ANCA-associated vasculitis patients treated in Polish intensive care units – retrospective characteristics based on the POLVAS registry

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Abstract

Background: ANCA-associated vasculitides (AAV) is a group of rare disorders where inflammation and damage of the small blood vessels lead to dysfunction of the supplied organs. In severe flares of the disease patients may require intensive care unit (ICU) admission and treatment. The study aims to characterize Polish patients with AAV who were admitted to the ICU and compare them to the others.

Methods: An observational, retrospective study based on the POLVAS – registry of Polish adult patients with AAV was carried out. Patients admitted to the ICU (ICU group) were identified and compared with the patients who did not require ICU admission (non-ICU group). Characteristics and comparison between groups were made using standard statistic descriptive methods.

Results: 30 patients admitted to the ICU were identified among 573 cases included in the registry. All patients in the ICU group with available data were ANCA positive. The clinical manifestations related to the ICU admission were respiratory, renal and central nervous system involvement. The treatment regimen for remission induction was similar in both groups. Almost half of the patients in the ICU-group (48.3%) required dialysis, whereas in the non-ICU group it was 21.8% ($P = 0.01$). Infections were also more frequent in the ICU group (72.4% vs. 36.9% $P < 0.001$). The mortality rate among patients who needed ICU treatment was significantly higher when compared to the rest of the patients (53.6% vs. 7.8%; $P < 0.001$).

Conclusions: In the Polish AAV cohort one in twenty patients required ICU admission. This group was characterized by multiple organ involvement and high mortality.

Key words: autoimmune diseases, vasculitis, intensive care unit, ANCA, ICU, AAV.

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TABLE 1. Types of ANCA-associated vasculitides

Name	Abbreviation	Most frequent type of antibodies	
		Indirect immunofluorescence (IIF) test	ELISA test
Granulomatosis with polyangiitis	GPA	c-ANCA	Anti-PR3
Microscopic polyangiitis	MPA	p-ANCA	Anti-MPO
Eosinophilic granulomatosis with polyangiitis	eGPA	p-ANCA	Anti-MPO

ELISA – enzyme-linked immunosorbent assay, p-ANCA – perinuclear immunofluorescence ANCA pattern, c-ANCA – cytoplasmic immunofluorescence ANCA pattern, MPO – myeloperoxidase, PR-3 – proteinase 3

ANCA-associated vasculitides (AAV) is a group of three disorders in which inflammation and damage of the small blood vessels are correlated with the presence of antineutrophil cytoplasmic antibodies (ANCA). It is considered to be a rare disease with an incidence between 12 and 33 cases/1 million population/1 year [1].

The main clinical types of AAV, based on Chapel Hill classification [2], are granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (eGPA). Details are listed in Table 1.

In rare cases, AAV may be diagnosed based on clinical presentation and pathological findings without ANCA antibodies present [3]. The main pathomechanism consists of an immune-mediated inflammatory process that occurs in the walls of small vessels. Necrosis of these vessels leads to dysfunction of the supplied organs [3]. The clear causes of such an autoimmune reaction are still unknown. Clinical manifestation of the AAV can vary from single-organ involvement to rapidly progressing systemic disease. Almost every organ can be involved, although in the intensive care unit (ICU) setting the most important and potentially life-threatening manifestations are pulmonary, renal, and neurological [4].

Pulmonary involvement may lead to diffused alveolar haemorrhage with symptoms like cough, dyspnoea, and haemoptysis. Laboratory and imaging findings include decrease of haemoglobin concentration (Hb) in complete blood count (CBC), ground-glass opacities in chest X-ray and chest computed tomography (CT), blood-stained discharge in bronchoscopy, and hemosiderin loaded macrophages in broncho-alveolar lavage (BAL) [5, 6]. Other common respiratory tract manifestations are lung granulomas, bronchi mucosa ulcers, and tracheal or subglottic stenosis.

Renal involvement presents as glomerulonephritis with progressive (often rapidly) renal failure. Initially it can be asymptomatic. Typical laboratory findings are proteinuria, active urinary sediment with red blood cells and granular casts, and increased creatinine and urea serum concentration [7].

Neurological manifestation typically presents as mononeuritis multiplex. Much rarer, but potentially far more dangerous, is the central nervous system (CNS) involvement, which may lead to ischaemic or haemorrhagic stroke [8].

