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Research Article

Predictors of all-cause Mortality and Infection During inducing Treatment in ANCA-Associated Vasculitis: a Population-Based Cohort Study

Wenbo Zhu, Wenyi Wu, Linlin Huang *, Sheng Chen

[1]The 2nd Department of Hepatobiliary Surgery, Meizhou People's Hospital, Meizhou, China.

[2]The 1st Department of Hepatobiliary Surgery, Meizhou People's Hospital, Meizhou, China.

***Corresponding Author:** Tianming Lian, The 2nd Department of Hepatobiliary Surgery, Meizhou People's Hospital, Meizhou, China.

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Abstract:

Objective. To determine the predictors of all-cause mortality and infection during inducing treatment in a population-based cohort of ANCA-associated vasculitis (AAV).

Methods. The study included 198 cases of AAV diagnosed in Renji Hospital from May 2011 through May 2020. Outcome data were collected during follow-up of AAV patients. Severe infection events (requiring hospitalization and treatment with intravenous antibiotics) during induction therapy were also identified. Demographic, clinical and laboratory results were tested as potential predictors in multivariable models.

Results. A total of 198 patients were followed from time of disease diagnosis to August 2020 or death. 168 patients are alive with a median follow up of 36 (20-60) months. Leading causes of death within 6 months were infection (42%), respiratory failure (25%) and renal failure (17%). After 6 months, the major causes of death were infection (39%), respiratory failure (22%), cancer (17%) and cardiovascular / cerebrovascular events (11%). Multivariable analysis showed that older age (>65 years), higher Birmingham Vasculitis Activity Score (BVAS, ≥15), infection at diagnosis or during induction therapy and disease relapse (≥2 times) were independent predictors of all-cause mortality. Besides, higher S-creatinine, BVAS ≥15, myeloperoxidase (MPO) positivity and lack of prophylaxis with sulfamethoxazole (SMZ) were predictors of severe infection events during induction therapy.

Conclusion. Patients with older age and higher disease activity are at increased risk of death. The events of infection at diagnosis or during induction therapy and disease relapse also threaten patients' lives. Besides, poorer renal function, high disease activity and MPO positivity are associated with severe infection. SMZ prophylaxis during induction therapy may be an effective measure to reduce the risk of infection.

Key words: ANCA-vasculitis; mortality; infection; predictors of all-cause mortality; predictors of severe infection

Introduction

ANCA-associated vasculitis (AAV) is a group of autoimmune-related inflammation of small blood vessels with characteristic antineutrophil cytoplasmic antibodies (ANCA)[1]. Three phenotypic variants of AAV are defined, based on clinicopathological features: granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA) and eosinophil granulomatosiswith polyangiitis (EGPA)[2]. With the precision of experimental indicators, the number of patients diagnosed with AAV is increasing in recent years[3,4].

Multiple organs may be involved in AAV, such as the respiratory tract, eyes, lungs, kidneys, skin, and nervous system. Among which, the kidney and lung particularly threatening patients' health and lives. Poor outcomes are attributed to inappropriate treatments that have insufficient effects and toxic effects[5]. The treatment strategy of AAV has optimized with two phases: first aimed at inducing remission and the latter to maintain remission and prevent relapse. The induction therapy is usually the first 6 months after diagnosis. Severe infection in the early stage of

the disease, including infection at disease diagnosis and infection during induction therapy, is quite common in clinical practice[6,7]. Infection not only aggravates the injury of organs, but also makes it more difficult to apply effective treatment, resulting in the increases of disease activity. Actually, mortality is extremely high in the first year after disease onset due to active vasculitis and infections.In cohorts observed in Western countries, one year survival rate was around 82~95%while that in Japan was 79.1%[8,9].In an earlier study, Bourgarit et al. showed that active vasculitis was the leading cause of deaths in the first year and that treatment with corticosteroid alone was associated with early death[10]. The study from Massachusetts General Hospital found that the most prominent cause of death for patients was still infection compared with the general population. Nowadays, with the widespread use of immunosuppressants, most patients are treated by glucocorticoid combined with CTX or RTX. Despite their quick effectiveness, the two drugs have comparable adverse effects[11] of increasing the incidence of infection by causing hypogammaglobulinemia and leukopenia. Insufficient treatment leads to the relapse of disease while aggressive treatment put patients under over-immunosuppressants[12,13]. Therefore, it is of great significance to balance the potent treatment as well as the adverse events alongside by exploring the prognosis of AAV patients and risk factors for infection.

