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Research Article

Impact of Various ANA Results and ANCA Serotypes on the Outcome of Patients with ANCA-Associated Vasculitis

ANCA İlişkili Vaskülitli Hastaların Sonuçlarına Çeşitli ANA Sonuçlarının ve ANCA Serotiplerinin Etkisi

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Aim: We aimed to investigate how antinuclear antibody (ANA) and antineutrophil cytoplasmic antibody (ANCA) serotypes affect the outcomes of patients with ANCA-associated vasculitis (AAV).

Material and Method: This retrospective cohort analysis analyzed 10726 Turkish tertiary hospital serum samples over 4.5 years. ANCA indirect immunofluorescence test (IIF) and ANCA Enzyme-Linked Immunosorbent Assay (ELISA) testing were requested for all patients.

Results: Among 10726 patients, there were 920 patients with positive ANCA IIF and/or ANCA ELISA test. Of the 920 patients, 18 of the 98 patients with AAV were concomitant connective tissue diseases, so 80 AAV patients were included in the study. Organ involvement in these very rare patients was pulmonary (63.8%), ear-nose-throat (43.8%), renal (38.8%), joint (38.8%), skin (20%), neurological (20%), ocular (17.5%), gastrointestinal (11.3%), and cardiac (2.5%) systems. Proteinuria (51.3%), renal biopsy (48.8%), hematuria (37.5%), hypertension (56.3%), chronic renal failure (36.3%), diabetes (21.3%), coronary artery disease (10%), and asthma (8.8%) were present. ANA was found in 42.5% of 80 patients. Renal involvement and coronary artery disease were more common in ANA-positive patients (p=0.025 and p=0.05). ANA-negative patients had higher ear, nose, and throat involvement (p=0.026). Ocular involvement was significantly more frequent in PR3-ANCA patients (p=0.020). In MPO-ANCA individuals, asthma was more common (p=0.002).

Conclusion: Patients with AAV and ANA had greater renal impairment compared to those without. Renal biopsy is recommended to distinguish AAV from systemic lupus erythematosus. Ocular involvement is more prominent in PR3-ANCA. The effects of ANA and ANCA serotypes in AAV patients need more study.

Keywords: Antinuclear antibody; Antineutrophil cytoplasmic antibody; Vasculitis; Organ involvement.

Amaç: Bu çalışmada, antinükleer antikor (ANA) ve antinötrofil sitoplazmik antikor (ANCA) serotiplerinin ANCA ile ilişkili vaskülit (AAV) hastalarının sonuçlarını nasıl etkilediğini göstermeyi amaçladık.

Gereç ve Yöntem: Bu retrospektif kohort analizi, 4,5 yıl boyunca üçüncü basamak bir hastanede 10726 serum örneğini analiz etmiştir. Tüm hastalardan ANCA indirekt immünofloresan testi (IIF) ve Enzim bağlı immünosorbent analiz (ELISA) istenmiştir.

Bulgular: Bulgular: 10726 hasta arasında, ANCA IIF ve/veya ANCA ELISA testi pozitif olan 920 hasta vardı. 920 hastanın 98'i AAV hastasıydı ve bunların 18'i eşlik eden bağ dokusu hastasıydı, bu nedenle 80 AAV hastası çalışmaya dahil edildi. Çok nadir görülen bu hastaların organ tutulumu pulmoner (%63,8), kulak-burun-boğaz (%43,8), renal (%38,8), eklem (%38,8), deri (%20), nörolojik (%20), oküler (%17,5), gastrointestinal (11,3%) ve kardiyak (2,5%) olmuştur. Proteinüri (%51,3), böbrek biyopsisi (%48,8), hematüri (%37,5), hipertansiyon (%56,3), kronik böbrek yetmezliği (%36,3), diyabet (%21,3), koroner arter hastalığı (%10) ve astım (%8,8) mevcuttu. ANA, 80 hastanın %42,5'inde saptandı. ANA pozitif hastalarda böbrek tutulumu ve koroner arter hastalığı daha yaygındı (p=0.025 ve p=0.05). ANA negatif hastalarda kulak, burun ve boğaz tutulumu daha yüksekti (p=0.026). PR3–ANCA'lı bireylerde göz tutulumu daha yüksekti (p=0.020). MPO-ANCA bireylerde astım daha yaygındı (p=0.002).