Diagnosis

The course of AAV is characterised by flares and remissions. In the case of a suspected flare of a disease, it is important to distinguish the disease from the complications of immunosuppressive treatment, like sepsis. Another ICU challenge is the diagnosis of the onset of the disease, which can be fulminant and life-threatening [9]. When pulmonary and renal disfunction coexist, so-called pulmonary-renal syndrome can be suspected and the diagnosis of vasculitis is very probable. Two main causes of pulmonary-renal syndrome are AAV and anti-glomerular basement membrane disease (Goodpasture syndrome – GPS) [10]. Immunological tests, such as ANCA screening, as well as rheumatology or clinical immunology consult, may allow a diagnosis to be made without delay. When possible, obtaining the samples for histopathological examination (e.g. kidney biopsy) may be extremely helpful to establish the diagnosis and severity of the disease, and hence to determine further procedures.

Treatment

Treatment of AAV is based on immunosuppression with glucocorticosteroids and additional immunosuppressants, such as cyclophosphamide or rituximab. It is carried out in two stages: intensive immunosuppressive treatment to induce disease remission, followed by milder maintenance therapy. In the ICU setting, in cases of AAV patients, induction therapy often requires an aggressive approach [11] and can be combined with interventions such as mechanical ventilation, continuous renal replacement therapy, and therapeutic plasma exchange [12]. There are also reports mentioning use of ECMO in diffused alveolar haemorrhage due to AAV [13, 14].

Patients with AAV admitted to an ICU can also suffer from severe infection and sepsis due to immunosuppressive treatment. Therefore, thorough

microbiological culture testing and broad-spectrum antibiotics when needed are essential.

POLVAS registry

The initiative named POLVAS is the Consortium of the Polish Vasculitis Registry, which was established to gather data on Polish adult vasculitis patients. A low incidence of AAV makes it impossible for a single centre to design and pursue clinical trials with a substantial number of patients; therefore, POLVAS was created by nine centres [15].

The presented research is based on the retrospective part of the POLVAS registry database. The main aim of the study is to characterise Polish patients with AAV who were admitted to the ICU and compare them to those who did not need such treatment.

METHODS

This is a multicentre, retrospective, observational, registry-based study on patients diagnosed with AAV between 1990 and 2016.

The study was carried out in accordance with the ethical principles of the Declaration of Helsinki developed by the World Medical Association. The study protocol was approved by the Jagiellonian University Bioethics Committee (Krakow, Poland) (approval no. 122/6120/25/2016). All POLVAS participating centres acquired Local Ethics Committee approval. Informed consent was obtained from the participants.

All included patients were diagnosed with vasculitis according to the American College of Rheumatology (ACR) classification criteria [16] and the 2012 Revised International Chapel Hill Consensus criteria [2]. Demographics, laboratory test results, clinical data, and treatment details were collected from the patients' medical records using an electronic form. The characteristics of the entire cohort are described in separate manuscripts [17, 18]. The presented analysis concerns comparison of the AAV patients admitted to the ICU (ICU group) to patients who did not require ICU admission (non-ICU group). ICU admission was defined in the form as "Severe disease flare requiring ICU admission".

Standard descriptive statistics were used. Normal distribution of variables was checked by the Shapiro-Wilk test, and homogeneity of variances was assessed by Levene's test. To compare the studied groups the χ^2 test (with Yates correction if needed) and Mann-Whitney *U* test were used. The *P*-value < 0.05 was considered as statistically significant, modified with Bonferroni correction when multiple comparisons were performed. The assumed level of significance for multiple comparisons according to Bonferroni correction equalled 0.017.

Calculations were performed with Statistica 13 software (StatSoft®, Tulsa, OK, USA).

RESULTS

Among 573 cases included in the retrospective POLVAS database, there were 30 cases (5.24%, 30/573; 18 males; $P = 0.21$) who were admitted to the ICU. Median time of observation (defined as the difference between the date of enrolment to the database and the date of the diagnosis) in the ICU group equalled three years (2.0–8.0), which was similar comparing to the non-ICU group (4 years, 2.0–8.0; $P = 0.98$). All patients in the ICU group were ANCA positive (in five cases there was no data regarding ANCA status), whereas 9% of cases in the non-ICU group were ANCA negative. MPA diagnosis, p-ANCA presence in IF test as well as anti-MPO presence in ELISA assay were associated with the risk of ICU admission ($P < 0.01$). The respiratory system was affected in 93.3% of ICU cases. Pulmonary, renal, CNS, and eye involvement were significantly more frequent in the ICU group ($P = 0.03$; $P = 0.01$; $P < 0.01$; $P = 0.03$). There were also more infections and more deaths in the ICU group compared to the non-ICU group (both $P < 0.01$). The details are presented in Table 2.

