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Research Article

Clinical investigation of patients with positive antineutrophil cytoplasmic antibodies (ANCA) test for autoimmunity and vasculitis

ANCA testi pozitif olan hastaların otoimmünite ve vaskülit açısından klinik olarak incelenmesi

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Abstract

Aim: Antineutrophil cytoplasmic antibodies (ANCA) autoantibodies target neutrophil and monocyte lysosomal granules. The objective of the study is to investigate patients with a positive ANCA test for autoimmune disorders and vasculitis.

Material and Methods: This 4-year retrospective cohort research analyzed 9.480 Turkish tertiary hospital serum samples. The baseline evaluation included 218 eligible ANCA-positive patients with vasculitis (AAV) and other autoimmune and non-autoimmune diseases.

Results: The other two groups have less pulmonary, renal, ear nose and throat (ENT), ophthalmic, and neurologic involvement than AAV individuals. AAV and autoimmune patients have higher joint and skin involvement than non-autoimmune patients. Hypertension and chronic renal failure are more common in AAV and non-autoimmune individuals than autoimmune ones. Chronic obstructive pulmonary disease (COPD) and proteinuria are higher in AAV patients than autoimmune patients. AAV patients have more hematuria than the other two. For non-autoimmune disease, myeloperoxidase (MPO) titer and erythrocyte sedimentation rate (ESR) are higher than for autoimmune disease. High proteinase 3 (PR3) titers are associated with AAV and autoimmune disorders. AAV patients have higher CRP and complement 3(C3) than the other two groups. In AAV, C4 and rheumatoid factor (RF) are higher than in non-autoimmune disease. Patients with autoimmune disease had higher initial GFR than the other two groups.

Conclusion: We concluded that ANCA may be positive in numerous AAV-like diseases. A tertiary hospital found a strong correlation between ANCA titers, particularly PR3, and AAV clinical diagnosis. ANCA titre and organ system extent may be clinical AAV diagnostic indicators.

Keywords: antineutrophil cytoplasmic antibody, myelopreoxidase, proteinase 3, autoimmune, vasculitis

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Öz

Amaç: Antinötrofil sitoplazmik antikorlar (ANCA), nötrofil ve monositlerde görülen lizozomal granülleri spesifik olarak hedef alan otoantikorlardır. Çalışmanın amacı, ANCA testi pozitif olan hastaları otoimmün bozukluklar ve vaskülit açısından araştırmaktır.

Gereç ve Yöntemler: Bu 4 yıllık retrospektif kohort araştırmasında üçüncü basamak bir hastaneye gönderilen 9.480 serum örneği analiz edilmiştir. Başlangıç değerlendirmesi, vaskülit ve diğer otoimmün ve otoimmün olmayan hastalıkları olan 218 uygun ANCA-pozitif hastayı içeriyordu.

Bulgular: ANCA ilişkili vaskülit (AAV) bireylerine göre diğer iki grup daha az pulmoner, renal, kulak burun boğaz (KBB), oftalmik ve nörolojik tutuluma sahiptir. AAV ve otoimmün hastalığı olanlarda eklem ve cilt tutulumu otoimmün hastalığı olmayanlara göre daha yüksektir. Hipertansiyon ve kronik böbrek yetmezliği AAV ve otoimmün olmayan bireylerde otoimmün olanlara göre daha yaygındır. Kronik obstrüktif akciğer hastalığı (KOAH) ve proteinüri AAV hastalarında otoimmün hastalığı olanlara göre daha yüksektir. AAV hastalarında diğer ikisine göre daha fazla hematüri vardır. Otoimmün olmayan hastalıkta miyelopereoksidaz (MPO) titresi ve eritrosit sedimentasyon hızı (ESR) otoimmün hastalığa göre daha yüksektir. Yüksek proteinaz 3 (PR3) titreleri AAV ve otoimmün hastalıklarla ilişkilidir. AAV hastalarında C reaktif protein (CRP) ve kompleman 3 (C3) diğer iki gruba göre daha yüksektir. AAV'de C4 ve romatoid faktör (RF) otoimmün olmayan hastalığa göre daha yüksektir. Otoimmün hastalığı olanların başlangıç glomerüler filtrasyon hızı (GFR) diğer iki gruptan daha yüksektir.

