P2.118 CENTRAL NERVOUS SYSTEM INVOLVEMENT IN PATIENTS WITH GRANULOMATOSIS WITH POLYANGIITIS: A SINGLE CENTER EXPERIENCE

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Objectives: Nerve involvement in patients with granulomatosis with polyangiitis (GPA) is encountered relatively frequently. However, central nervous system (CNS) manifestations, are reported to occur in about 10% of them. We aimed to estimate the prevalence of CNS involvement among patients with GPA, describe the related clinical characteristics, and compare patients with and without CNS involvement, in terms of the vasculitic manifestations and long term outcomes.

Methods: The medical charts of all patients with ANCA-associated and biopsy proven small vessel vasculitis (AAV), diagnosed in our hospital between 1985-2015, were reviewed retrospectively and patients with GPA and CNS involvement were identified.
Demographics, serological and clinical features at the time of AAV diagnosis and during the follow up time were recorded. Comparisons of disease characteristics and outcomes, were performed between GPA patients with and without CNS involvement.

Results: There were 77 patients with GPA in our AAV registry. Of these, 9 (11.7%), 7 men/2 women, all c/PR3-ANCA-positive developed CNS manifestations, either at clinical presentation (33.3%) or during the follow up time (66.7%). At the time of CNS diagnosis, their median (range) age was 46 (26-73) years old. They have been subsequently followed up for 14 (0-121) months and in total for 60 (13-144) months. GPA patients with CNS involvement, compared with those without, had ENT involvement more frequently (77.8% vs. 25.4%, p = 0.004) at initial AAV diagnosis, while throughout the disease course lung vasculitis was less frequent in this group (44.4% vs. 79.4%, p = 0.02) (Table-1). Furthermore, comparison of GPA patients with CNS involvement, to those without, did not reveal any difference in terms of long-term outcomes, i.e patient survival, relapse rate and treatment-related adverse events (Table-1).

Conclusions: CNS involvement was recorded in 11.7% of our GPA patients, either during the initial phase of the disease, or later in sequelae. According to our results, CNS involvement was more common amongst patients who had ENT involvement at initial diagnosis and did not affect the long term outcomes of these patients.

P2.120 FACTORS ASSOCIATED WITH DEVELOPMENT OF TRACHEOBRONCHIAL STENOSSES (TBS) IN GRANULOMATOSIS WITH POLYANGIITIS (GPA)
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TBS in GPA are severe as they impose high morbidity and are potentially lethal. They can occur even when disease is in remission. Factors associated with their development have not been identified. Objective: To identify factors associated with TBS development in GPA.

Methods: Retrospective study of patients with (n=29) and without TBS (n=76) within our GPA cohort of 155 patients in a respiratory centre, classified according to EMA algorithm.

Results: We have separately described the characteristics of those 29 patients with TBS. In all of them, TBS were diagnosed bronchoscopically. Patients who only had subglottic stenoses were not included in this study.

Statistical analysis: Comparisons between those who developed and did not develop TBS by X2 with Yates correction or two-tailed exact Fisher test.

Results: Of the total 108 patients analysed, 69 had limited and 39 generalised disease. Since 2009 we have identified 29 patients with TBS. Factors identified in those who developed TBS are expressed as the variable, percentage and significance. Those positively associated with TBS presence were: general symptoms-72.4%-p < 0.001; rhinosinusal involvement-89.7%-p < 0.009; musculoskeletal symptoms-55.2%-p = 0.006. At time of TBS diagnosis, patients had a higher VDI (mean 15 vs 6; p < 0.006) and higher CVI (mean 42 vs 14; p < 0.025). Other factors negatively associated with TBS were: PDN dose > 10 mg qd-93.7% in those without TBS-p < 0.009 and history of azathioprine intake-30.4%-p = 0.025. Interestingly, extent of disease, either generalised or limited was not associated with TBS development, nor were ANCA levels.