Initial treatment for remission induction was analysed. The main trends based on glucocorticosteroids and cyclophosphamide were the same in both groups; however, the cyclophosphamide cumulative dose was significantly higher in the non-ICU group, reaching 8.0 g (median: 4.7–15.0 g, $P < 0.01$). Therapeutic plasma exchange was used similarly in both groups, but intravenous immunoglobulins were more frequently given to the patients who needed ICU treatment during the course of disease (17.2% vs. 4.8%, $P < 0.01$). Almost half of the patients in the ICU-group (48.3%) required dialysis treatment at some point, whereas in the non-ICU group it was only 21.8% ($P = 0.01$). Details about the treatment are given in Table 3.

DISCUSSION

Generally, ANCA-associated vasculitides are diagnosed and treated in specialised internal medicine departments, like rheumatology, nephrology, or pulmonology. The majority of the patients do not require ICU admission. The data in the registry were gathered in the academic centres from across Poland, covering about 60% of the Polish population [17]. Our study shows that only 5.24% of all investigated AAV patients were admitted to an ICU. This is a relatively low number compared with other studies, in which 12–14% of AAV patients were treated in an ICU [19, 20]. This is probably due to the retrospective character of the presented part of the reg-

TABLE 2. The differences between the subgroup of cases who were admitted to the ICU and the subgroup of cases who were not treated in the ICU

Parameter	ICU group	Non-ICU group	*P*-value
Cases	30	543	–
Men	18/30, 60%	262, 48.3%	0.2101
Median observation (years)	3.0 (2.0–8.0)	4.0 (2.0–8.0)	0.9790
MPA	12/30, 40.4%	93/543, 17.1%	**MPA/GPA: 0.0047*** MPA/EGPA: 0.0371* GPA/EGPA: 0.4733*
GPA	17/30, 56.7%	385/543, 70.9%	
EGPA	1/30, 3.3%	65/543, 12.0%	
p-ANCA presence	13/25, 52.0 %	110/466, 23.6 %	**0.0014**
c-ANCA presence	12/25, 48.0%	310/466, 66.5%	0.0576
No ANCA	0/25, 0.0%	42/466, 9.0%	–
Anti-MPO presence	14/26, 53.8%	119/462, 25.8%	**0.018**
Anti-PR3 presence	13/26, 50.0%	321/477, 67.3%	0.0690
Cigarette smoking	5/16, 31.3%	143/375, 38.1%	0.5783
Infections	21/29, 72.4%	188/510, 36.9%	**0.0001**
Deaths	15/28, 53.6%	41/529, 7.8%	**< 0.0001**
Organ involvement			
Constitutional symptoms	24/30, 80.0%	455/540, 84.3%	0.5353
Musculo-skeletal system	21/30, 70.0%	307/538, 57.1%	0.1627
Skin	10/30, 33.3%	180/539, 33.4%	0.9944
ENT	16/30, 53.3%	354/543, 65.2%	0.1861
Eye	11/30, 36.7%	108/536, 20.1%	**0.0307**
Respiratory system	28/30, 93.3%	401/539, 74.4%	**0.0335**
Cardiovascular system	6/29, 20.7%	83/541, 15.3%	0.4396
Gastrointestinal system	7/30, 23.3%	63/541, 11.6%	0.0574
Renal	26/30, 86.7%	332/539, 61.6%	**0.0101**
CNS	7/30, 23.3%	42/539, 7.8%	**0.0031**
Peripheral neurological system	9/30, 30.0%	114/534, 21.3%	0.2642

Statistically significant *P*-values are shown in bold (assumed level of significance = 0.05)

* Assumed level of significance for multiple comparisons according to Bonferroni correction equals 0.017

ICU – intensive care unit, MPA – microscopic polyangiitis, GPA – granulomatosis with polyangiitis, EGPA – eosinophilic granulomatosis with polyangiitis, ANCA – antineutrophil cytoplasm antibodies, p-ANCA – perinuclear immunofluorescence ANCA pattern, c-ANCA – cytoplasmic immunofluorescence ANCA pattern, MPO – myeloperoxidase, PR-3 – proteinase 3

