Neurologijos seminarai 2023; 27(95): 53–59
DOI: 10.15388/NS.2023.27.7

ANCA-Associated Vasculitic Neuropathy: A Case Report

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Summary. Anti-neutrophil cytoplasmic antibodies (ANCA)-associated vasculitides (AAVs) are a group of autoimmune diseases that can affect many vital organs and tissues, as well as the nervous system. When peripheral nerves are affected, one of the clinical features of AAVs can be vasculitic neuropathies (VNs). This usually causes asymmetric weakness and numbness in the distal parts of extremities, frequently followed by pain. Nerve conduction study (NCS) reveals axonal loss in multiple individual nerves, whereas a length-dependent process may also be observed. As AAV is a rare diagnostic entity and can manifest with a vast variety of symptoms, it is seldom involved in the initial diagnostic work-up of polyneuropathy. Moreover, the only definitive diagnostic method for VN is peripheral nerve biopsy. Therefore, AAV represents one of the greatest difficulties in the differential diagnosis of neuropathies. In this article, we present a clinical case of a 73-year-old patient admitted to the hospital due to blurry vision, headache, and limb weakness. The findings of the neurological examination were consistent with polyneuropathy. A thorough examination was performed to determine the exact diagnosis. However, due to the lack of typical symptoms, VN was diagnosed only after acute renal failure evolved. This case demonstrates the many symptoms that AAV can present with and the importance of a thorough differential diagnosis for multiple mononeuropathies.

Keywords: ANCA-associated vasculitis, microscopic polyangiitis, multiple mononeuropathies, vasculitic neuropathies.

INTRODUCTION

Anti-neutrophil cytoplasmic antibodies (ANCA)-associated vasculitides (AAVs) are a group of autoimmune diseases that predominantly affect small vessels. Clinical presentations of AAV, in addition to non-specific clinical features of systemic inflammation, such as fever, fatigue, and weight loss, include dysfunction of the heart, kidneys, lungs, gastrointestinal tract, and many other internal organs and tissues, as well as peripheral nerves [1]. AAVs are categorized into three subgroups, namely microscopic polyangiitis (MPA), eosinophilic granulomatosis with polyangiitis (Churg-Strauss syndrome), and granulomatosis with polyangiitis (Wegener's granulomatosis), which are defined based on clinical features and results of immunological tests.

AAV is a rare disease, and according to recent epidemiological studies its prevalence reaches approximately 300-421 cases per million people in the world [1]. Despite its rarity, neurologists should be aware of this diagnostic entity because it can cause serious lesions of the nervous system, especially peripheral nerves, which progress rapidly, and may mimic other diseases such as Guillain-Barre syndrome (GBS). The incidence of AAV-induced vasculitic neuropathies (VNs) varies due to the lack of research, yet recent studies show that VNs may be present in 65% of cases of Churg-Strauss syndrome, 23% of microscopic polyangiitis, and 19% of Wegener's granulomatosis [2].

Since AAV is an autoimmune condition, the pathogenesis of this disease is rather complex and not fully understood. The main cause of VN is ischemic vasculitic nerve damage resulting from cell-mediated immunity and immune complex deposition [3]. Accordingly, Peripheral

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Table 1. **Main laboratory test results during treatment**

Variable	1st admission (June 23, 2021)	2nd admission (July 05, 2021)	ARF onset (July 09, 2021)	Discharge (August 13, 2021)	Reference range
RBC ($\times 10^{12}$/L)	3.97	4.22	3.42	3.30	3.9-5.03
NE ($\times 10^{9}$/L)	14.1	16.4	13.8	3.6	1.9-8.2
PLT ($\times 10^{9}$/L)	474	507	479	79	179-408
ALT (IU/L)	–	–	27	–	0-35
AST (IU/L)	–	–	23	–	0-35
HGB (g/L)	112	115	93	94	120-155
WBC ($\times 10^{9}$/L)	16.2	18.4	19.0	4.3	3.8-11.8
CRP (mg/L)	228.4	240.5	200.1	19.9	0-5
Procalcitonin (g/L)	0.91	1.43	15.38	–	0-0.1
Urea (mmol/L)	5.2	13.0	19.6	12.3	2.8-7.2
Creatinine (mol/L)	43.0	143.0	407.0	389.0	45-84
GFR (mL/min/1.73m^2)	97	31	9	9	60
CK (IU/L)	77	–	–	–	0-145
Cerebrospinal fluid					
Red-cell count ($\times 10^{9}$/L)	0	25	–	–	0
Total nucleated cells ($\times 10^{6}$/L)	0	29	–	–	0-5
PMN ($\times 10^{6}$/L)	0	18.4	–	–	
MMN ($\times 10^{6}$/L)	0	36.4	–	–	
Total protein (g/L)	0.34	0.47	–	–	0.15-0.45
Glucose (mmol/L)	3.15	3.59	–	–	2.2-3.9