The aim of our study is to determine the predictors of all-cause mortality and infection during induction therapy in a population-based cohort of ANCA-associated vasculitis (AAV) patients from Renji Hospital.

Patients and Methods

Population

We identified an incident cohort of AAV patients diagnosed between May 2011 and May 2020 in Renji Hospital. All had a clinical diagnosis of small vessel vasculitis established by clinical manifestation, diagnostic histology, and positive ANCA. The patients were followed up until death or the end (November 2020) of the study. Patients with incomplete information or secondary vasculitis including allergic purpura, tumor, drug associated AAV and cryoglobulinemia were excluded. As well as those followed up less than 6 months. All patients met the 2012 Chapel Hill Consensus Conference definition. The study was approved by the Ethics Committee of Renji Hospital, and all patients gave written informed consent. The study was performed in accordance with the Declaration of Helsinki.

Definition

The activity of AAV was assessed by Birmingham Vasculitis Activity Score (BVAS). Relapse was defined as the new appearance or recurrence of one or more symptoms with a BVAS ratio ≥1 after remission and it was likely to be attributable to vasculitis disease activity and not to alternative pathology such as infection after appropriate investigation[14]. Infection in the early stage of the disease included infection at diagnosis without treatment and infection during induction therapy. Severe infection was defined as an infection that required hospitalization and intravenous antibiotics for at least 3 days. End stage renal disease (ESRD) was defined by demand of chronic dialysis and/or an eGFR < 15 ml/min/1.73 m^2 for 3 months. Estimated glomerular filtration rate (eGFR) was calculated by the Modification of Diet in Renal Disease formula[15].

Data collection

The baseline characteristics of these patients was recorded at diagnosis. The following data were collected: age, gender, ANCA serology, hemoglobin, albumin, erythrocyte sedimentation rate(ESR), C-reactive protein(CRP), 24h urine protein and creatinine(Cr). Severe infection during induction therapy was recorded. The follow-up was conducted by outpatient service or telephone calls. During the follow-up, major events including relapse, severe infection, ESRD, and death were recorded. Same laboratory data were collected at each relapse event. BVAS was calculated at diagnosis and each hospitalization. Death were also recorded. Of patients that died outside of hospital the exact cause of death could not be identified and was therefore designated unknown in these cases. Data of type and site of infection, pathogen results, and prescription of antibiotics were recorded when available.

Data analysis

Data are presented as median with interquartile range(IQR) when appropriate. Cox regression analysis was applied to investigate predictors of all-cause mortality. The following variables were selected: age at diagnosis, sex, organ involvement, BVAS, clinical diagnosis, ESR, CRP, 24h protein urine, albumin, S-creatinine, ANCA serology, relapse. Risk factors for all-cause mortality with a p-value < 0.10 in the univariate analysis were incorporated in the multivariate Cox regression model. Student's t-test for normally distributed variables or the Mann–Whitney U test fornon-normally distributed data was used to compare groups. Differences among categorical variables were quantified by Chi-square test or fisher's exat test—when Chi-square test was not appropriate.

To study the predictors of severe infection during induction therapy, only patients who did not get infected at diagnosis were included. Univariate and multivariate logistic regression models were developed to determine predictors of severe infection during induction therapy. The following variables were selected for univariate analysis: age at diagnosis, sex, BVAS, Hemoglobin, 24h protein urine, albumin, S-creatinine, ANCA serology, induction therapy, prophylaxis with SMZ. Risk factors for severe infection with a p-value < 0.10 in the univariate analysis were selected in the multivariate Cox regression model. IBM SPSS Statistics software v.25 and GraphPad Prism v.8 were used for all statistical calculations and diagram drawing. A P-value <0.05 was considered significant.