Sonuç: ANCA'nın birçok AAV benzeri hastalıkta pozitif olabileceği sonucuna vardık. Üçüncü basamak bir hastanede ANCA titreleri, özellikle PR3 ile AAV klinik tanısı arasında güçlü bir korelasyon bulunmuştur. ANCA titresi ve organ sistemi kapsamı klinik AAV tanı göstergeleri olabilir.

Anahtar Kelimeler: antinötrofil sitoplazmik antikor, miyelopreoksidaz, proteinaz 3, otoimmün, vaskülit

Introduction

Antineutrophil cytoplasmic antibodies (ANCA) are autoantibodies that target neutrophil and monocyte lysosomal granules [1]. Since its discovery in 1982 in necrotizing glomerulonephritis patients, ANCA testing has been used to diagnose, exclude, or monitor small vessel vasculitides, specifically ANCA-associated vasculitides (AAV) [2]. ANCA testing is also requested for non-AAV [3]. Indirect immunofluorescence (IIF) and enzyme-linked immunosorbent assay (ELISA) test ANCA. ANCA binding to peripheral blood granulocytes can be assessed using the IIF method. Ethanol and formalin-fixed granulocytes are ANCA substrates [4].

c-ANCA targeting PR3 causes increased ear, nose, eye, and lung symptoms, but renal involvement is rare. They have a higher risk of relapse, poor prognosis, and mortality [5]. Patients with p-ANCA against MPO commonly develop pauci-immune segmental necrotizing glomerulonephritis. This condition may occur alone or with systemic symptoms. Relapses and ear, nose, eye, and lung difficulties are less common in these patients [6].

Non-vasculitic diseases can have ANCA. ANCA positivity has also been reported in primary sclerosing cholangitis, drug-induced vasculitis, systemic lupus erythematosus (SLE), autoimmune hepatitis, interstitial lung disease, cystic fibrosis, ulcerative colitis, rheumatoid arthritis, and particular infections including subacute bacterial endocarditis [7]. Clinicians must use ANCA status to rule out vasculitis mimickers in the absence of histology AAV. Clinical findings strongly suggest ANCA pathogenicity, while in vitro studies have identified neutrophil and monocyte activation, complement-mediated inflammation, and neutrophil extracellular trap release that damages endothelial cells [8,9]. The study examines ANCA-positive patients with autoimmune diseases and vasculitis.

Material and Methods

This study reviewed 9.480 ANCA-tested blood samples from February 2019 to December 2022 at the Ankara Bilkent City Hospital, Medical Microbiology Laboratory. ANCA-positive patients with vasculitis and other autoimmune and non-autoimmune diseases at Ankara Bilkent City Hospital from February 2019 to December 2022 were the patient population and baseline evaluation. All vasculitis patients met these criteria: (1) Adhere to AAV classification criteria from the American College of Rheumatology (1990) or International Chapel Hill Consensus Conference (2012); (2) Age ≥18 years.

If a patient had numerous serum samples, only the first was included in the study. Only 218 of 9.480 patients had positive ANCA IIF/ELISA testing. Out of 218 patients, 80 had AAV, 65 had autoimmune disorders, and 73 had non-autoimmune diseases. Most of the 218 ANCA-positive serum samples came from rheumatology, nephrology, neurology, gastrointestinal, pulmonology, internal medicine, and others. All patients demanded ANCA IIF and ELISA. The Non-Interventional Clinical Research Ethics Committee of Ankara Bilkent City Hospital approved this study on July 12, 2023, under Decision Number. E2-23-4480. The research followed the Helsinki Declaration and publishing ethics.

The hospital records were utilized to acquire data on complement 3 (C3), C4, initial glomerular filtration rate (GFR), erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and rheumatoid factor (RF). These data were recorded concurrently with the IIF assessment or within one week thereafter. Clinical information including age, gender, and diagnosis was also recorded. Sera were tested for ANCA presence using IIF-based ANCA testing (Euroimmun, Lubeck, Germany) and anti-PR3 and anti-MPO using ELISAs. Hep-2-neutrophil substrate, formalin-fixed neutrophils, and ethanol-fixed neutrophils are in the IIF-based ANCA diagnostic kit. Before incubation, patient sera were diluted to one-tenth and put to substrate wells. These antibodies bind to immobilized neutrophil antigens in ANCA-containing serum. FITC-labeled anti-human antibody reaction is examined using fluorescence microscopy. ELISA kits cover reagent wells with pure recombinant PR3, natural PR3, and MPO antigens. PR3 and MPO ELISA have 20 relative units (RU)/mL cutoffs.