Discussion: We have aimed to initially identify some factors associated with TBS development, especially as this complication can arise late in the disease course, and not be associated with other clinical manifestations of the disease. Although not associated with the classic definition of limited disease, those with more general and rhinosinusal symptoms seem to be more prone to this complication, while sustained prednisone use seem to avoid it. However, this is not symptoms related to TBS. Females were predominantly affected (76%). Mean ± SD age was 42 ± 12 years. Most patients had limited GPA (n=20, 69%). Main TBS symptoms: dysphonia (n=25, 86%), stridor and dyspnoea (n=23 each; 79%). All had trachitional involvement and 12 (41%) additional bronchial stenosis. Other accompanying manifestations by organ/system: rhinosinusal (n=26;87%), musculoskeletal (n=16;53%), ocular (n=13;45%), pulmonary (n=12;41%), renal (n=8;27%), mucocutaneous (n=5;17%), neurological (n=4;13%). In 16 patients, biopsies were performed, acute and/or chronic inflammation was reported in 14 (48%), vasculitis in 2 (7%), granulomatous inflammation in 1 (3.5%). 59% were PR3-ANCA and C-ANCA positive at TBS diagnosis; data on the remaining percentage were either negative or not performed at that time. Treatment: 58% had simultaneous medical (n=8; 27%) with glucocorticoid therapy; n=9; 31% with immunosuppressants) and surgical therapy, while the rest only the latter at time of TBS diagnosis. Associated comorbidities were: arterial hypertension in 7 (24%), diabetes mellitus type 2 in 3 (10%) and 2 patients already had ESRD at time of TBS development. Frequent relapses were present: 18 (82%) had one, 11 (38%) two, 9 (31%) three and 2 (7%) > 4 relapses.

Conclusions: TBS are serious and can develop with GPA in remission. These features are common to all reported series, but factors which identify its development are still to be known. Timely diagnosis, and optimal treatment and follow-up remain as unmet needs.

P2.119 TRACHEOBRONCHIAL STENOSES (TBS) IN GRANULOMATOSIS WITH POLYANGIITIS (GPA)
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Tracheobronchial stenoses (TBS) occur in GPA between 10-20% of cases. In our centre, it is vital to know its characteristics due to its scope. Objective: to describe the frequency, characteristics and treatment of TBS in GPA at our institution.

Methods: retrospective study of all identified TBS cases in whom their demographics, clinical and paraclinical features, and treatment were analysed. Statistical analysis: descriptive statistics.

Results: we identified 29 TBS in 155 (19%) GPA patients (classified according to the EMA algorithm). Twenty cases sought attention due to symptoms related to TBS. Females were predominantly affected (76%). Mean ± SD age was 42 ± 12 years. Most patients had limited GPA (n=20, 69%). Main TBS symptoms: dysphonia (n=25, 86%), stridor and dyspnoea (n=23 each; 79%). All had tracheobronchial involvement and 12 (41%) additional bronchial stenosis. Other accompanying manifestations by organ/system: rhinosinusitis (n=26;87%), musculoskeletal (n=16;53%), ocular (n=13;45%), pulmonary (n=12;41%), renal (n=8;27%), mucocutaneous (n=5;17%), neurological (n=4;13%). In 16 patients, biopsies were performed, acute and/or chronic inflammation was reported in 14 (48%), vasculitis in 2 (7%), granulomatous inflammation in 1 (3.5%). 59% were PR3-ANCA and C-ANCA positive at TBS diagnosis; data on the remaining percentage were either negative or not performed at that time. Treatment: 58% had simultaneous medical (n=8; 27%) with glucocorticoid therapy; n=9; 31% with immunosuppressants) and surgical therapy, while the rest only the latter at time of TBS diagnosis. Associated comorbidities were: arterial hypertension in 7 (24%), diabetes mellitus type 2 in 3 (10%) and 2 patients already had ESRD at time of TBS development. Frequent relapses were present: 18 (82%) had one, 11 (38%) two, 9 (31%) three and 2 (7%) > 4 relapses.

Conclusions: TBS are serious and can develop with GPA in remission. These features are common to all reported series, but factors which identify its development are still to be known. Timely diagnosis, and optimal treatment and follow-up remain as unmet needs.
unsatisfactory as long-standing glucocorticoid use can also lead to added comorbidity. Currently, there are no consensus on how to treat and timely diagnose TBS, and this constitutes an unmet need.