Abbreviations: RBC – red blood cells; NE – neutrophil count; PLT – platelet count; ALT – alanine transaminase; AST – aspartate transaminase; HGB – hemoglobin; WBC – white blood cells; CRP – C reactive protein; GFR – glomerular filtration rate; CK – creatine kinase; PMN – polymorphonuclear leukocytes; MMN – monomorphonuclear leukocytes; ARF – acute renal failure.

Nerve Society Guidelines instruct that the only diagnostic method for pathologically definite vasculitic neuropathy is a peripheral nerve biopsy which meets the histopathological view [4]. Although it is the only definite answer, studies show that nerve biopsy is performed only in 12% of all AAV patients, and clinicians tend to make the diagnosis exclusively by clinical symptoms [2].

Clinical features of VNs are usually asymmetric or multifocal sensory or sensorimotor deficits that predominantly affect the lower extremities, primarily their distal ends. Rarely, symmetric sensorimotor polyneuropathy or plexopathy may also occur. VNs are also frequently followed by pain in the affected limbs [5]. Inevitably, other clinical symptoms vary depending on the ANCA-targeted organs. Evidence illustrates that VNs are frequently accompanied by musculoskeletal, dermatological, and cardiovascular symptoms. In contrast, clinical features including renal, eye, and gastrointestinal involvement are less common in patients with VN [2]. VNs usually progress over several weeks, although acute attacks are also common [6]. Nerve conduction study (NCS) usually shows axonal damage involving multiple individual sensory and motor nerves in an asymmetric fashion [4].

We present the case of a 73-year-old woman who was diagnosed with VN caused by ANCA-associated vasculitis. This case illustrates the vast variety of symptoms that AAV can present with and therefore the importance of a thorough differential diagnosis.

CASE PRESENTATION

A 73-year-old female was admitted to the Department of Neurology of the Hospital of Lithuanian University of Health Sciences Kauno Klinikos due to polyneuropathy of unknown origin.

Two weeks prior to the admission, the patient arrived at the Emergency Department (ED) of the Hospital of Lithuanian University of Health Sciences Kauno Klinikos complaining of blurry vision, headache, and limb weakness. However, on neurological examination, the patient showed no cranial nerve pathology, muscle weakness, or sensory disturbances. Complete blood count and biochemical blood tests showed extensive inflammation (Table 1). However, urinalysis, chest X-ray, and abdominal ultrasound did not reveal any source of inflammation. A head CT and CT venogram were performed and showed no abnormalities. A lumbar puncture was performed to rule out possible neuroinfection, but cerebrospinal fluid (CSF) analysis was normal. Due to an infection of unknown origin, the patient was hospitalized in the Department of Pulmonology, and empiric antibiotic therapy with cefuroxime was started. However, the patient spent only one inpatient day there because she refused to continue the prescribed treatment.

Two days later, the patient sought care at another hospital. There, repeated blood tests revealed the exacerbation of the inflammation. The patient was admitted to the de-

partment of internal medicine and the same empirical antibiotic therapy was continued. However, the primary source of infection was not found. In addition, the patient's condition was complicated by the episode of paroxysmal atrial fibrillation, which was terminated by intravenous amiodarone. After four inpatient days, the patient refused further medical treatment and was discharged home.