Statistical Analysis

The Statistical Package for Social Sciences (SPSS) version 22.0 (IBM SPSS Inc., Chicago, IL) was employed for statistical analysis. Data normality was determined via the Kolmogorov-Smirnov test. Median was used for non-normally distributed values and mean and standard deviation for regularly distributed ones. Two-group comparisons used the Mann-Whitney U test for numerical variables without a normal distribution and the t-test for independent samples for numerical variables with a normal distribution. Based on appropriateness, the Chi-square or Fisher's exact test compared categorical data. Value of $p < 0.05$ were considered statistically significant.

Results

Of 218 randomly selected ANCA-positive patients, 124 (56.8%) were female and 94 (43.1%) were male. Women aged 54.9 ± 16.4 years, whereas men lived 53.5 ± 17.3 years. In these patients, 139 (63.8%) were c-ANCA positive, 70 (32.1%) were p-ANCA positive, and 9 (4.1%) were both. Six (27.8%) were MPO and PR3 positive, 101 (46.3%) were MPO positive, and 111 (50.9%) were PR3 positive.

Of the AAV patients, 60 had granulomatosis with polyangitis (GPA), 12 had microscopic polyangiitis (MPA) and 8 had eosinophilic granulomatosis with polyangiitiis (EGPA).

The range of diseases associated with autoimmune diseases includes rheumatoid arthritis (12/65), ulcerative colitis (11/65), primary sclerosing cholangitis (5/65) and systemic lupus erythematosus (5/65), Crohn's disease (4/65), Sjogren's syndrome (3/65), ankylosing spondylitis (3/65), and familial Mediterranean fever (3/65), leukocytoclastic vasculitis (3/65), and intestinal lung diseases (3/65), scleroderma (2/65), uveitis (2/65), autoimmune hepatitis (2/65), and gout (2/65), Behçet's disease (1/65), primary biliary cirrhosis (1/65), polyneuropathy (1/65), polymyositis (1/65), and systemic connective tissue disease (1/65).

The range of diseases associated with non-autoimmune diseases includes chronic kidney disease (33/73), cerebrovascular events (10/73), pneumonia (5/73), hypothyroidism (3/73), pulmonary hypertension (2/73), gallstones (2/73), nephrotic syndrome (2/73), chronic obstructive pulmonary disease (COPD) (2/73), liver cirrhosis (2/73), and bronchiectasis (2/73), lung abscess (1/73), bronchitis (1/73), benign prostatic hypertrophy (1/73), gastritis (1/73), lichen planus (1/73), breast cancer (1/73), membranous glomerulonephritis (1/73), prostate cancer (1/73), tubulopathy (1/73), and sepsis (1/73).

Discussion

The other two groups have less pulmonary, renal, ENT, ophthalmic, and neurologic involvement than AAV individuals. AAV and autoimmune patients have higher joint and skin involvement than non-autoimmune patients. Hypertension and chronic renal failure are more common in AAV and non-autoimmune individuals than autoimmune ones. COPD and proteinuria are higher in AAV patients than autoimmune patients. AAV patients have more hematuria than the other two. For non-autoimmune disease, MPO titer and ESR are higher than for autoimmune disease. High PR3 titers are associated with AAV and autoimmune disorders. AAV patients have higher CRP and C3 than the other two groups. In AAV, C4 and RF are higher than in non-autoimmune disease. Patients with autoimmune disease had higher initial GFR than the other two groups (Table 1).

Table 1. Baseline clinical characteristics and laboratory data of ANCA positive vasculitis, autoimmune and non-autoimmune patients according to ANCA type.