Methods: We carried out a retrospective analysis of 218 patients GPA, treated in rheumatology department from 2010 till 2014 to evaluate frequency of ophthalmic manifestations and their relations to disease variant. Women were affected twice as often as men (143/75). The age of patients ranged from 16 to 86 with the mean age 51 years old.

Results: Upper airways were affected in 94.0% of cases (205/218), lungs in 62.4% (136/218) and kidneys in 48.6% (106/218). Local variant was diagnosed in 27.5% (60/218) and systemic (including early systemic and generalized) in 72.5% (158/218). Ophthalmic manifestations were present in 48.2% of patients (105/218). Eye (i.e. globe and adnexa) was affected slightly more often in localized GPA, but this is not statistically reliable. The most common ophthalmic manifestations were orbital masses - 22.9% (50/218) and episcleritis - 14.7% (32/218), frequency of other ophthalmic manifestations is described in Table. In further analysis we included two groups of patients: those with orbital masses (n = 50) and those with globe involvement (n = 51). Within each group patients were divided depending on variant of GPA. Orbital masses were more common in localized disease (40% vs. 16%, p < 0.001) and globe involvement (episcleritis, scleritis, keratitis, uveitis, retinal damage) was associated with systemic variant (12% vs. 27%, p<0.05). Table. Ocular manifestations in GPA.

Conclusions: The eye is the common target organ in GPA, affected in 48.2% of cases. Ophthalmic manifestations are variable with orbit and episclera being involved more frequently. Statistical analysis showed that orbital damage is more common in localized variant and globe affection is more common in patients with systemic involvement.

Conflicts of Interest: None.

\[\text{Table. Ocular manifestations in GPA.}\]

<table>
<thead>
<tr>
<th>Manifestation</th>
<th>Prevalence in localized GPA (n=50)</th>
<th>Prevalence in systemic GPA (n=158)</th>
<th>All (n=218)</th>
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</thead>
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<tr>
<td>Orbital masses</td>
<td>40%</td>
<td>16.4%</td>
<td>22.9%</td>
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<tr>
<td>Medial orbital wall destruction</td>
<td>8.3%</td>
<td>5.7%</td>
<td>6.4%</td>
</tr>
<tr>
<td>Dacryoanadentis</td>
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<td>5.0%</td>
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<td>Dacryocystitis</td>
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<td>5.7%</td>
<td>6.0%</td>
</tr>
<tr>
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<td>2.3%</td>
</tr>
<tr>
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<td>15.8%</td>
<td>14.7%</td>
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<td>Necrotizing scleritis</td>
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<td>Keratitis</td>
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<td>5.0%</td>
</tr>
<tr>
<td>Optic neuropathy</td>
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<td>4.1%</td>
</tr>
<tr>
<td>Uveitis</td>
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<td>Retinal damage</td>
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<td>3.2%</td>
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</table>

P2.12 CERABLYRULAR MANIFESTATIONS IN GPA: ASSOCIATION WITH DISEASE VARIANTS

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Objective: To study prevalence of ophthalmic manifestations and its relationship to the GPA variant.

Methods: We identified 30 patients with orbital inflammatory masses associated with autoimmune diseases including granulomatosis with polyangiitis GPA, formerly Wegener’s granulomatosis, eosinophilic granulomatosis with polyangiitis eGPA, Immunoglobulin G4 related disease IgG4RD. Clinical and laboratory data was collected from electronic clinical records. Comprehensive diagnostic criteria were used for IgG4RD and Chapel Hill criteria for GPA and eGPA. Statistical analysis was performed by GraphPad software. Continuous variables were compared using non-parametric Mann-Whitney test and categorical variables were compared by Fisher exact test.