After one more week, the patient returned to the Lithuanian University of Health Sciences Hospital Kauno klinikos ED complaining of general weakness, headache, progressive weakness, numbness, and pain in both legs and arms. Neurological examination showed a decrease in strength on the Medical Research Council Scale for Muscle Strength: strength in both arms proximally was 5/5, distally 3/5 on the right and 4/5 on the left, in both legs proximally – 4/5, dorsiflexion on both sides – 1/5, plantar flexion on the right – 3/5, on the left – 4/5. Reflexes were 2+ and symmetric in the arms and absent in the legs. Sensation to the light touch and pain was decreased in the right hand, in both feet, and on the lateral side of the right calf. Vibration sense was absent in the feet. Laboratory tests showed anemia, leukocytosis, thrombocytosis, elevated C reactive protein, and slightly altered renal function (Table 1). Urinalysis revealed slight glucosuria and proteinuria. A head CT was repeated; however, it showed no abnormalities. Based on the progressive motor and sensory deficits in the extremities, acute inflammatory demyelinating polyneuropathy was suspected and a second lumbar puncture was performed. Although CSF analysis was complicated by artificial blood contamination, only a slightly elevated protein level was found (0.47 g/L; Table 1). The patient was admitted to the Department of Neurology for further investigation and treatment.

During hospitalization in the Department of Neurology, a thorough investigation was performed to determine the cause of polyneuropathy and the source of the inflammation. Blood and urine cultures were negative. Chest X-ray, abdominal and cardiac ultrasound, and examination of the oral cavity and teeth showed no evidence of infection. Empiric antibiotic therapy was administered starting with cefuroxime. However, after two days, inflammatory markers remained highly elevated and antibiotic therapy was escalated to ampicillin/sulbactam. Laboratory tests for chronic infections (syphilis, human immunodeficiency virus, Lyme disease, and tick-borne encephalitis) were negative. However, an elevated erythrocyte sedimentation rate was found (95 mm/h). NCS showed abnormalities consistent with axonal sensorimotor polyneuropathy. In the upper extremities, NCS revealed asymmetric axonal damage involving both sensory and motor nerves, predominantly in the right. In the lower extremities, symmetric motor axonopathy was found, while the conduction of sensory nerves was intact (Table 2). Although the results of NCS ruled out the possibility of acute inflammatory demyelinating polyneuropathy, the etiology of peripheral nerve lesions remained unknown. Electromyography showed minimal fibrillations in the first dorsal intraosseous muscle without any spontaneous activity in L4-S2 and C8-T1 myotomes. Voluntary movements were impossible to assess due to the patient's weakness.

Table 2. **Patient NCS findings**

Nerve site	R/L	Response amplitude average (mV)	Distal latencies average (ms)	NCV average (m/s)
Motor				
n. medianus	R	0.25	10.2	42.7
	L	2.9	8	48.5
n. ulnaris	R	0.25	8.4	51.8
	L	2.1	8.4	49.2
n. peroneus	R	0.2	2.7	76
	L	0.2	3.6	47
n. tibialis	R	0.4	8.5	53
	L	0.2	8.7	40
Sensoric				
n. medianus, n. ulnaris	R	1.7	–	42.8
	L	15.2	–	56.5
n. radialis superficialis	R	45.8	–	82
	L	40.1	–	58
n. peroneus superficialis	R	6.1	2.1	48.5
	L	6.3	1.8	54
n. suralis	R	12.1	1.9	56
	L	10.2	1.9	54

R – right; L – left; NCV – nerve conduction velocity.

During the investigation period, the patient's symptoms progressed and she developed generalized myalgia and arthralgia. On the 5th day of hospitalization, biochemical blood tests revealed significantly increased creatinine and urea levels indicating the development of acute renal failure (Table 1). The patient was consulted by a nephrologist and additional laboratory tests were recommended including antinuclear antibodies, anti-neutrophil cytoplasmic antibodies, antibodies to myeloperoxidase (anti-MPO), leukocyte proteinase 3 and glomerular basement membrane, kappa and lambda free light chains ratio, serum protein electrophoresis, and monoclonal protein in the blood.

On the 6th day, the patient complained of dyspnea and abdominal pain. A thoracic X-ray was done and showed bilateral hydrothorax with pulmonary edema. The same evening oxygen therapy was started. The patient's saturation was 96% while she was receiving oxygen through a nasal cannula at a rate of 7 L/min. However, the oxygen demand gradually increased to 15 L/min, and the patient developed tachypnea and cyanosis. As a result of acute respiratory insufficiency, the patient was transferred to the intensive care unit (ICU). Due to the progression of acute renal failure, hemodialysis was started on the same night and continued throughout the whole stay in the hospital. The patient spent 3 inpatient days in the ICU, after which she was transferred back to the Department of Neurology.