	AAV n=80	ANCA(+) Autoimmune disease(AI) n=65	ANCA(+)Non-autoimmune disease(NAI) n=73	AAV/AI (p)	AAV/NAI (p)	AI/NAI (p)
Demographic						
Female, n (%)	48.8%	53.8%	68.5%	0.542	0.013	0.077
Age, years	52.8±16.2	49±18.1	60±14.8	0.188	0.005	<0.001
Organ involvement at ANCA determination						
Lungs, n (%)	51 (63.7%)	14 (21.5%)	12 (16.4%)	<0.001	<0.001	0.444
Kidney, n (%)	31 (38.8%)	6 (9.2%)	12 (16.4%)	<0.001	0.002	0.210
Ear, nose, throat, n (%)	35 (43.8%)	3 (4.6%)	2 (2.7%)	<0.001	<0.001	0.556
Joints, n (%)	49 (61.3%)	32 (49.2%)	21 (28.8%)	0.147	<0.001	0.014
Skin, n (%)	16 (20%)	6 (9.2%)	1 (1.4%)	0.072	<0.001	0.036
Gastrointestinal, n (%)	9 (11.3%)	1 (1.5%)	3 (4.1%)	0.022	0.101	0.369
Eye, n (%)	14 (17.5%)	3 (4.6%)	0 (0%)	0.016	<0.001	0.063
Neurological, n (%)	16 (20%)	2 (3.1%)	5 (6.8%)	0.002	0.018	0.313
Comorbidities						
Hypertension, n (%)	45 (56.3%)	22 (33.8%)	44 (60.3%)	0.007	0.614	0.002
Diabetes, n (%)	17 (21.3%)	18 (27.7%)	25 (34.2%)	0.367	0.072	0.407
Asthma, n (%)	7 (8.8%)	4 (6.2%)	5 (6.8%)	0.557	0.662	0.869
Chronic renal failure, n (%)	29 (36.3%)	9 (13.8%)	27 (37%)	0.002	0.925	0.002
Chronic obstructive lung disease, n (%)	8 (10%)	1 (1.5%)	5 (6.9%)	0.036	0.501	0.123
Laboratory data						
Hematuria, n (%)	30 (37.5%)	10 (15.4%)	13 (17.8%)	0.003	0.007	0.703
Proteinuria, n (%)	41 (51.2%)	16 (24.6%)	28 (38.4%)	0.001	0.109	0.084
MPO ANCA titer, median (min-max)	2.0 (2-200)	15.6 (2-200)	44.9 (2-200)	0.136	<0.001	0.003
PR3 ANCA titer, median (min-max)	87.3 (2-200)	25.3 (2-200)	2 (2-200)	<0.001	<0.001	0.143
MPO, n (%)	27 (33.8%)	31 (47.7%)	47 (64.4%)	0.088	<0.001	0.048
PR3, n (%)	53 (66.3%)	37 (56.9%)	28 (38.4%)	0.250	<0.001	0.029
Initial GFR (mL/min per 1.73 m2) median (min-max)	68.5 (4.90-154)	94.5 (6.57-186)	47.2 (3.27-126)	0.002	0.193	<0.001
CRP (mg/L) median (min-max)	17.6 (0.800-327)	6.10 (0.400-143)	7.30 (0.400-192)	<0.001	0.027	0.168
C3 (g/L) median (min-max)	1.25 (0.293-2.27)	1.03 (0.377-1.56)	1.16 (0.388-5.60)	<0.001	0.045	0.008
C4 (g/L) median (min-max)	0.296 (0.0519-1.24)	0.214 (0.0550-0.434)	0.279 (0.117-0.530)	<0.001	0.577	0.001
ESR (mm/h) median (min-max)	29 (0.253-127)	24.5 (4.10-123)	44.5 (3-139)	0.539	0.114	0.030
RF (IU/ml) median (min-max)	11.5 (3.2-474)	9.8 (3.0-91)	6.9 (2.20-107)	0.061	0.023	0.486

ANCA is linked to various diseases other than AAV, as shown by others and our data. Chronic inflammatory diseases, malignancies, infections, and propylthiouracil usage have been described [10,11]. In AAV-deficient patients, ANCA positivity is unclear. Animal studies show MPO and PR3 ANCA are pathogenic. Infused mice with ANCA auto-antibodies had glomerulonephritis symptoms and histology [12,13]. There is little clinical evidence that ANCA is pathogenic in people. Transplacental transmission of maternal MPO antibodies causes MPA in newborns, according to a common case report. Plasma exchange in severe AAV proves pathogenicity indirectly [14,15]. Immunoglobulin A (IgA) nephropathy patients with incidental positive ANCA serology had worse clinical and histological features [16]. This indicates ANCA pathogenicity, even in individuals without AAV.