Results: Reference Table. The median age was 44 years (range 29-76). 14 patients were diagnosed with GPA, 11 patients had IgG4RD, 1 patient with IgG4 lymphoma, 1 with eGPA, 1 unspecified vasculitis, 1 IgA dacroanadentis, 1 non-specific granuloma. 7/11 patients with IgG4-RD had isolated orbital masses whereas all 14 GPA patients suffered extra-ocular manifestations (p < 0.01), usually sino-nasal or pulmonary disease. IgG4 levels were elevated pre-treatment in IgG4 RD patients (median 2.46 g/l range 1.2-23.7) and dropped to 1.25 g/l (range 0.37-10.4) after therapy (p < 0.05). Immunoglobulin subclass levels were not checked routinely in GPA. All 11 patients with IgG4-RD underwent diagnostic orbital biopsy vs 3/14 GPA (p = 0.0001). All 30 patients were treated with corticosteroids (used alone in 3/11 IgG4-RD patients).

P2.123 FEATURES OF ORBITAL INFLAMMATORY DISEASE AND RESPONSE TO THERAPY

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Objective: To characterize a single centre retrospective case series of patients with orbital inflammatory masses associated with autoimmune diseases including granulomatosis with polyangiitis GPA, formerly Wegener’s granulomatosis, eosinophilic granulomatosis with polyangiitis eGPA, Immunoglobulin G4 related disease IgG4RD.

Methods: We identified 30 patients with orbital inflammatory masses on MRI imaging. Clinical and laboratory data was collected from electronic clinical records. Comprehensive diagnostic criteria were used for IgG4RD and Chapel Hill criteria for GPA and eGPA. Statistical analysis was performed by GraphPad software. Continuous variables were compared between IgG4RD and GPA groups using non-parametric Mann-Whitney test and categorical variables were compared by Fisher exact test.

Results: Reference Table. The median age was 44 years (range 29-76). 14 patients were diagnosed with GPA, 11 patients had IgG4RD, 1 patient with IgG4 lymphoma, 1 with eGPA, 1 unspecified vasculitis, 1 IgA dacroanadentis, 1 non-specific granuloma. 7/11 patients with IgG4-RD had isolated orbital masses whereas all 14 GPA patients suffered extra-ocular manifestations (p < 0.01), usually sino-nasal or pulmonary disease. IgG4 levels were elevated pre-treatment in IgG4 RD patients (median 2.46 g/l range 1.2-23.7) and dropped to 1.25 g/l (range 0.37-10.4) after therapy (p < 0.05). Immunoglobulin subclass levels were not checked routinely in GPA. All 11 patients with IgG4-RD underwent diagnostic orbital biopsy vs 3/14 GPA (p = 0.0001). All 30 patients were treated with corticosteroids (used alone in 3/11 IgG4-RD patients).
ANCA ASSOCIATED VASCULITIS REMEDIABLE WITH MILD IMMUNOSUPPRESSIVE THERAPY

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Objectives: A new disease concept, otitis media with ANCA-associated vasculitis (AAV), has recently been advocated. We clarify the clinical features of otitis media with AAV (OMAAV).

Methods: One hundred and eighty two patients (85 males and 97 females) with AAV (93 microscopic polyangiitis, 63 granulomatosis with polyangiitis (GPA), and 26 eosinophilic GPA) were admitted to Niigata University Hospital from 1989 through 2016 October. Thirty two patients were diagnosed as having definite or probable GPA and in E only group were 110 ml and 43.2 U/ml, respectively (p = 0.04). Azathioprine was chosen in 46% of GPA group and 13% of E only group (p < 0.01). Relapse was observed in 54% of GPA group and 13% of E only group (p < 0.05).

Conclusions: The prevalence of OMAAV was increasing. Although cyclophosphamide is a standard immunosuppressant for GPA, azathioprine might be enough for E only OMAAV.

P2.125 CUTANEOUS MANIFESTATIONS OF ANCA-ASSOCIATED VASCULITIS

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Cutaneous Manifestations of ANCA-Associated Vasculitis:

Micheletti RG, Chiesa Fuxench ZC, Craven A, Watts RA, Lugman RA, Merkel PA for the Diagnostic and Classification Criteria in Vasculitis Study (DCVAS) Investigators.

Title: Cutaneous manifestations of anti-neutrophil cytoplasmic antibody-associated vasculitis.