A few days later, the test of IgG lambda light chain monoclonal protein was positive. Moreover, laboratory tests showed elevated levels of both lambda (111 mg/L) and kappa free light chains (485 mg/L), and their ratio (4.37). The patient was consulted by a hematologist, and the diagnosis of multiple myeloma was suspected. To confirm or reject it, a trepanobiopsy was performed. However, it showed no abnormalities.

Shortly thereafter, positive results for perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA) (titre 1:1000) and anti-MPO (titre 1:320) were received. The patient was consulted by a rheumatologist, and a preliminary diagnosis of systemic vasculitis was stated. For further examination and treatment, the patient was transferred to the Department of Rheumatology. On the 2nd day in the Department of Rheumatology, the patient developed a second episode of acute respiratory failure and a sharp increase in blood pressure (220/140 mmHg), which was complicated by impaired consciousness. Therefore, she was transferred to the ICU for the second time. A day later, vital signs stabilized, and the patient was transferred back to the Department of Rheumatology. However, two days later, respiratory insufficiency reappeared, and the patient was transferred to ICU for the third time where she spent another 2 days.

When the patient's condition finally stabilized, a renal biopsy was performed to confirm the diagnosis of systemic vasculitis. The results were positive for active focal crescentic glomerulonephritis caused by ANCA-associated vasculitis, a microscopic polyangiitis. To induce remission, physicians administered methylprednisolone 48 mg orally and cyclophosphamide 1000 mg intravenously. For neuropathic pain, gabapentin 300 mg per day was prescribed. Throughout the entire hospital stay, the patient underwent 25 sessions of hemodialysis. Although the patient's condition was not cured (Table 1), the patient refused further stay in the hospital, thus she was discharged. It was recommended to continue methylprednisolone 32 mg per day, antihypertensive medications, and regular hemodialyses in outpatient treatment. Moreover, two weeks after discharge, a second cyclophosphamide infusion was planned in the Day Care Unit, but the patient did not appear.

One month after admission to the Department of Neurology, the patient's neurological condition was reassessed, but it remained stable.

DISCUSSION

We present a clinical case of systemic ANCA-associated vasculitis that illustrates the rapidly progressive course of this disease involving many vital organs, as well as the nervous system. In this case, the first clinical presentation of systemic vasculitis was peripheral nerve damage, which means that the neurologist was the leading physician who examined and treated the patient. It emphasizes the importance of our ability to recognize this systemic disease as early as possible in order to be one step ahead of life-threatening complications.

At the time of the diagnosis, our patient was 73 years old. Research shows that MPA out of all AAVs has the latest onset (on average 61.8 years), while Wegener's granulomatosis and Churg-Strauss syndrome usually develop before the fifth decade [7].

In our case, the primary symptoms of the patient were blurry vision, headache, and limb weakness. According to research, AAV mostly presents with general (93.6%), renal (63.8%), and respiratory symptoms (59.6%) [8], however, the first symptoms in our patient did not meet the typical presentation. At first, the patient had only general symptoms, which presented with general weakness, headache, and elevated inflammatory markers in the blood. Subsequently, acute renal failure developed, although typical pulmonary symptoms were absent. Evidence shows that approximately 80% of patients with MPA have pulmonary complaints such as cough, hemoptysis, and dyspnea. On chest X-rays, pulmonary infiltrates are present in 92% of cases [9]. However, at the beginning of the patient's examination, a chest X-ray was performed several times and showed no abnormalities. Although acute pulmonary failure developed later, it was an expression of decompensation of acute renal failure.