During sustained observation, Turkish patients with ANCA-positive AAV and autoimmune and non-autoimmune disorders were examined clinically and biochemically. In AAV, multisystem dysfunction and different clinical symptoms occur. Common symptoms include fever, tiredness, weight loss, and cavity effusion. The organs most frequently affected by AAV are the kidneys, lungs, joints, and ENT, which will be discussed individually.

Studies show 40%–76% kidney involvement in AAV [17-20]. One study reported 60% renal involvement in AAV, 10% in the autoimmune group, and 13% in the non-autoimmune group. It was much higher in AAV than the other two groups [21]. We found 38% renal involvement in AAV, much greater than the other two groups.

Various studies [17–20] show 55%–84% pulmonary involvement in AAV. AAV was substantially more common than the other two categories, with pulmonary involvement in 63% of AAV patients, 21% in the autoimmune group, and 21% in the non-autoimmune group. In our analysis, AAV had a much greater prevalence of lung involvement—63.7% compared to 21.5% in the autoimmune group and 16.4% in the non-autoimmune group.

Studies show 27%–73% joint AAV involvement [17-20]. A study found joint involvement in 25% of AAV cases, 52% in the autoimmune cohort, and 11% in the non-autoimmune cohort, with a statistically significant increase in the autoimmune group [21]. Joint involvement was 61.3% in AAV, 49.2% in the autoimmune group, and 28.8% in the non-autoimmune group in our study. AAV had significantly higher rates.

ENT involvement was seen in 31% of AAV cases, 3% of the autoimmune group, and 11% of the non-autoimmune group, with AAV having a statistically significant greater prevalence [21]. AAV had a much greater prevalence of ENT involvement—43.8% compared to 4.6% in the autoimmune group and 2.7% in the non-autoimmune group.

In a previous study conducted in the Netherlands, among 118 patients without a diagnosis of AAV, 87 were found to have alternative diagnoses, which included: inflammatory bowel disease (n = 24), renal insufficiency from other etiologies (n = 17), rheumatoid arthritis (n = 11), infections (n = 11), other forms of vasculitis (n = 8), malignancy (n = 4), and miscellaneous conditions (n = 12). In our investigation, inflammatory bowel disease, renal failure, rheumatoid arthritis, and other conditions emerged as non-AAV diseases [22].

MPO titers were higher in non-autoimmune diseases than the other two groups. In the non-autoimmune group, chronic renal failure, inflammation and infection are possible causes. One study found that AAV patients had higher MPO titers than the other two [21]. As with a previous study, PR3 titer was higher in AAV than the other two groups [21]. Our findings show that MPO and PR3 ANCA can be positive in many clinical settings. Our patients with AAV had higher ANCA levels, notably PR3, and more damaged organ systems.

The significance of ANCA positivity in patients without AAV is unclear. Evidence from animal studies, which is supported by a case report of transplacental transfer of MPO-ANCA resulting in neonatal pulmono-renal syndrome, suggest that MPO-ANCA are pathogenic [23]. It has also been shown that naturally occurring anti-MPO autoantibodies can exist in healthy individuals [24]. However, these anti-MPO autoantibodies have epitope specificities that are different from those present in ANCA disease implicating immunodominant epitopes in the pathology of AAV [25]. It is therefore possible that anti-MPO positivity in patients without AAV, like healthy controls, are directed against non-vasculitic disease-causing epitopes, but specifically testing for these remains outside standard clinical care. In a retrospective review of 236 ANCA-positive patients, patients with an alternative diagnosis to AAV were more often anti-MPO positive than anti-PR3 positive (58% vs. 42%) [22].