Sonuç: Sonuç: ANA'lı AAV hastalarında, ANA'sız hastalara göre böbrek yetmezliği daha yaygındı. AAV'yi sistemik lupus eritematozustan ayırt etmek için böbrek biyopsisi önerilir. Göz tutulumu PR3-ANCA'da daha yüksektir. AAV hastalarında ANA ve ANCA serotiplerinin etkileri daha fazla araştırılmalıdır.

Anahtar Kelimeler: Antinükleer antikor; Antinötrofil sitoplazmik antikor; Vaskülit; Organ tutulumu.

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INTRODUCTION

Antineutrophil cytoplasmic antibodies ANCA are autoantibodies that target the lysosomal granules present in neutrophils and monocytes (1). Since its initial determination in 1982 in patients with necrotizing glomerulonephritis, ANCA testing has been employed for diagnosing, excluding, or monitoring small vessel vasculitides, namely ANCA-associated vasculitides (AAV) (2). Furthermore, ANCA testing is requested when there is a suspicion of non-AAV (3). ANCA tests are performed utilizing two methodologies: indirect immunofluorescence (IIF) and enzyme-linked immunosorbent assay (ELISA). The IIF method, utilizing a substrate derived from peripheral blood granulocytes, enables the assessment of the binding pattern of ANCA to granulocytes. Ethanol and formalin-fixed granulocytes are utilized as ANCA substrates for this purpose (4).

Patients with c-ANCA targeting PR3 are more likely to exhibit symptoms associated with the ear, nose, eyes, and lungs; however, renal involvement is not consistently observed. They show a heightened tendency for relapse and frequently have a worse prognosis, along with a higher mortality rate (5). Patients with p-ANCA targeting MPO typically show distinct form of renal inflammation known as pauci-immune segmental necrotizing glomerulonephritis. This disease may manifest itself independently or in conjunction with other systemic symptoms. These individuals generally experience fewer problems with their ears, nose, eyes, and lungs, and are less likely to experience recurrence (6).

ANA may contribute to organ or tissue damage through several mechanisms, including immune complex deposition, cytokine-mediated stimulation, and receptor binding. Previous studies have reported that 18–66% of AAV patients are positive for serum ANA; however, the clinical and pathological significance of this finding remains unclear (7,8).

In this study, we aimed to investigate the impact of different ANA patterns and ANCA serotypes on clinical outcomes in Turkish AAV patients.

MATERIALS AND METHODS

Study Design and Data Collection

This study retrospectively assessed 10726 blood samples submitted for ANCA testing to the Ankara Bilkent City Hospital Medical Microbiology Laboratory from February 2019 to July 2023. The patient population and baseline evaluation comprised eligible individuals diagnosed with AAV at Ankara Bilkent City Hospital from February 2019 to July 2023. All patients met the subsequent criteria: 1) Compliance with the AAV classification criteria established by the American College of Rheumatology in 1990 or the International Chapel Hill Consensus Conference in 2012 (9, 10) 2) Age ≥18 years individuals with any of the following circumstances were excluded: with other comorbidities, connective tissue disorders; ANCA antibody negative or doubly positive.

If there were multiple samples from the same patient, the first serum sample from the patient was included in the study, while the other samples were excluded. Among 10726 patients, there were 920 patients with positive ANCA IIF and/or ANCA ELISA test. Of the 920 patients, 18 of the 98 patients with AAV were concomitant connective tissue patients, so 80 AAV patients were included in the study. A total of 80 AAV serum samples were sent, primarily from the departments of rheumatology, nephrology, neurology, gastroenterology, pulmonology, internal medicine, and others. AAV is a relatively rare disease with an estimated prevalence of 200–400 cases per million people. Since it is a very rare disease, we only recorded 80 AAV patients who requested ANA tests in the clinical records of our 4000-bed hospital. 59 patients have granulomatous polyangiitis (GPA), 13 have microscopic polyangiitis (MPA), and 8 have eosinophilic granulomatous polyangiitis (EGPA). The Ankara Bilkent City Hospital Non-Interventional Clinical Research Ethics Committee approved this study (Date: 12.07.2023 and Decision no: E2-23-4480). The study was carried out in conformity with the Helsinki Declaration and publishing ethics.

