (CYC) pulse intravenous - all 27 patients (mainly 1x monthly 6 months) 15 mg/kg body weight (A 25 % reduction in CYC dose was made for those aged over 65 years and for a glomerular filtration rate under 50 mL/min).

A1. CYC pulse + methylprednisolone pulse (500 mg iv three consecutive days), later prednisolone *per os* 1 mg/kg 4 - 6 months, then tapered prednisolone to 10 mg per day - 17 patients.

A2. CYC pulse + prednisolone *per os* - 10 patients.

B. plasmapheresis - 6 patients (mainly for patients with pulmonary haemorrhage or severe renal failure).

2. Maintenance therapy

- Azathioprine (2 mg/kg than 1.5 mg/kg) plus prednisolone 12-30 months (mainly 18-24) - 17 patients.

- Mycophenolate mofetil 2 g daily - 2 patients

- CYC pulses 1000 mg iv every 3 months - total 1 year - 2 patients.

The maintenance therapy was not given for patients who had definitely end stage renal disease (ESRD) and didn't have other serious disease manifestation.

3. Therapy of relapse

CYC pulse - 3 patients and mycophenolate mofetil - one patient.

Rituximab was not available for patients.

4. Other therapy

Trimethoprim sulphametoxazol mainly to prevent the occurrence of *Pneumocystis carinii* infection, upper respiratory tract infection and subsequent relapse of disease.

Uromitexan was prescribed with CYC for prevention haemorrhagic cystitis.

Statistical analysis was performed using IBM SPSS Statistics 21. Continuous variables are presented as median and interquartile range (IQR) or mean $\pm$ standard deviation, depending on data distribution. Categorical variables are presented as absolute numbers and percentages. The normality of distribution was assessed using appropriate tests. For comparison between two groups, the Mann-Whitney U test was used for

non-normally distributed variables, while the independent samples t-test was used for normally distributed variables. Associations between categorical variables were analysed using the Chi-square test or Fisher's exact test when appropriate. Kaplan-Meier survival analysis was used to assess patient and renal survival. A p-value < 0.05 was considered statistically significant.

Results

Demographic and clinical data

There were 13 female and 14 males. The overall median age at diagnosis was 57.8 (23-76) years. The median age for patient with MPA was 60.9, for those with GPA 55 and for patients with RLV 48.8 years. There was no statistical difference between these three groups. Median follow up for all patients was 66.4 months (59 for MPA, 90 for GPA and 50.2 for RLV) and there was no statistical difference between three groups. Patients were tested for ANCA by immunofluorescence and ELISA. ANCA was detected in all patients. MPO-ANCA was positive in 20 patients and PR3-ANCA in 7 patients, three patients were both MPO- and PR3-ANCA positive. Organ involvement at presentation is summarised in Table 1. By definition, all patients had renal involvement, which was biopsy-proven in 77.8 % patients.

There was no statistical difference between MPA, GPA and RLV regarding age, gender, level of proteinuria, initial creatinine level, presence of microhaematuria and acute haemodialysis at diagnosis. The patients with GPA had statistical significantly higher involvement of eyes and upper respiratory tract. Except patients with RLV, all others patients had general symptoms ("flu-like syndrome") and loss of body weight.

Lung involvement was most often clinically manifested as pulmonary haemorrhage. Two patients were on artificial ventilation. Upper respiratory tract symptoms in patient with GPA were rhinosinusitis. Repeated rhinosinusitis caused loss of nasal cartilage ("saddle nose") in one patient. All patients with nervous system symptoms had mono- or polyneuritis. Median initial creatinine level overall was 387.4 μ mol/L and there was no statistical difference between three groups. At diagnosis, 5 patients were on haemodialysis. Kidney biopsy was performed in 21 patients and 13 patients had crescents in > 50 % of glomeruli.

Table 1: Demographic and clinical data of patients with antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis

Parameters	Group of respondents				*p-value
	Overall	MPA	GPA	RLV	
Number of patients (%)	27 (100.0 %)	17 (63.0 %)	6 (22.2 %)	4 (14.8 %)	< 0.050
Age (years) (mean ± SD)	57.8 ± 14.0	60.9 ± 14.2	55.0 ± 15.9	48.8 ± 6.3	0.266
Female	13 (48.1 %)	10 (37.0 %)	2 (7.4 %)	1 (3.7 %)	0.339
Male	14 (51.9 %)	7 (25.9 %)	4 (14.8 %)	3 (11.2 %)	
Proteinuria (g/24 h)	2.276	2.239	2.073	2.740	0.774
Initial creatinine level (µmol/L)	387.4	384.7	375.8	416.5	0.949
Acute haemodialysis et diagnosis	5 (18.5 %)	3 (17.6 %)	1 (16.7 %)	1 (25.0 %)	0.415
Microhaematuria	25 (92.6 %)	15 (88.2 %)	6 (100.0 %)	4 (100.0 %)	0.530
Organ involvement					
Renal involvement	27 (100.0 %)	17 (100.0 %)	6 (100.0 %)	4 (100.0 %)	-
Lung	10 (37.0 %)	7 (41.2 %)	3 (50.0 %)	-	0.233
Upper respiratory tract	3 (11.0 %)	-	3 (50.0 %)	-	0.003
Skin	5 (18.5 %)	4 (23.5 %)	1 (16.7 %)	-	0.547
Arthralgias	4 (14.8 %)	3 (17.6 %)	1 (16.7 %)	-	0.664
Peripheral nervous system	5 (18.5 %)	4 (23.5 %)	1 (16.7 %)	-	0.547
Gastrointestinal system	5 (18.5 %)	5 (29.4 %)	-	-	0.165
Eyes	2 (7.4 %)	-	2 (33.3 %)	-	0.023
Lymphadenopathy	3 (11.1 %)	2 (11.8 %)	1 (16.7 %)	-	0.707
Kidney biopsy	21 (77.8 %)	12 (70.6 %)	5 (83.3 %)	4 (100.0 %)	0.415

MPA: microscopic polyangiitis; GPA: granulomatosis with polyangiitis; RLV: renal-limited vasculitis; SD: standard deviation;

Adverse events are shown in Table 2.

Table 2: Adverse event of patients with antineutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis

Adverse event	N	%
Infections	8	29.6
Sepsis	1	3.7
Bacterial (urinary tract infection, pneumonia)	6	22.2
Viral (Herpes zoster)	1	3.7
Cardiovascular	4	14.8
Cardiac failure	2	7.4
Pulmonary embolus	1	3.7
Cerebrovascular event	1	3.7
Malignancy	1	3.7
Thyroid cancer	1	3.7

Remission

Complete remission: The absence of symptoms or signs attributable to active vasculitis, with a Birmingham Vasculitis Activity Score of 0-1.¹⁹ The absence of renal disease activity was indicated by stable or falling creatinine levels and the absence of erythrocyte cell casts.⁸ Partial remission: Clear-cut suppression of the disease, with improvement or stopping in disease progression

and stabilisation of renal function. Complete or partial remission after induction therapy in this study was achieved in 22 patients (81.5 %). The remission persisted on the end of the follow up period in 16 patients.

Patient survival

Overall survival was better in younger patients (Table 3). Cumulative survival of all patients at 1-, 3- and 5-years was 96.3 %, 92 % and 88.2 %, respectively. Cumulative survival of patients with MPA at 1-, 3- and 5-year was 94.1 %, 87.5 % and 81.8 %. Patients with GPA and RLV had cumulative survival at 1-, 3- and 5-years 100 %.

Four patients (14.8 %) died. Causes of death were sepsis (one patient), pulmonary embolism (one patient) and ischaemic cardiovascular disease (two patients). All patients who died had MPA. There were no deaths in the GPA and RLV groups. However, differences in outcomes between groups were not statistically significant (Table 4).

There was no statistically significant difference regarding mortality between group of patients with and without pulmonary manifestation of disease and initial serum creatinine level was not prognostic factor for patient’s survival.

Table 3: Difference in average age between patients who died and surviving patients (t-test for independent samples)

Outcome	N	Average age	SD	p-value
Died	4	69.75	6.70	0.012
Remission or dialysis	23	55.69	14.01	
Overall	27	57.78	14.04	

SD: standard deviation;

Table 4: Outcome of disease (at the end follow up)

Outcome	MPA (n = 17)	RLV (n = 4)	GPA (n = 6)	Overall
Death	4 (23.5 %)	0 (0.0 %)	0 (0.0 %)	4 (14.8 %)
ESRD (dialysis)	5 (29.4 %)	1 (25.0 %)	1 (16.7 %)	7 (25.9 %)
Remission	8 (47.1 %)	3 (75.0 %)	5 (83.3 %)	16 (59.3 %)

$\chi^2 (4) = 3.81, p = 0.432$; MPA: microscopic polyangiitis; GPA: granulomatosis with polyangiitis; RLV: renal-limited vasculitis; ESRD: end-stage renal disease;

Renal survival

At the end of follow up period 7 patients (25.9 %) had ESRD and were dialysis dependent. It is characteristic that 4 patients developed ESRD in the first 7 months of the diagnosis. Patients who developed ESRD and were dependent on haemodialysis had higher initial creatinine levels than those who didn't need dialysis (Table 5).