Results

Baseline characteristics and major events during follow up

The characteristics of 198 included patients(52% female, median age 64-year-old) were displayed in Table1. Among who, 168 patients (85%) were alive with a median follow up of 36 (20-60) months. Cases' median course of disease at diagnosis was 3 months with median BVAS 13. The number of PR3-ANCA potive and MPO-ANCA positive patients is 37 (19%) and 154 (78%) respectively while 6 patients (3%) were ANCA-negative. At the time of diagnosis, 153 patients (77%) exhibited involvement of multiple organ systems. Of these,113 patients (57%)presented with respiratory system involvement while 145 patients (53%) demonstrated renal involvement, with over half of these cases advancing irreversibly to end-stage renal disease. Renal(73%), pulmonary(57%) and muscular system(18%) were the top 3 most commonly involved organ systems. During follow up, 139(71%) patients got infected, 48(24%) experienced relapse more than 2 times and 62(31%) advanced to ESRD.

Predictors of all- cause mortality

In the course of observation, 30 (15%) of 198 patients died with median observe time of 7months (IQR 2-14). Main causes of death within 6 months were infection (43%), interstitial lund disease (21%) and renal failure (14%). After 6 months, the top3 causes turned into infection (38%), cancer (19%), cardiovascular and cerebrovascular events (13%) and renal failure (13%). The detailed information are summarized in Table 2. The cumulative mortality at 6 months, 1 year, and 2 years were 7%, 12%, and 16%, respectively. Univariate analysis exhibited age (>65 years)[HR,2.574; 95%CI 1.178-5.626; p=0.018], BVAS (≥15)[HR 3.233; 95%CI 1.535-6.810; p=0.002], female[HR 0.489; 95%CI 0.232-1.028; p=0.059], S-creatinine (>177μmol/l)[HR 3.218; 95%CI 1.529-6.77; p=0.002], infection in the early stage of AAV[HR 13.0721; 95%CI 1.780-95.993; p=0.012] and relapse (≥2times)[HR 8.353; 95%CI 4.051-17.225; p<0.001] were significantly differently between survivors and nonsurvivors of AAV patients (Table 2). Putting the above variables into the multivariable regression, the results showed that only age (>65 years)[HR 3.472; 95%CI 1.504-8.016; p=0.004], BVAS (≥15)[HR 2.561; 95%CI 1.103-5.492; p=0.029], infection in the early stage of AAV[HR 8.861; 95%CI 1.131-69.441; p=0.038] and relapse (≥2times)[HR 14.000; 95%CI 6.290-31.162; p<0.001] independently predicted the occurrence of death. Figure 1 demonstrated the survival relative to selected variables.

Predictors of severe infection during induction therapy

Table 4 recorded the detailed characteristics of 139 AAV patietns infected in the early stage of disease, with 97 at diagnose and 42 during induction therapy. Most of them were renal(109[78%])or lung(86[61%]) involved, which consisted with the overall population. The most significant site of infection was respiratory tract(86[62%]), followed by urinary tract (25[18%]) and gastrointestinal tract(6[4%]). Most infections in our cohort were caused by bacteria (91[65%]), with some caused by fungus (37[27%]) and a smaller proportion caused by virus(28[20%]). Among patients who were not infected at diagnosis, 42 of them got severe infection during induction treatment. Factors associated with the occurrence of severe infection were included in the univariable analysis:age, gender(female), BVAS(≥15), multiple systemic involvement, 24h protein urine(g/24h), S-creatinine, ANCA positivity, dose of prednisone, use of cyclophosphamide and prevention with SMZ (Table 5). However, only age[OR 1.037; 95%CI 1.001-1.072; p=0.038], BVAS(≥15)[OR 6.053; 95%CI 2.196-16.681; p<0.001], S-creatinine[OR 1.008; 95%CI 1.003-1.012; p<0.001], MPO-ANCA+[OR 0.172; 95%CI 0.045-0.658; p=0.01] and prevention with SMZ[OR 0.136; 95%CI 0.049-0.377; p<0.001] reached significance. The following multivariable analysis exhibited that only BVAS[OR 4.324; 95%CI 1.017-18.387; p=0.047], S-creatinine[OR 1.007; 95%CI 1.001-1.013; p=0.015], MPO-ANCA+[OR 8.291; 95%CI 1.175-58.492; p=0.034]and prevention with SMZ[OR 0.025; 95%CI 0.004-0.154; p<0.001] can independently predict the occurrence of severe infection.