Usually, when the nervous system is affected, patients first complain of deep pain in the proximal parts of extremities, burning pain in the skin, and focal weakness in the region of one nerve. These symptoms are usually of acute to subacute onset [6], which correlates to our patient's clinical course. As the disease progresses, it affects other individual nerves, and multiple mononeuropathies are the main presentation of the disease. Alternatively, many overlapping mononeuropathies can sometimes imitate symmetric or asymmetric distal polyneuropathy [10]. This clinical case is an excellent example of this phenomenon, exemplified by bilateral asymmetric muscle weakness in the distal ends of both upper and lower extremities. Moreover, patellar and Achilles reflexes were absent in both legs. Research shows, that pain is significantly more common in mononeuropathy and mononeuropathy multiplex. However, considering the individual nerves affected in mononeuropathies, 32% of these cases are painless [7]. It should be noted that our patient complained of the whole-body muscle and joint pain, which was of acute-subacute origin. Furthermore, although NCS showed symmetric polyneuropathy in the lower extremities, neurological examination also revealed features of multiple mononeuropathies. Despite bilateral distal muscle weakness, the patient's sensory symptoms were asymmetric: she had hypoesthesia in the right palm, both feet, and on the lateral side of the right calf. Interestingly, the most frequent nerve to be affected by AAV is found to be the peroneal nerve [7], which corresponds to the hypoesthesia in the lateral side of the right calf in our patient.

NCS is usually performed in all patients with acute or subacute polyneuropathy to differentiate its type and exclude neurological emergencies such as GBS. NCS in VNs

Table 3. **Findings consistent with VN in the presented clinical case**

Factors	Case findings consistent with VN
Course of the symptoms	Acute to subacute
Age	Elderly patient
Accompanying symptoms	Renal insufficiency, blurry vision, headache, arthralgia, myalgia
Neuropathy	Multiple sensorimotor mononeuropathies
Nerve conduction study	Axonal damage symmetric in the legs and asymmetric in the arms
Cerebrospinal fluid	Slightly elevated protein level
Laboratory results	Elevation of the inflammatory markers, positive p-ANCA, positive anti-MPO
Renal biopsy	ANCA-associated focal crescentic glomerulonephritis

Abbreviations: p-ANCA – perinuclear anti-neutrophil cytoplasmic antibodies; anti-MPO – anti-myeloperoxidase antibodies; ANCA – antineutrophil cytoplasmic antibodies.

usually shows reduced sensory and motor amplitudes with normal or mildly reduced conduction velocities, indicative of axon loss. Moreover, these changes are frequently found in multiple individual nerves indicating mononeuritis multiplex, whereas length-dependent axonal damage may also be present [7, 11]. In the presented clinical case, NCS showed axonopathy, which was asymmetric in the upper extremities and symmetric in the lower extremities. Due to the broad differential diagnostic of axonal polyneuropathies, it is challenging to identify its etiology exclusively by NCS and clinical presentation. Evidence shows that sonography of the large arm and leg nerves and brachial plexus can be performed to diagnose VN. Sonographic enlargement of the nerve can indicate VN and be a specific and sensitive method for this diagnosis [12]. However, to date, peripheral nerve biopsy remains the only diagnostic method to confirm VN [4]. Conversely, peripheral nerve biopsy can sometimes be uninformative since vasculitic lesions are focal and dispersed along the course of the nerve [13].

In our case, the lumbar puncture was performed in order to differentiate the cause of neuropathy and exclude a possible neuroinfection or GBS. Research shows that in VN, lumbar puncture can show elevated protein levels (>0.45 mg/g) in 51.4% of cases, whereas in 8.1% of cases, CSF can show pleocytosis. It was found that protein levels higher than 1.10 g/L were found only in cases when vasculitis was excluded [14]. In our case, there were two lumbar punctures performed. The first one showed no abnormalities, while the second one, although contaminated by blood, showed a slightly elevated protein level (0.47 g/L) which is consistent with the research findings.

During the initial examination of the patient, autoimmune etiology was considered. However, due to the lack of more common clinical features, selective biomarkers of vasculitis were tested only after an acute renal insufficiency occurred. Some studies show that eventually, 50-95% of AAV patients develop renal involvement. In the case of MPA, the most common renal pathology is a rapidly progressive glomerulonephritis [15]. Usually, proteinuria, microscopic hematuria, and sediment abnormalities are the most prevalent parameters indicating kidney involvement [16]. It should be noted that the patient's urinalysis revealed leukocyturia, glucosuria, and proteinuria only on her second admission to the ED. This suggests that a more thorough examination should have been performed earlier, especially as the renal disease is a negative prognostic feature in AVV and is negatively associated with patient survival [16].