Background/Purpose: The cutaneous manifestations of anti-neutrophil cytoplasmic antibody-associated vasculitis (AAV), including granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA), are varied and have not been well characterized. This study aimed to describe the spectrum and extent of dermatologic features of AAV.

Methods: A large, international, collaborative effort to collect comprehensive clinical data on a large cohort of patients with vasculitis with a goal of developing new classification and diagnostic criteria.

Results: Data from 1274 patients with AAV from 130 centers worldwide were available for this study: 702 (55%) with GPA, 331 (26%) with MPA, and 241 (19%) with EGPA. Cutaneous findings were common in patients with AAV (Table 1), affecting 239 (34%) patients with GPA, 97 (29%) patients with MPA, and 113 (47%) patients with EGPA. The most frequent cutaneous manifestations in each type of AAV were as follows: GPA: petechiae or purpura (N = 113; 16%), painful skin lesions of any type (N = 66; 9.4%), and maculopapular rash (N = 47; 6.7%); MPA: petechiae or purpura (N = 33; 10%), livedo reticularis or racemosa (N = 25; 7.6%), and maculopapular rash (N = 20; 6.0%); and EGPA: petechiae or purpura (N = 50; 21%), maculopapular rash (N = 36; 15%), pruritus (N = 30; 13%), and urticaria (N = 19; 7.9%). Cutaneous findings were also analyzed by ANCA type (PR3, MPO, and ANCA-negative), as shown in Table 1.

Conclusions: This is the largest study of cutaneous manifestations of AAV ever conducted. Utilizing data collected comprehensively via a standard protocol, it demonstrates that skin lesions are quite common and varied in GPA, MPA, and EGPA. Ongoing analyses will focus on examining the association of specific cutaneous manifestations of AAV with other organ system involvement and laboratory findings.

P2.124 INCREASING PREVALENCE OF OTITIS MEDIA WITH ANCA ASSOCIATED VASCULITIS REMEDIABLE WITH MILD IMMUNOSUPPRESSIVE THERAPY

Takehiko Nakatsu1, Yukiko Nozawa1, Hiroe Sato2, Yoko Wada1, Takeshi Kuroda3, Masashi Nakano1 and Ichiel Nair1
1Division of Clinical Nephrology and Rheumatology, Graduate School of Medical and Dental Sciences, Niigata University Niigata, Japan, 2Division of Medical Technology, School of Health Sciences, Faculty of Medicine, Niigata University Niigata, Japan

Objectives: The prevalence of OMAAV was increasing. Although cyclophosphamide is a standard immunosuppressant for GPA, azathioprine might be enough for E only OMAAV.
P2. 125 ONE THIRD OF EGPA PATIENTS HAVE INFLAMMATORY HEART DISEASE

Eli García1, Len Harty1 and David Jayne1
Vasculitis/Manchester Metropolitan University Manchester/UK

Objective: To establish the prevalence of EGPA inflammatory heart disease and develop an algorithm for heart disease screening in EGPA patients.

Methods: A single centre audit of tests performed on all EGPA patients was undertaken. No parametric statistics are given as percentages and median (IQR).

Results: 96 (73%) of 131 EGPA patients (47% men) underwent cardiac evaluation. Median age was 50 years (38-58), 37% were ANCA-positive and asthma preceded diagnosis by median 97 months (36-240). 41 (43%) were symptomatic for heart disease: dyspnoea (47%), chest pain (29%), limb oedema (24%), palpitations (13%), syncope (4%), abdominal discomfort (2%) and shock (2%). 27/96 (28%) patients had inflammatory heart disease, present in 20 at EGPA diagnosis, 5 upon EGPA relapse and preceded EGPA diagnosis in two. 59% (24) of those who were symptomatic and 5% (3) of those who were asymptomatic for heart disease had inflammatory heart disease. 15% patients had myocarditis, 6% pericarditis, 5% myopericarditis and 1 coronary vasospasm. One patient with pericarditis also had periarthritis. Cardiac abnormalities of any sort were found in 52% patients. Patients who had inflammatory EGPA heart disease were younger (46 [28 - 52] V 50 [41 - 59]; p = 0.005) compared to inactive AAV-patients and healthy controls (p = 0.001). Patients with inflammatory EGPA heart disease have more aggressive systemic disease with higher serum and clinical markers of inflammation. EGPA patients should have ECG, echocardiography and troponin. 60% of those symptomatic for heart disease and 5% of those asymptomatic being affected. EGPA patients with inflammatory heart disease have more aggressive systemic disease with higher serum and clinical markers of inflammation.