Characteristics	Total (n=198)	Survivors (n=168)	Nonsurvivors (n=30)	P-value
Age, median (IQR), year	64 (57-71)	64 (57-71)	67 (61-72)	0.002
Female, n (%)	103 (52)	92 (55)	11 (37)	0.068
Course of disease, median (IQR), month	3 (1-8)	3 (1-8)	4 (1-12)	0.448
ANCA positivity, n (%)				
MPO- ANCA	154 (77)	130 (77)	24 (80)	0.751
PR3-ANCA	37 (19)	32 (19)	5 (17)	0.758
Double negative	6 (3)	5 (3)	1 (3)	0.932
BVAS, median (IQR)	13 (9-15)	11 (7-14)	17 (9-19)	0.142
System involvement, n (%)				
Multiple systemic involved	153 (77)	130 (77)	23 (77)	0.931
Kidney	145 (73)	121 (72)	24 (80)	0.363
Lung	113 (57)	94 (56)	19 (63)	0.452
Muscle	36 (18)	30 (18)	6 (20)	0.779
ENT	33 (17)	30 (18)	3 (10)	0.426
Nervous system	21 (11)	18 (11)	3 (10)	0.975
Eyes	22 (11)	18 (11)	4 (13)	0.752
Laboratory results, median (IQR)				
ESR, mm/h	70 (40-94)	67 (38-98)	71 (35-87)	0.217
CRP, mg/L	24 (6-71)	18 (3-67)	24 (7-59)	0.630

24h protein urine, g/24h	0.8 (0.4-2.0)	0.8 (0.4-2.0)	0.9 (0.5-2.0)	0.942
S-creatinine, μmol/l	109 (61-257)	114 (62-245)	252 (97-474)	＜0.001
Therapy, n (%)				
CTX	117 (59)	95 (57)	22(73)	0.375
RTX	25 (13)	20 (12)	5 (17)	0.756
Others	56 (28)	53 (32)	3 (10)	0.248
Major events, n (%)				
Infection at diagnose or during induction therapy	139 (71)	111 (66)	28 (97)	0.001
ESRD	62 (31)	47 (28)	15 (50)	0.017
Relapse (≥2times)	48(24)	31 (18)	17 (57)	＜0.001

ENT: ear nose and throat; ESR: erythrocyte sedimentation rate;CRP: C-reactive protein;MPA: microscopic polyangiitis; GPA: granulomatosis with polyangiitis; EGPA: eosinophil granulomatosis with polyangiitis; MPO: myeloperoxidase; PR3: proteinase-3; GC: glucocorticoid;CTX: cyclophosphamide;RTX: rituximab;BVAS: Birmingham vasculitis activity score (range 0–63);IQR:interquartile range;ESRD: end stage renal disease

Table 1: Baseline characteristics of the cohort by outcome

Cause of death	Total (n=30)	Survival time ≤ 6months(n=14)	Survival time >6months (n=16)
Infection, n (%)	12 (40)	6 (43)	6 (38)
Interstitial lung disease, n (%)	5 (17)	3 (21)	2 (5)
Renal failure, n (%)	4 (13)	2 (14)	2 (13)
Cancer, n (%)	3 (10)	0	3 (19)
Alveolar hemorrhage, n (%)	2 (7)	1 (7)	1 (6)
Cardiovascular and cerebrovascular event, n (%)	2 (7)	0	2 (13)
Undetermined, n (%)	2 (7)	2 (14)	0

Table 2: Cause of death of nonsurvivors among ANCA associated vasculitis patients

Variable	Univariable		Multivariable	
	HR(95%CI)	p value	HR(95%CI)	p value
Demographic characteristics				
Age, >65 years	2.574(1.178-5.626)	0.018	3.472(1.504-8.016)	0.004
Sex, Female	0.489(0.232-1.028)	0.059	0.986(0.435-2.235)	0.973
Clinical manifestations				
Multiple system involved	1.010(0.433-2.354)	0.982		
Kidney	1.614(0.659-3.952)	0.295		
Lung	1.328(0.632-2.791)	0.453		
Muscle	1.044(0.426-2.557)	0.925		
ENT	0.526(0.159-1.733)	0.291		
Nervous system	0.897(0.272-2.959)	0.859		
Eyes	1.102(0.384-3.163)	0.856		
BVAS, ≥15	3.233(1.535-6.810)	0.002	2.561(1.103-5.492)	0.029
Laboratory data				
ESR, >70mm/h	1.229(0.601-2.515)	0.572		
CRP, >20mg/L	1.359(0.663-2.786)	0.403		
24h protein urine, >0.85g/24h	1.177(0.574-2.412)	0.656		
S-creatinine, >177μmol/l	3.218(1.529-6.770)	0.002	2.208(0.987-4.938)	0.054
MPO-ANCA+ (ref. PR3-ANCA+)	0.822(0.314-2.156)	0.691		
Major events during follow up				
Relapse,≥ 2 times	8.353(4.051-17.225)	<0.001	14.000(6.290-31.162)	<0.001