Clinical features differ between PR3-AAV and MPO-AAV. The combination of upper and/or lower respiratory tract involvement with renal involvement is frequently seen in PR3-AAV as opposed to vasculitis limited to the kidney in MPO-AAV. The frequency of pulmonary involvement is similar in both serotypes, but differs in nature. While fibrosing lesions are more frequent in MPO-AAV, cavitating lesions are more often found in PR3-AAV. Nodules and masses on chest radiography are more often observed in PR3-AAV and patchy infiltrates more often in MPO-AAV. Initially, alveolar lung hemorrhage was reported more frequently in MPO-AAV but more recent studies have reported it to be more frequent in PR3-AAV [26].

Our study has limitations, the most significant of which being its retrospective design and the identification of patients only based on their ANCA status. This indicates that 'ANCA-negative' AAV patients were disregarded, despite these variants being predominantly renal, infrequently systemic, and remaining unusual [27]. Consequently, we assert that this limitation does not substantially affect our message. Notably, other small-vessel vasculitides necessitated either the presence of a particular marker (i.e., cryoglobulinemia) and/or histological confirmation of the condition. Moreover, not all diagnoses were confirmed by histology evidence. A key aspect of our work is its pragmatic and clinical methodology in addressing diagnostic difficulties encountered in daily practice.

In conclusion, we determined that ANCA may be positive in several conditions that mimic AAV. A substantial association was detected between ANCA titers, particularly PR3 titers, and a clinical diagnosis of AAV in ANCA-positive patients at a tertiary

hospital in Turkey. The ANCA titre and the extent of affected organ systems may function as diagnostic markers in AAV within clinical practice. Future studies may aid clinicians in more accurately differentiating between AAV and other diseases.

Ethics Committee Approval

The Non-Interventional Clinical Research Ethics Committee of Ankara Bilkent City Hospital approved this study on July 12, 2023, under Decision Number. E2-23-4480.

Conflict of Interest

None declared by the authors.

Financial Disclosure

None declared by the authors.

Authors Contribution

ES: Conceptualization, Data Collection, Methodology, Writing, Formal Analysis, Original Draft. AT: Supervision, Writing, Review & Editing. Both authors have read and approved the final version of the manuscript.