Conclusions: 27% EGPA patients have inflammatory heart disease with 60% of those symptomatic for heart disease and 5% of those asymptomatic being affected. EGPA patients with inflammatory heart disease have more aggressive systemic disease with higher serum and clinical markers of inflammation. EGPA patients should have ECG, echocardiography and troponin in patients with suspicion for cardiac disease.

P2. 127 GASTROINTESTINAL INVOLVEMENT IN 6% OF 216 CONSECUTIVE PATIENTS WITH ANTI-NEUTROPHIL CYTOPLASMIC ANTIBODY (ANCA) ASSOCIATED VASCUITIS

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Objectives: The ANCA-associated vasculitides granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA) involve several organs sometimes including the gastrointestinal (GI) tract. The aim was to study GI involvement in a large series of patients.

Methods: All GPA and MPA patients from a restricted area were prospectively registered since 1997. Patients diagnosed before 1997 were also investigated. GI manifestations of vasculitis were defined as proposed by Pagnoux in 2005. Data were retrieved from the patient charts.

Results: Fourteen (6%) of 216 patients had GI involvement starting after diagnosis of vasculitis in 8/14 patients, most commonly abdominal pain and GI bleeding. Radiology was important for detection of GI disease. Gastroscopy showed red and swollen mucosa in 3/5 where histology confirmed vasculitis in 2 patients. Colonoscopy was uncharacteristic in 1/3 patients but histology showed vasculitis. Because of perforation, 5 individuals were operated with hemicolecotomy or small intestinal resection. Primary anastomosis was created in 2/5 and enterostomy in 3/5 patients. One patient had a hemicolecotomy because of lower GI bleeding. One sigmoid abscess was treated with drainage and antibiotics. One intra-abdominal bleeding was treated with arterial coiling of the inferior mesenteric artery. Conservative therapy was sufficient in 6 patients. Two patients died because of GI vasculitis, but patient survival was similar in the two groups.

Conclusions: In this population-based study, GI involvement of ANCA-associated vasculitides was found in 6% of 216 patients. Patients only need surgery in case of GI perforation or severe bleeding. A multidisciplinary approach of these patients is strongly recommended.

P2. 128 VENOUS THROMBOEMBOLISM (VTE) IN PATIENTS WITH ANTI-NEUTROPHIL CYTOPLASMIC ANTIBODY (ANCA)-ASSOCIATED VASCULITIS - UNDERLYING PROTHROMBOTIC CONDITION IN ACTIVE DISEASE?

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1Unit of Rheumatology, Department of Medicine, Karolinska Institutet Stockholm, Sweden, 2Department of Renal Medicine, CLINTEC, Karolinska Institutet Stockholm, Sweden

Objectives: Patients with anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides (AAVs) entail an increased incidence of venous thromboembolism (VTE) compared to the general population.

Aims: To investigate the influence of the ANCA-specificity, disease phenotype and disease duration as well as classical VTE risk factors on the development of VTE in AAV-patients.

Methods: This was a retrospective cohort study using patient data from a database on AAV-patients at the Karolinska University Hospital, Stockholm, Sweden. 187 ANCA-positive patients with granulomatosis with polyangiitis (GPA) or microscopic polyangiitis (MPA) diagnosed between 2005 and 2014 were included and the occurrence of VTE was evaluated from 3 months before AAV diagnosis until end of follow-up. Plasma samples were obtained at the time of diagnosis and/or active flare from AAV-patients (n = 19) sex- and age-matched AAV-patients during inactive period of the disease (n = 15) and healthy controls (n = 15). We assessed haemostasis in a pilot study using two global haemostatic methods: endogenous thrombin potential (ETP) and overall fibrinolytic potential (OPF) in these three groups.