Infection at diagnosis or during induction therapy	13.072(1.780-95.993)	0.012	8.861(1.131-69.441)	0.038

Table 3: Predictors at diagnosis for all-cause mortality during follow-up in AAV patients (n = 198)

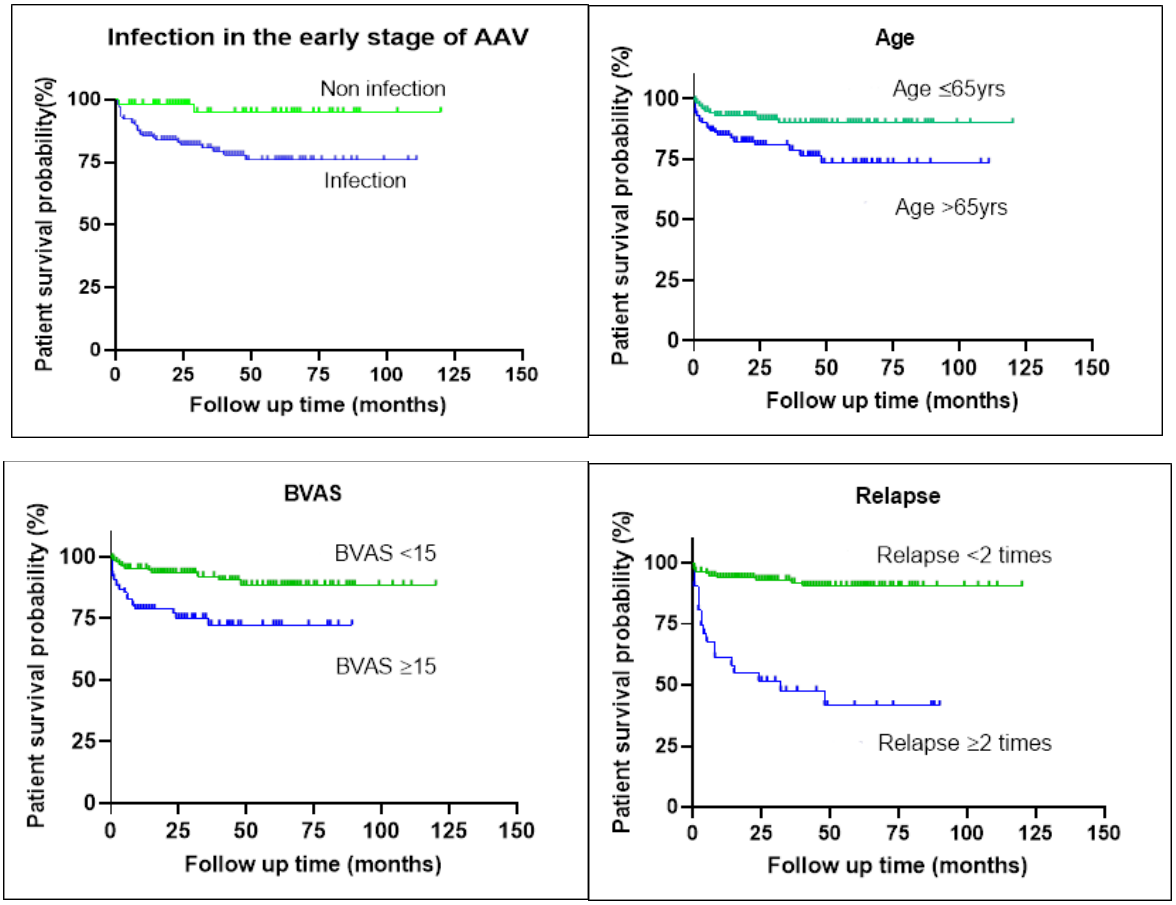

Figure 1: Survival by age group, BVAS at diagnosis, infection at diagnosis or during induction therapy, relapse at diagnosis