Table 5: Initial serum creatinine level as prognostic factor for ESRD (dialysis dependent) (t-test)

Outcome	Number patients	Average serum creatinine (μmol/L)	SD	p-value
No dialysis	20	318.4	139.1	0.001
Dialysis	7	584.7	204.9	
Overall	27	387.4	194.9	

ESRD: end-stage renal disease; SD: standard deviation;

Table 6: Crescent formation as prognostic factor for ESRD (dialysis dependent) (t-test)

Outcome (without death)	Overall	> 50 % crescents	< 50 % crescents	p-value
Dialysis	7	6	1	0.039
No dialysis	11	4	7	
Total	18	10	8	

ESRD: end-stage renal disease;

Crescents on kidney biopsy had prognostic value. Patients with > 50 % crescent had worse outcome (Table 6).

Cumulative renal survival for all patients at 1-, 3- and 5- years was 81.5 %, 72 % and 53.3 % (Figure 1). Cumulative renal survival for patients with MPA at

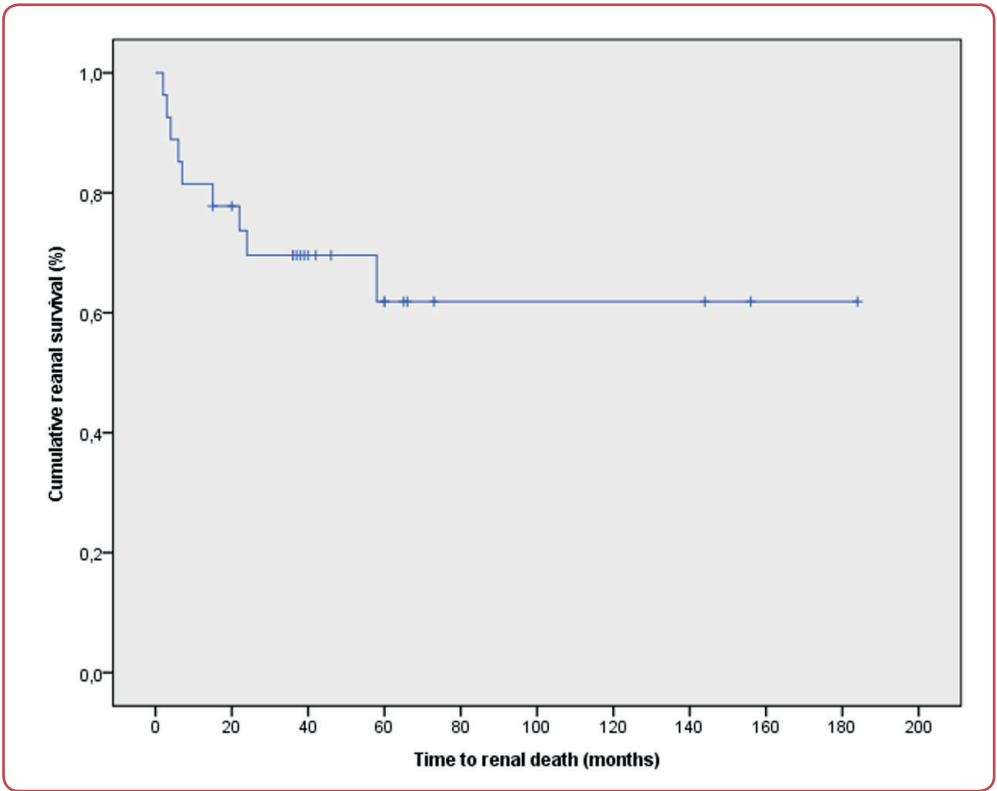


Figure 1: Cumulative renal survival for all patients (Kaplan Meyer analysis)

1-, 3- and 5-years was 76.5 %, 68.8 % and 44.4 %. Patients with GPA had cumulative renal survival at 1-, 3- and 5-years 100 %, 80 % and 75 % and those with RLV 75 %, 75 % and 50 %, respectively.

Relapse

Renal relapse was defined as recurrence of haematuria with or without increased proteinuria or an increase in the serum creatinine in patients who achieved remission. Relapse occurred in four patients (14.8 %), two patients with GPA and two patients with MPO. One patient with GPA had two relapses.