The hospital records were used to obtain data on immunoglobulin (Ig) G, Ig A, Ig M, platelets, neutrophils, lymphocytes, hemoglobin, complement 3 (C3), C4, initial creatinine, initial glomerular filtration rate (GFR), erythrocyte sedimentation rate (ESR), and rheumatoid factor (RF). These values were measured at the same time as the ANA evaluation or within one week after. Additionally, the records provided clinical data such as age, gender, and diagnosis.

ANCA and ANA testing

Sera were tested for the presence of ANCA using IIF-based ANCA testing (Euroimmun, Lubeck, Germany) and anti-PR3 and anti-MPO using ELISAs (Euroimmun, Lubeck, Germany). The ANCA diagnostic kit based on IIF contains ethanol-fixed neutrophils as well as formalin-fixed neutrophil substrate and ethanol-fixed Hep-2-neutrophil substrate. Patient sera were diluted one-tenth and applied to the substrate wells before being incubated. These antibodies

bind to antigens on fixed neutrophils in the presence of ANCA-containing serum. Fluorescence microscopy is then used to observe the reaction with FITC-labeled anti-human antibodies. Reagent wells in ELISA kits are coated with a mixture of purified recombinant PR3 and natural PR3, as well as purified MPO antigens. Both PR3 and MPO ELISA have a cutoff value of 20 relative units (RU)/mL. The indirect immunofluorescence method, which is the gold standard method for ANA detection, was used.

Statistical analysis

Statistical analyses were performed using the Statistical Package for Social Sciences (SPSS) for Windows, version 22.0 (IBM Corp., Chicago, IL, USA). The Kolmogorov–Smirnov test was used to assess the normality of data distribution. Normally distributed continuous variables were expressed as mean ± standard deviation (SD), whereas non-normally distributed variables were presented as median (interquartile range). Comparisons between two groups were performed using the independent samples t-test for normally distributed variables and the Mann–Whitney U test for non-normally distributed variables. Categorical variables were compared using the Chi-square test or Fisher's exact test, as appropriate. A p-value <0.05 was considered statistically significant.

RESULTS

Eighteen patients were excluded due to the presence of concomitant connective tissue diseases (n=11) or ANCA negativity/dual positivity (n=7). The remaining 80 patients were included in the analysis and followed for a mean duration of 15 months (range, 0.3-52 months). Baseline characteristics according to ANA status are presented in Table 1. The mean age at diagnosis was 53.6±15.7 years. Forty-two patients (52.5%) were female, and 70 patients (87.5%) were involved of more than one organ at diagnosis.

Organ involvement included pulmonary (63.8%), ear–nose–throat (43.8%), renal (38.8%), joint (38.8%), skin (20%), neurological (20%), ocular (17.5%), gastrointestinal (11.3%), and cardiac (2.5%) systems. Additional findings included proteinuria (51.3%), renal biopsy (48.8%), and hematuria (37.5%). Comorbid conditions were hypertension (56.3%), chronic renal failure (36.3%), diabetes mellitus (21.3%), coronary artery disease (10%), and asthma (8.8%).

ANA positivity was detected in 42.5% of patients. The mean age of ANA-positive and ANA-negative patients was 54.1±16.8 years and 52.4±15.3 years, respectively, with no significant difference (p=0.647). Female sex was observed in 46.2% of ANA-positive patients (p=0.519). Renal involvement and coronary artery disease were significantly more frequent in ANA-positive patients (p=0.014 and p=0.050, respectively), whereas ear–nose–throat involvement was more common in ANA-negative patients (p=0.026).