Characteristics	Results
Demographic characteristics	
Age, median (IQR), year	66 (59-72)
Female (%)	69 (49)
Clinical manifestations, n (%)	
Multiple system involved	111 (79)
Kidney	109 (78)
Lung	86 (61)
Muscle	23 (16)
ENT	22 (16)
Nervous system	16 (11)
Eyes	15 (10)
BVAS, ≥15	66 (47)
Laboratory data	
ESR, mm/h	68 (38-93)
CRP, mg/L	23 (5-69)
24h protein urine, g/24h	0.85 (0.47-1.92)
S-creatinine, μmol/l	156 (71-321)
MPO-ANCA+, n (%)	114 (81)
Sites and pathogens of infection	
Respiratory tract infection	86 (62)
Urinary tract infection	25 (18)
Gastrointestinal tract infections	6 (4)
Bacterium	91(65)
Fungus	37 (27)

Virus	28(20)

Table 4: Characteristics of 139 patients infected in the early stage of AAV

Variable	Univariable		Multivariable	
	OR(95%CI)	p value	OR(95%CI)	p value
Age, year	1.037(1.001-1.072)	0.038	1.030(0.979-1.083)	0.251
Female Sex	1.071(0.421-2.373)	0.835		
BVAS, ≥15	6.053(2.196-16.681)	＜0.001	4.324(1.017-18.387)	0.047
Multiple systemic involvement	0.781(0.294-2.073)	0.620		
24h protein urine, g/24h	1.199(0.760-1.892)	0.436		
S-creatinine	1.008(1.003-1.012)	＜0.001	1.007(1.001-1.013)	0.015
MPO-ANCA+ (ref. PR3-ANCA+)	0.172(0.045-0.658)	0.01	8.291(1.175-58.492)	0.034
Prednisone, ≥120mg/d	2.235(0.618-8.091)	0.220		
CTX (ref. RTX)	0.886(0.447-1.755)	0.728		
Prevention with SMZ	0.136(0.049-0.377)	<0.001	0.025(0.004-0.154)	<0.001

Table 5: Predictors for infection during inducing treatment in AAV patients (n = 84)

Discussion

This population-based study followed 198 AAV patients, analysing their courses of infection and death, and discovered that age (>65 years) and BVAS (≥15) were the common predictors to infection and death. Infection in the early stage and relapse(≥2times) independently contributed to mortality while high levels of S-creatinine, MPO-ANCA+ and lack of prevention with SMZ increased the incidence of severe infection. The findings is instrumental in making clinical treatment strategies on the purpose of reducing patients mortality and infection rates, ultimately serving better clinical remission of AAV patients.

In the previous prognostic analysis of AAV, few studies concerning on the survival and infection of the Oriental population. Firstly, higher BVAS score means severer condition of the AAV patients and some studies believed that it predicted the refractory and recurrence of MPA and GPA[16,17]. Which is confirmed in our study that patients with BVAS(≥15) were prone to infection and death. The refractory and recurrence of AAV result in mroe damage to organs and require a higher dose of immunosuppressants. However, BVAS score is not only designed for small-vessel vasculitis, but also includes other vasculitis such as Takayasu arteritis. Therefore, an improved grading system specifically designed for AAV based on BVAS may be more sensitive and accurate. Secondly, it was not surprising that increasing age was associated with growing probability of mortality. A South Korean cohort of the elderly prognosis study also defined age as a risk factor for prognosis[18]. The older is more likely to occur treatment complications and infection. Besides, Rennie L. Rhee's study [19] finded that the baseline creatinine level was a basic predictor of ESRD or death in a cohort of 554 patients. Severe renal dysfunction was identified a predictor of mortality in patients with AAV in other similar studies as well[20]. Although in our study, creatinine was not an independent risk factor in the multivariate cox analysis of prognosis (p=0.054), it affected the survival in univariate analysis. To be notice, the level of creatinine at diagnose is part of the BVAS grading, which may explained why creatinine is not an independent predictor while BVAS is.