Discussion

Most frequent clinical manifestation of AAV in our study was MPA (63 %), followed by GPA (22.2 %) and RLV (14.8 %). MPA was the most frequent AAV in studies by Jennette and Nachman³ and Cisternas et al²⁰ where the percent of MPA within AAV is 49-52.8 %. In Chinese population this percent is even higher, 92.8 % in study by Hong et al¹³ and 78.8 % in study by Kim SM et al.²¹ In some study the most frequent AAV was GPA.^{22,23}

In study by Weidner et al the patients' average age for MPA was 66.8 years, for RLV 64.7 and for GPA 54.7 years.²³ Patients were younger in study by Jennette and Nachman, MPA 57.5 years, RLV 59.8 and GPA 47.9 years.³ In this study the patients' average age for MPA was 60.9 years, for GPA 55 and RLV 48.8 years.

Renal involvement in RLV is 100 % and in MPA and GPA varies between 68 % - 100 %.^{10,13,20} All presented patient had renal disease. Lung involvement is more frequent in patients with GPA than MPA, in study by Jennette and Nachman 81.2 % vs 59.5 %, in study by Cisternas et al 62 % vs 28 %, in study by Weidner et al 43.7 vs 28.6 %²³ and in presented study 50 vs 41.2 %.

Upper respiratory tract involvement is characteristic of GPA and varies 28.5 %-57 %.^{13,23,24} In this study it was 50 %. Peripheral nervous system involvement is more frequent in MPA than GPA, in our study 23.6 % vs 16.7 % and in study by Weidner et al 39.3 % vs 9.3 %.²³ In study by Cisternas et al only MPA group had the peripheral nervous system involvement.²⁰ Eye involvement is most frequent in GPA, 24.2 %-26 %.^{13,23,24} In this

study two patients had the eye involvement, both with GPA. Cutaneous manifestations are more frequent in patients with GPA than MPA, in study by Rihova et al 24.2 % vs 20 %²² and in presented study 23.5 % vs 16.7 %.

Percentage of relapses in presented study was 14.8 %. This is lower than a data from literature, 33 % according to Weidner et al²³ and 44.7 % in study by Rihova et al.²²

Patients with AAV have a very poor outcome if the vasculitis is not diagnosed and treated promptly. The introduction of immunosuppressive therapy has improved patient survival dramatically, with over 90 % of patients achieving remission compared to a mortality of 80 % at 1 year in untreated patients. Mortality is highest in the first year after diagnosis, but the excess risk of mortality persists also after the first year from diagnosis.¹⁴ In presented cohort the mortality rate was 14.8 % (4 patients). Two patients died within the first year of diagnosis (one from sepsis and one from pulmonary embolism). Two other patients died later due to ischaemic cardiovascular disease.

Median follow up in this study was 66.6 ± 42.6 months. On the end of the follow up 14.8 % patients died, 25.9 % had ESRD (dialysis dependent) and 59.3 % patients were in remission or had chronic kidney disease (CKD). These results are similar as results in study by Weidner et al²³ and Binda et al.²⁴

In presented study 1-year survival was very good, 96.3 %. Similar results had Galešić et al, 94.2 %.²⁵ Lower 1-year survival was found in a study by Weidner et al, 86 %²³ and Booth et al 82%.¹⁰

In different studies, 5-year-patient-survival with AAV was 76 %-81 %.^{10,23,24} This study got better results with 5-year-patient-survival 88.2 %.

Weidner et al found higher mortality in GPA patients vs MPA patients (35 % vs 6 %).²³ This is opposite to presented results. In this study, 4 patients (14.8 %) died and all had MPA. In study by Galešić et al 1-year renal survival was 70.6 %²⁵ and in this study, it was 81.5 %. Five-year renal survival in study by Rihova et al was 69.2 %²² and in this study, it was 53.3 %. Patients who developed ESRD had higher initial serum creatinine level and > 50 % crescents on kidney biopsy. This is consistent with literature data.¹⁸

Conclusion

AAV with renal involvement is still a severe disease, especially if it is not timely detected and promptly treated. In presented study, mortality and renal survival were comparable to similar reports in the literature. In view of the severity of the renal disease and the length of follow-up, mortality and renal survival in presented patients was good.

Ethics

The study was approved by the Ethics Committee of the University Clinical Centre of the Republic of Srpska, Banja Luka, The Republic of Srpska, Bosnia and Herzegovina under approval number No: 01-19-178-2/26, dated 17 April 2026.

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Conflicts of interest

The authors declare that there is no conflict of interest.

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Data access

The data that support the findings of this study are available from the corresponding author upon reasonable individual request.

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