Table 1. Baseline clinical characteristics and laboratory data of ANCA-associated vasculitis patients according to ANCA type.

	ANA (-)	ANA (+)	P value
Demographic			
Female, n (%)	53.8%	46.2%	0.519
Age, years	52.4±15.3	54.1±16.8	0.647
Organ involvement			
Lungs	67.4%	58.8%	0.431
Kidney	28.3%	52.9%	0.025
Ear, nose, throat	54.3%	29.4%	0.026
Joints	34.8%	44.1%	0.397
Skin	15.2%	26.5%	0.214
Gastrointestinal	15.2%	5.9%	0.191
Ocular	17.4%	17.6%	0.976
Neurological	19.6%	20.6%	0.910
Cardiac	0	5.9%	0.096
Laboratory data			
Ig M (g/L)	0.630 (0.440-0.778)	0.686 (0.496-1.24)	0.355
Ig G (g/L)	10.7 (6.20-11.9)	10.5 (6.92-16)	0.313
Ig A (g/L)	1.73±0.946	1.79±0.870	0.802
Platelets($\times10^9$/L)	277 (228-392)	287 (203-325)	0.448
Neutrophils($\times10^9$/L)	6.61 (4.02-8.77)	5.28 (3.19-7.54)	0.164
Lymphocytes($\times10^9$/L)	1.35 (0.980-1.75)	1.29 (0.860-1.61)	0.431
Hemoglobin(g/L)	10.8±2.44	10.9±2.28	0.909
C3(g/L)	1.26±0.368	1.27±0.310	0.945
C4(g/L)	0.296 (0.218-0.377)	0.296 (0.233-0.365)	0.816
Initial creatinin(µmol/L)	1.18 (0.863-2.40)	1.44 (0.760-2.98)	0.687
Initial GFR(mL/min per 1.73 m2)	71.9 (24.8-99.5)	50.4 (20.9-103)	0.866
CRP(mg/L)	16.2 (4.35-35)	22 (8-52.1)	0.390
ESR(mm/h)	29 (13.5-64.8)	30.5 (10.8-76.8)	1
RF (IU/ml)	14 (6.60-39)	10 (7.35-24.1)	0.553
Proteinuria	50%	52.9%	0.795
Hematuria	39.1%	35.3%	0.726
Comorbidities and other parameters			
Hypertension	52.2%	61.8%	0.393
Diabetes mellitus	21.7%	20.6%	0.901
Chronic kidney disease	32.6%	41.2%	0.431
Coronary artery disease	4.3%	17.6%	0.050
Asthma	6.5%	11.8%	0.412
Hemodialysis	17.4%	20.6%	0.717
Renal biopsy	47.8%	50%	0.848

Among ANA-positive patients, speckled (AC-4/5) patterns were observed in 12 patients, homogeneous (AC-1) patterns in 10 patients, nucleolar (AC-8/9/10) patterns in 5 patients, and mixed patterns in 7 patients. Among ANA-positive patients, 28 had a low-positive ANA titer and 6 had a higher ANA titer according to the manufacturer's grading system.

Table 2. Baseline clinical characteristics and laboratory data of ANCA-associated vasculitis patients according to ANCA types