Infection has always been a troublesome adverse events of AAV. The impact of infection on the outcome of other rheumatic disease and chronic kidney diseases has been identified in a number of studies. However, the researches paid attention to that of AAV are limited. Our study showed that infection in the early stage of AAV contributed to morality independently. Which may resulted from the increasing disease activity along with infection aggravates the injury of multiple organs. Meanwhile, the severe infection could get in the way of the promptly use of immunosuppressants. To further determine the susceptible population during induction therapy, werestricted analysis to thoseweren't infected at diagnosis. Itshowed that patients with poor renal function and high disease activity wereat higher risk of infection during induction therapy, which was in accordance with the prognosis analysis. It is also worth noticing that MPO-ANCA+ patients were more susceptible toinfection than PR3-ANCA+ patients, which was in accordance of thefindings of the research of Jens Rathmann et al[21]. Moreover, use of SMZ was a protective factor against infection. Hence, SMZ should be taken into consideration for patients with high risk of infection during induction therapy .Previous studies have demonstrated that high cumulative doses of CTX increased the risk of infection[22]. Hower, in our study, we did not observe significant difference between CTX and RTX recipients.In our cohort, the one-year survival rate was 88%. In a study from Massachusetts General Hospital, the leading cause of death in AAV patients was cardiovascular disease and infection[23]. Other Western studies have reached similar conclusions[24-26]. Intersetingly, in our cohort, interestingly, cardiovascular disease was not so significant as it was in Western researches. There differences may owing to different races, which highlighted further researches on Asian populationsto explain this difference.

Our study also has some limitations. Firstly, it is a single center study which may not be representative of all AAV patients, external validation is needed to confirm our findings. Secondly, detailed records of the dose of corticosteroids are not complete. Thus, the effect of the conticosteroids on prognosis and infection could not be analyzed. The are also some strengths in the study. Firstly, this study fills the gap in the Asian region of AAV patients of this kind of observation study. Then, by dividing the timespan into six months and later, it informs clinicians about different treatment priorities during induction therapy or maintenance therapy. During induction therapy, it is of great necessity to control infection while exerting potent immunosuppressants on patients to quickly contain the advancing of disease.