Moreover, multiple myeloma was suspected due to acute renal insufficiency, as well as a positive IgG lambda light chain monoclonal protein, elevated concentrations of lambda and kappa free light chains, and their ratio. To date, there is no conclusive explanation for the light chain elevation in AAVs, but there are clinical cases describing similar findings [17, 18]. On the other hand, a case report of multiple myeloma with positive ANCA, PR-3, and MPO antibodies has been published [19]. It is hypothesized that inflammatory stimulation in the context of autoimmune disease may trigger the development of hematological malignancies including monoclonal gammopathy of undetermined significance [20]. It emphasizes the role of biopsy in the diagnostic work-up of patients presenting with contradictory findings of laboratory tests. Our patient underwent both a trepanobiopsy and a renal biopsy, whereas only the latter was pathological and confirmed the diagnosis of ANCA-associated vasculitis.

VN is difficult to diagnose because its symptoms vary. In the presented case, 42 days passed from admission to the ED to the diagnosis of MPA. This suggests that differential diagnosis for this disease remains challenging. Although a plethora of research has been conducted to better categorize the symptoms of this disease, due to its rarity and various clinical manifestations it is always a diagnosis of exclusion. Despite the magnitude of different symptoms, there were several key elements in our case that were particularly consistent with VN (Table 3). The combination of these findings can help to suspect the vasculitic origin of neuropathy.

CONCLUSIONS

AAVs are a group of rare autoimmune diseases that can affect many organs and tissues, as well as the nervous system. However, because of its rarity, AAV is frequently

overlooked in the initial examination of the patient with acute onset polyneuropathy. We presented the case of a 73-year-old woman with microscopic polyangiitis, whose primary symptoms were headache, blurry vision, and muscle weakness. In this case, VN manifested as both symmetric and asymmetric neuropathies with evidence of axonal damage in NCS. Despite a broad differential diagnosis and extensive examinations, the final clinal diagnosis of ANCA-associated vasculitic neuropathy was reached only after renal insufficiency occurred. Due to a wide variety of symptoms, their rarity, and alterations, vasculitic neuropathies can cause one of the greatest difficulties in differential diagnosis in cases of acute onset polyneuropathies.

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SU ANCA SUSIJUSIO VASKULITO SUKELTA POLINEUROPATIJA: KLINIKINIO ATVEJO PRISTATYMAS

Santrauka

Su antineutrofiliniais citoplazminiais antikūnais (ANCA) susiję vaskulitai (AAV) yra autoimuninių ligų grupė, kuri gali pažeisti daugelį organų ir audinių, taip pat nervų sistemą. Esant periferinių nervų pažeidimui, AAV gali sukelti vaskulitinę neuropatiją (VN). VN dažniausiai sukelia asimetrinį raumenų silpnumą ir jutimo sutrikimus, skausmą distalinėse galūnių dalyse. Elektroneuromiografija įprastai parodo kelių atskirų nervų aksonopatiją, nors taip pat galimas ir nuo ilgio priklausantis nervinių skaidulų pažeidimas. Kadangi AAV yra reta liga ir gali pasireikšti įvairiais klinikiniais požymiais, ji retai įtraukiama į pradinį paciento, sergančio polineuropatija, ištyrimą. Be to, vienintelis VN diagnosti-

kos metodas, patvirtinantis ligą, yra periferinio nervo biopsija. Dėl šių priežasčių AAV sukeltos VN yra vienas didžiausių iššūkių, siekiant diferencijuoti polineuropatijos kilmę. Šiame straipsnyje pristatome 73 metų amžiaus pacientę, kuri kreipėsi į Skubios pagalbos skyrių dėl neryškaus matymo, galvos skausmo ir galūnių silpnumo. Neurologinio ištyrimo duomenys parodė polineuropatijai būdingus pakitimus. Pacientei buvo atlikti išsamūs diagnostiniai tyrimai, tačiau, nesant tipinių sisteminių simptomų, VN buvo diagnozuota tik išsivysčius ūmiam inkstų funkcijos nepakankamumui. Šis atvejis parodo AAV klinikinių simptomų įvairovę ir išsamios diferencinės diagnostikos svarbą pacientams, tiriamiems dėl dauginės mononeuropatijos.

Raktažodžiai: su ANCA susiję vaskulitai, mikroskopinis poliangitas, dauginės mononeuropatijos, vaskulitinės neuropatijos.

Gauta: 2023 01 27

Priimta spaudai: 2023 04 21