TABLE 3. Remission induction treatment modalities between ICU and non-ICU groups

Parameter	ICU group	Non-ICU group	*P*-value
GCs oral	15/29, 51.7%	338/543, 62.2%	0.2560
GCs iv	24/29, 82.8%	402/543, 74.0%	0.2101
CYC	27/29, (93.1%)	435/543, 80.1%	0.1368
RTX	4/29, 13.8%	40/543, 7.4 %	0.3640
TPE	6/29, 20.7%	65/535, 12.1%	0.1769
IVIG	5/29, 17.2%	26/543, 4.8%	0.0039
GS pulses*	24/29, 82.8%	347/475, 73.1%	0.2496
CTX cumulative dose in grams (median)	5.0 (2.0–8.0)	8.0 (4.7–15.0)	0.0084
RTX cumulative dose in grams (median)	2.4 (1.15–3.75)	2.0 (1.5–2.8)	0.7311
Dialysis	14/29, 48.3%	116/533, 21.8%	0.0010

Statistically significant *P*-values are bolded (assumed level of significance = 0.05)

* GS pulse was defined as at least one dose of ≥ 500 mg of methylprednisolone (or equivalent)

GCs – glucocorticoids, CYC – cyclophosphamide, RTX – rituximab, TPE – therapeutic plasma exchange, IVIG – intravenous immunoglobulins

istry with no follow-up. In addition, ICU admission criteria vary across countries [21, 22].

Knowledge of the main clinical manifestations can be valuable for the intensivists. Published studies show vasculitis as one of the most frequent autoimmune disease in the ICU [12, 23].

It is reported that AAV can have fulminant onset, and a significant number of diagnoses – reaching 10% – were first diagnosed in the ICU [9]. Unfortunately, in our study we lack data on whether the patients were diagnosed at the ICU. Moreover, information on whether it was the first or a subsequent flare that resulted in the ICU admission was also not included in the registry.

All patients with disease flares requiring ICU treatment presented multiorgan involvement. The most common manifestations in the ICU group were respiratory (93%) and renal (83%), which is consistent with other studies [9, 19, 20, 24]. Respiratory, re-

nal, and central nervous system manifestations were found more often in the ICU-group than in the non-ICU group. In the study by Demiselle *et al.*, patients with AAV treated in an ICU were compared to the control group treated in medical units; respiratory and CNS involvement were also more common in the ICU group, but not renal involvement [4]. It could be influenced by the fact that the control group was enrolled from the nephrology centres, where renal emergencies can be managed outside the ICU.

The treatment of the AAV has significantly changed over time. The last decade has brought a better understanding of immunosuppressive treatment with reduced doses of glucocorticosteroids and cyclophosphamide or new biological immunosuppressants, like rituximab. The registry's inclusion period is large; therefore, treatment differences may result from the state of the knowledge at the time as well as the availability of some methods. Our study shows that the general remission induction treatment regimen had no relation to the need for ICU treatment. However, a higher cumulative dose of cyclophosphamide in the non-ICU group could indicate that more aggressive immunosuppression can lead to better results, especially because the mortality was almost seven times higher in the ICU group, reaching 53.6%. Such high mortality may also be related to the fact that infections were twice as frequent as in the non-ICU group.

In another study by our team, we showed that patients with vasculitides have a worse prognosis than other autoimmune disease patients treated in the ICU [25].

AAV patients who needed ICU admission more often required intravenous immunoglobulins (IVIG). This may be due to the aforementioned registry's inclusion period, because 20 years ago IVIG therapy was more available than therapeutic plasma exchange. On the other hand, therapeutic plasma exchange frequency was comparable in both groups. Recently published results of the PEXIVAS trial by Walsh *et al.* showed that in ANCA-positive patients with renal exacerbation or alveolar haemorrhage, plasma exchange did not reduce end-stage kidney disease incidence or death [26].

Our study has several major limitations. It is based on retrospective data gathered in a registry and is focused on the overall medical characteristics of the cohort. We lack data on specialist ICU procedures and prognostic scales scores. Moreover, we were not able to establish from the registry's dataset any details of the ICU admissions. One of the biggest disadvantages of POLVAS registry is the scarcity of data on infectious complications. However, to our knowledge, it is the first attempt to estimate the problem of AAV in Polish ICUs. Hopefully the prospective part of the POLVAS registry will give more details on the matter.

CONCLUSIONS

In the Polish AAV cohort one in 20 patients required ICU admission. In this group respiratory, renal and central nervous system involvement was more often observed. The mortality was high. More prospective observational studies are needed to provide the full characteristics of AAV treated in ICUs in the Polish population.

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