	MPO (+)	PR3 (+)	P value
Demographic			
Female, n (%)	43.6%	56.4%	0.069
Age, years	59.7±16.1	49.8±14.8	0.008
Organ involvement			
Lungs	31.4%	68.6%	0.551
Kidney	35.5%	64.5%	0.794
Ear, nose, throat	28.6%	71.4%	0.388
Joints	22.5%	77.4%	0.093
Skin	37.5%	62.5%	0.723
Gastrointestinal	22.2%	77.8%	0.438
Ocular	7.1%	92.9%	0.020
Neurological	37.5%	62.5%	0.723
Cardiac	50%	50%	0.623
Laboratory data			
Ig M (g/L)	0.839 (0.642-0.1.35)	0.609 (0.340-0.760)	0.026
Ig G (g/L)	13.1 (11-17.2)	8.84 (6.2-11.4)	0.002
Ig A (g/L)	2.16±1.03	1.59±0.803	0.031
Platelets($\times10^9$/L)	282 (217-350)	283 (225-334)	0.811
Neutrophils($\times10^9$/L)	5.38 (3.56-7.02)	6.35 (3.49-8,78)	0.304
Lymphocytes($\times10^9$/L)	1.28 (0.880-1.58)	1.35 (0.990-1.77)	0.596
Hemoglobin(g/L)	10.6±2.43	10.9±2.34	0.588
C3(g/L)	1.31±0.328	1.24±0.350	0.404
C4(g/L)	0.304 (0.260-0.378)	0.279 (0.211-0.354)	0.184
Initial creatinin(µmol/L)	1.56 (0.833-3.05)	1.13(0.800-2.48)	0.602
Initial GFR(mL/min per 1.73 m2)	43.5 (17.4-87.4)	71.8 (27.4-104)	0.148
CRP(mg/L)	26.3 (8.3-52.9)	15 (5.2-47)	0.120
ESR(mm/h)	34.4 (15-79)	36.5 (10.5-68)	0.379
RF (IU/ml)	10 (6.7-15.9)	13.8 (7.5-39)	0.687
Proteinuria	66.7%	65.9%	0.939
Hematuria	68%	63.3%	0.669
Comorbidities and other parameters			
Hypertension	71.4%	62.2%	0.388
Diabetes mellitus	66.7%	64.7%	0.879
Chronic kidney disease	66.7%	65.5%	0.917
Coronary artery disease	65.3%	75%	0.581
Asthma	71.2%	14.3%	0.002
Hemodialysis	70.8%	46.7%	0.075
Renal biopsy	68.3%	64.1%	0.692

Immunoblot analysis was performed in 32 ANA-positive patients, yielding positive results in 22 patients and negative results in 10 patients. Among the 22 patients with positive immunoblot results, the following autoantibodies were detected: anti-dsDNA (n=3), anti-Ro-52 (n=3), anti-PM-Scl (n=2), anti-SS-B (n=2), anti-Scl-70 (n=2), anti-Mi-2 (n=2), anti-SS-A (n=1), anti-DFS70 (n=1), anti-Jo-1 (n=1), anti-Ku (n=1), anti-CENP-B (n=1), anti-Sm/RNP (n=1), anti-histone+nucleosome (n=1), and combined anti-SS-A+anti-PM-Scl positivity (n=1).

Of the 80 patients, 33.75% were MPO-ANCA positive and 66.25% were PR3-ANCA positive. MPO-ANCA-positive patients were diagnosed at a significantly older age compared with PR3-ANCA-positive patients (59.7±16.1 vs. 49.8±14.8 years, p=0.008). Ocular involvement was significantly more frequent in PR3-ANCA–positive patients (p=0.020), whereas asthma was more prevalent in the MPO-ANCA-positive group (p=0.002).

No significant differences were observed between ANCA subgroups regarding platelet count, neutrophil count, lymphocyte count, hemoglobin, C3, C4, baseline creatinine, baseline GFR, CRP, ESR, or RF levels. However, IgM, IgG, and IgA levels were significantly higher in the MPO-ANCA-positive group (Table 2).

DISCUSSION

This study evaluated the clinical and laboratory characteristics of ANCA serotypes and ANA status in Turkish patients with AAV during extended follow-up. PR3-ANCA was the predominant serotype, accounting for 66.3% of cases. Several clinical differences were observed between patients with PR3-ANCA and MPO-ANCA. Data from the POLVAS registry similarly reported that MPO-ANCA–positive patients were generally older than PR3-ANCA-positive patients (11), which is consistent with our findings.

A notable finding of our study was the higher frequency of ocular involvement in PR3-ANCA-positive patients. Previous studies have shown that ear, nose, throat, and ocular involvement is less common in MPO-ANCA-positive patients and more prevalent in those with PR3-ANCA (12,13). In contrast, no significant differences were observed between the two serotypes regarding nervous system, skin, pulmonary, or renal involvement. Although MPO-ANCA is often associated with more severe renal pathology due to extensive glomerular damage (14), ANCA specificity alone may not reliably predict renal outcomes (15).