References

1. Tsukui D, Kimura Y, Kono H. Pathogenesis and pathology of anti-neutrophil cytoplasmic antibody ANCA - associated vasculitis. J Transl Autoimmun 2021; 4:1 00094.
2. de Groot K, Csernok E, van der Woude D. History of antineutrophil cytoplasmic autoantibodies: Milestones in rheumatology. Z Rheumatol 2025; 84: 219-224.
3. Savige J, Trevisin M, Pollock W. Testing and reporting antineutrophil cytoplasmic antibodies (ANCA) in treated vasculitis and non-vasculitic disease. J Immunol Methods. 2018; 458: 1-7.
4. Folci M, Ramponi G, Solitano V, Brunetta E. Serum ANCA as Disease Biomarkers: Clinical Implications Beyond Vasculitis. Clin Rev Allergy Immunol 2022; 63: 107-123.
5. Qi F, Hao J, Wei W. Impact of different ANCA serotypes on the long-term outcome of ANCA-associated vasculitis patients. Ann Med 2023; 55: 2289614.
6. Jiang C, Liu J, Wang H, Liu H, Fu Z, Li M et al. Anti-neutrophil cytoplasmic antibody patterns can predict clinical relapse in ANCA-associated vasculitis: overall population and subgroups. Clin Exp Rheumatol 2023; 41: 848-55.
7. Walulik A, Łysak K, Błaszkiewicz M, Górecki I, Gomułka K. The Role of Neutrophils in ANCA-Associated Vasculitis: The Pathogenic Role and Diagnostic Utility of Autoantibodies. Int J Mol Sci 2023 7; 24: 17217.
8. Sun XJ, Li ZY, Chen M. Pathogenesis of anti-neutrophil cytoplasmic antibody-associated vasculitis. Rheumatol Immunol Res 2023; 4: 11-21.
9. Kitching AR, Anders HJ, Basu N, Brouwer E, Gordon J, Jayne DR et al. ANCA-associated vasculitis. Nat Rev Dis Primers 2020; 6: 71.
10. Morita TCAB, Trés GFS, Criado RFJ, Sotto MN, Criado PR. Update on vasculitis: an overview and dermatological clues for clinical and histopathological diagnosis - part I. Ann Bras Dermatol 2020; 95: 355-371.
11. Sayınalp B, Bilgin E, Güler AC, Kalyoncu U. c-ANCA positivity in a patient with relapsing polychondritis: Co-incidence or not? A case and review of the literature. J Turk Soc Rheumatol 2021; 13: 88-90.
12. Huang F, Lv Y, Liu S, Wu H, Liu Q. Animal models for anti-neutrophil cytoplasmic antibody-associated vasculitis: Are current models good enough? Animal Model Exp Med 2023; 6: 452-63.
13. Shochet L, Holdsworth S, Kitching AR. Animal Models of ANCA Associated Vasculitis. Front Immunol 2020; 11: 525.
14. Ashok A, Russell L, Dey M, Kouranloo K. Incidence, clinical features, management and outcomes of ANCA-associated vasculitis in pregnancy- a systematic literature review. Sarcoidosis Vasc Diffuse Lung Dis 2023; 40: e2023040.
15. Paroli M, Gioia C, Accapezzato D. New Insights into Pathogenesis and Treatment of ANCA-Associated Vasculitis: Autoantibodies and Beyond. Antibodies (Basel) 2023; 12: 25.
16. Tota M, Donizy P, Byrska M, Krajewska M, Kusztal M. An unsuspected histopathological finding -concomitant IgA nephropathy in a patient with ANCA-associated vasculitis: a case report and literature review. Front Immunol 2023; 14: 1227878.
17. Shobha V, Fathima S, Prakash R. Granulomatosis with polyangiitis: clinical course and outcome of 60 patients from a single center in South India. Clin Exp Med 2018; 18: 347-53.
18. Kim HW, Song YW. ANCA-associated vasculitis: report from Korea. Clin Exp Nephrol 2013; 17: 708-11.
19. Holle JU, Gross WL, Latza U, Nölle B, Ambrosch P, Heller M, Fertmann R, Reinhold-Keller E. Improved outcome in 445 patients with Wegener's granulomatosis in a German vasculitis center over four decades. Arthritis Rheum 2011; 63: 257-66.
20. Koç E, Aşık MA, Erdoğan EK, Arslan D, Özbek S, Özer HTE. ANCA associated vasculitis: clinical course and outcome of 44 patients from a single center in Turkey. Rheumatol Q 2023; 1: 6-13.
21. Merindol J, Levraut M, Seitz-Polski B, Morand L, Martis N. Diagnostic significance of antineutrophil cytoplasmic antibody (ANCA) titres: a retrospective case-control study. RMD Open 2023; 9: e003113.

22. Houben E, Bax WA, van Dam B, Slieker WAT, Verhave G, Frerichs FCP, van Eijk IC, Boersma WG, de Kuyper GTM, Penne EL. Diagnosing ANCA-associated vasculitis in ANCA positive patients: A retrospective analysis on the role of clinical symptoms and the ANCA titre. Medicine (Baltimore) 2016; 95: e5096.
23. Bansal PJ, Tobin MC. Neonatal microscopic polyangiitis secondary to transfer of maternal myeloperoxidase-antineutrophil cytoplasmic antibody resulting in neonatal pulmonary hemorrhage and renal involvement. Ann Allergy Asthma Immunol 2004; 93: 398-401.
24. Cui Z, Zhao MH, Segelmark M, Hellmark T. Natural autoantibodies to myeloperoxidase, proteinase 3, and the glomerular basement membrane are present in normal individuals. Kidney Int 2010; 78: 590-7.
25. Roth AJ, Ooi JD, Hess JJ, van Timmeren MM, Berg EA, Poulton CE et al. Epitope specificity determines pathogenicity and detectability in ANCA-associated vasculitis. J Clin Invest 2013; 123: 1773-83.
26. Hilhorst M, van Paassen P, Tervaert JW; Limburg Renal Registry. Proteinase 3-ANCA Vasculitis versus Myeloperoxidase-ANCA Vasculitis. J Am Soc Nephrol 2015; 26: 2314-27.
27. Fijołek J, Wiatr E. Antineutrophil cytoplasmic antibodies (ANCA) - their role in pathogenesis, diagnosis, and treatment monitoring of ANCA-associated vasculitis. Cent Eur J Immunol 2020; 45: 218-27.