References

1. Watts R, Mahr A, Mohammad A, et al. Classification, epidemiology and clinical subgrouping of antineutrophil

cytoplasmic antibody (ANCA)-associated vasculitis[J]. Nephrology, dialysis, transplantation : official publication of the European Dialysis and Transplant Association - European Renal Association, 2015: i14-22.
2. Jennette J C, Falk R J, Bacon P A, et al. 2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides[J]. Arthritis and Rheumatism, 2013, 65(1): 1-11.
3. Fauci A S, Katz P, Haynes B F, et al. CYCLOPHOSPHAMIDE THERAPY OF SEVERE SYSTEMIC NECROTIZING VASCULITIS[J]. New England Journal of Medicine, 1979, 301(5): 235-238.
4. Stone J H, Merkel P A, Spiera R, et al. Rituximab versus Cyclophosphamide for ANCA-Associated Vasculitis[J]. New England Journal of Medicine, 2010, 363(3): 221-232.
5. Little M A, Nightingale P, Verburgh C A, et al. Early mortality in systemic vasculitis: relative contribution of adverse events and active vasculitis[J]. Annals of the Rheumatic Diseases, 2010, 69(6): 1036-1043.
6. Holle J U, Gross W L, Latza U, et al. Improved Outcome in 445 Patients With Wegener's Granulomatosis in a German Vasculitis Center Over Four Decades[J]. Arthritis and Rheumatism, 2011, 63(1): 257-266.
7. Cartin-Ceba R, Golbin J M, Keogh K A, et al. Rituximab for remission induction and maintenance in refractory granulomatosis with polyangiitis (Wegener's) Ten-year experience at a single center[J]. Arthritis and Rheumatism, 2012, 64(11): 3770-3778.
8. Tan J, Dehghan N, Chen W, et al. Mortality in ANCA-associated vasculitis: ameta-analysis of observational studies[J]. Annals of the rheumatic diseases, 2017, 76(9): 1566-1574.
9. Yamagata K, Usui J, Saito C, et al. ANCA-associated systemic vasculitis in Japan: clinical features and prognostic changes[J]. Clinical and Experimental Nephrology, 2012, 16(4): 580-588.
10. Bourgarit A, Le Toumelin P, Pagnoux C, et al. Deaths occurring during the first year after treatment onset for polyarteritis nodosa, microscopic polyangiitis, and Churg-Strauss syndrome - A retrospective analysis of causes and factors predictive of mortality based on 595 patients[J]. Medicine, 2005, 84(5): 323-330.
11. Peh C, Jones R, Walsh M, et al. RITUXIMAB VERSUS CYCLOPHOSPHAMIDE IN ANCA ASSOCIATED RENAL VASCULITIS[J]. Nephrology, 2010, 15: 49-49.
12. Flossmann O, Berden A, De Groot K, et al. Long-term patient survival in ANCA-associated vasculitis[J]. Annals of the Rheumatic Diseases, 2011, 70(3): 488-494.
13. Jennette J C, Falk R J. Small-vessel vasculitis[J]. New England Journal of Medicine, 1997, 337(21): 1512-1523.
14. Luqmani R A, Bacon P A, Moots R J, et al. BIRMINGHAM VASCULITIS ACTIVITY SCORE (BVAS) IN SYSTEMIC NECROTIZING VASCULITIS[J]. Quarterly Journal of Medicine, 1994, 87(11): 671-678.
15. Smitten A L, Choi H K, Hochberg M C, et al. The risk of hospitalized infection in patients with rheumatoid arthritis[J]. Journal of Rheumatology, 2008, 35(3): 387-393.
16. Oh Y, Ahn S, Park E, et al. Chest and renal involvements, Birmingham vascular activity score more than 13.5 and five factor score (1996) more than 1 at diagnosis are significant predictors of relapse of microscopic polyangiitis[J]. Clinical and experimental rheumatology, 2017(1): 47-54.
17. Yoo J, Kim H, Jung S, et al. Birmingham vasculitis activity score of more than 9.5 at diagnosis is an independent predictor of refractory disease in granulomatosis with polyangiitis[J]. International journal of rheumatic diseases, 2017, 20(10): 1593-1605.
18. Mcgovern D, Williams S, Parsons K, et al. Long-term outcomes in elderly patients with ANCA-associated vasculitis[J]. Rheumatology (Oxford, England), 2020, 59(5): 1076-1083.
19. Rhee R, Hogan S, Poulton C, et al. Trends in Long-Term Outcomes Among Patients With Antineutrophil Cytoplasmic Antibody-Associated Vasculitis With Renal Disease[J]. Arthritis & rheumatology (Hoboken, N.J.), 2016, 68(7): 1711-20.
20. De Joode A a E, Sanders J S F, Stegeman C A. Renal Survival in Proteinase 3 and Myeloperoxidase ANCA-Associated Systemic Vasculitis[J]. Clinical Journal of the American Society of Nephrology, 2013, 8(10): 1709-1717.
21. Rathmann J, Jayne D, Segelmark M, et al. Incidence and predictors of severe infections in ANCA-associated vasculitis: a population-based cohort study[J]. Rheumatology (Oxford, England), 2020.
22. De Groot K, Harper L, Jayne D R W, et al. Pulse Versus Daily Oral Cyclophosphamide for Induction of Remission in Antineutrophil Cytoplasmic Antibody-Associated Vasculitis A Randomized Trial[J]. Annals of Internal Medicine, 2009, 150(10): 670-U3.
23. Wallace Z, Fu X, Harkness T, et al. All-cause and cause-specific mortality in ANCA-associated vasculitis: overall and according to ANCA type[J]. Rheumatology (Oxford, England), 2020, 59(9): 2308-2315.
24. Luqmani R, Suppiah R, Edwards C, et al. Mortality in Wegener's granulomatosis: a bimodal pattern[J]. Rheumatology (Oxford, England), 2011, 50(4): 697-702.
25. Tan J, Choi H, Xie H, et al. All-Cause and Cause-Specific Mortality in Patients With Granulomatosis With Polyangiitis: A Population-Based Study[J]. Arthritis care & research, 2019, 71(1): 155-163.
26. Jardel S, Puechal X, Le Quellec A, et al. Mortality in systemic necrotizing vasculitides: A retrospective analysis of the French Vasculitis Study Group registry[J]. Autoimmunity Reviews, 2018, 17(7): 653-659.

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