AAV is characterized by multisystem involvement and heterogeneous clinical manifestations. Non-specific symptoms such as fever, fatigue, weight loss, and serositis are frequently observed. In our cohort, renal involvement was present in 38.8% of patients. Although renal involvement has been reported in up to 83.5% of AAV patients in previous studies (16), it was less frequent in our population, possibly reflecting differences in disease presentation, referral patterns, or diagnostic practices.

A single-center retrospective study from Spain reported ear–nose–throat involvement in 31.4% and ocular involvement in 19.5% of patients (17,18). In comparison, ear–nose–throat involvement was more frequent and ocular involvement less

common in our cohort. These discrepancies may be related to geographic, environmental, or genetic factors and warrant further investigation.

Pulmonary involvement has been identified as a risk factor for disease relapse in the Glomerular Disease Collaborative Network cohort (19). However, lung involvement was not associated with relapse in our study, consistent with findings from the French Vasculitis Study Group (20).

AAV is a rare autoimmune disease associated with substantial morbidity and mortality if diagnosis is delayed. Its prevalence is estimated at 13–30 cases per million individuals in North America and Europe (21,22). Genetic and clinical studies suggest that differences in outcomes between PR3-AAV and MPO-AAV are more pronounced than those between GPA and MPA, emphasizing the pathogenetic importance of antigen specificity (23,24). The distribution of ANCA serotypes varies geographically, with MPO-ANCA predominating in East Asian populations and PR3-ANCA being more common in Europe and the United States (25). Consistent with the literature, PR3-ANCA was the dominant serotype in our cohort.

A previous study reported no difference in baseline renal function between PR3-ANCA–positive and MPO-ANCA–positive patients, supporting our findings (26). Previous investigations have demonstrated ANA positivity in 18–66% of patients with AAV, although its clinical significance remains unclear (27). In our study, 42.5% of patients were ANA positive. Among patients with a homogeneous ANA pattern, 31% exhibited MPO positivity in a prior study (28), which is consistent with our findings, where 4 of 10 patients with a homogeneous pattern were MPO positive.

It has been suggested that ANA may contribute to renal injury through multiple mechanisms (29). However, the clinical impact of ANA positivity in AAV remains controversial. In our study, ANA-positive AAV patients exhibited a higher prevalence of renal involvement. Previous studies have also reported that ANA-positive AAV patients have more severe renal disease, including higher rates of ESRD, proteinuria, hypertension, reduced eGFR, elevated serum creatinine, and increased frequency of renal biopsy (16). These findings support the hypothesis that ANA may exacerbate renal dysfunction in AAV, although the underlying mechanisms require further clarification.

This study has several limitations. Its retrospective design and inclusion of patients based solely on ANCA positivity may have led to the exclusion of ANCA-negative AAV cases. Additionally, treatment-specific outcomes could not be evaluated. The absence of renal pathology in some patients also limited differentiation between ANA-positive AAV and lupus nephritis with ANCA positivity.

CONCLUSION

In conclusion, ANA-positive AAV patients exhibited more severe renal involvement compared with ANA-negative patients. Renal biopsy remains essential for distinguishing AAV from systemic lupus erythematosus in ANA-positive cases. Ocular involvement was more prominent in PR3-ANCA-positive patients, whereas asthma was more frequent among MPO-ANCA-positive patients. These findings highlight the clinical relevance of ANA status and ANCA serotypes in AAV and underscore the need for further studies to clarify their prognostic implications.

Declarations

Ethics Committee Approval: Ethics committee approval was obtained from the Human Research Ethics Committee of a hospital (Date: July 12, 2023, Decision No: E2-23-4480)). This study was conducted according to the principles of the Declaration of Helsinki.

Authors' Contributions

ES designed the study. Data collection was performed by ES. Statistical analysis was performed by ES. Manuscript was written by ES. All authors read and approved of the final manuscript.

Conflict of Interest: No conflict of interest was declared by the authors.

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