Results: 28 VTEs occurred in 24 patients over a total follow-up time of 1020 person-years. A total incidence rate of 2.74 cases of VTE per 100 person-years. A majority of the VTEs occurred in close temporal proximity to the AAV-diagnosis, with more than half of the patients having a VTE within the first year after the AAV-diagnosis (Figure 1). ANCA-specificity was not significantly associated with VTE development, nor were AAV-diagnosis (GPA/MPA), sex or renal involvement. High age (p < 0.01) and previous VTE (p < 0.05) were significantly more common in the VTE group. ETP was significantly increased and OPF significantly decreased in plasma from active compared to inactive AAV-patients and healthy controls (p < 0.05, p < 0.01, respectively).

Conclusions: The main finding of this study is the striking prevalence of VTE in AAV-patients within the first year after the AAV-diagnosis. High age at AAV-diagnosis and previous VTE should be taken into account when estimating VTE-risk. The results of the pilot study indicate disturbances in the haemostatic balance towards prothrombotic condition in active AAV-patients, where ETP and OPF may be useful markers for identifying patients at high risk.
lead to a higher incidence of erosive and ulcerative lesions and thrombotic or bleeding complications.

Conflicts of Interest: None.

<table>
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<tr>
<th>Condition</th>
<th>Control Group (n=130)</th>
<th>GPA Group (n=165)</th>
<th>P-value</th>
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<td>Upper respiratory tract involvement</td>
<td>24 (75.9%)</td>
<td>29 (79.2%)</td>
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<tr>
<td>Lung disease</td>
<td>24 (75.9%)</td>
<td>27 (70.1%)</td>
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<td>Hemorrhagic alveolitis</td>
<td>8 (23.8%)</td>
<td>6 (16%)</td>
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<td>Kidney disease</td>
<td>19 (58.8%)</td>
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<td>15 (45.5%)</td>
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<td>Vein thrombosis</td>
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<td>Mortality</td>
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P2.130 MALIGNANT LYMPHOMA IN GRANULOMATOSIS WITH POLYANGIITIS

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Objective: Our aim was to assess the clinical characteristics and treatment of patients with GPA complicated by lymphoma and to describe the lymphoma subtypes, sites, prognosis and possible association with Epstein-Barr virus in a population-based setting.

Method: From the Swedish population based patient register all individuals with the diagnosis of GPA between 1964 and 2012 were identified (n=3,224). Through linkage with the Swedish cancer register all lymphoproliferative malignancies (ICD7:200-202) registered after the first discharge listing GPA were identified. Through medical records, the GPA diagnosis was evaluated using the EMEA Consensus Algorithm for Classification of Vasculitis. All diagnostic biopsies were retrieved and classified according to the latest WHO classification. The presence of EBV in lymphoma tissue was analyzed using EBV-encoded RNA (EBER) in situ hybridization. Clinical data of both GPA and lymphoma were collected from the medical files.

Results: 23 GPA-patients with malignant lymphoma were identified. Clinical characteristics of the study patients at the time of lymphoma diagnosis, characteristics of the lymphoma and survival are shown in table 1. The majority had severe and generalized GPA disease. Most (78%) had been treated with cyclophosphamide for their GPA, the median cumulative dose was 42g (1.9-511). The majority (83%) of the lymphomas were of B-cell origin, the single most common subtype was diffuse large B-cell lymphoma (DLBCL). Only one of the lymphomas was localized to the ear, nose and throat (ENT) area. EBV-positivity in lymphoma tissue was detected in only 1 of 18 examined cases. The majority of the lymphomas were aggressive, the median survival time after lymphoma diagnosis was only 4 months.

Discussion: We report that the lymphoma patients mainly had severe and relapsing GPA disease treated with potent immunosuppressive drugs during a prolonged period of time, the distribution of lymphoma subtypes and the engaged sites were broadly in line with what is seen in the general population. EBV did not seem to have an important role in the lymphomagenesis in GPA patients. The prognosis for these patients was markedly poor, even with modern treatment strategies, and the poor prognosis seemed associated with high cumulative CYC doses given for GPA.