AB0686

ARTERIAL ANEURYSMS IN BEHCET'S DISEASE: A RETROSPECTIVE DESCRIPTIVE ANALYSIS AND LONG-TERM OUTCOME

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Background: Behcet's Disease (BD) was described in 1937 after Hulusi Behcet; as a triad of recurrent aphthous and genital ulcers together with iridocyclitis.^{1,2} Arterial disease is seen in 3%–5% of the patients.^{3,4} Thrombosis and/or aneurysmal formation are common sequelae mainly false aneurysms.⁵ In spite of the fact that BD is not uncommon in the clinical practice in Egypt; the paucity of data that looked specifically to the arterial aneurysms in the Egyptian patients with BD had prompted the present study.

Objectives: Arterial aneurysm in Behcet's disease is a rare manifestation of Behcet's disease. The presence of Arterial aneurysm in Behcet's disease change the course of disease and it is management remains a challenge for rheumatologists.

Methods: A retrospective revision of files of 160 patients admitted and followed up in the rheumatology department, Cairo University Hospitals between 2004–2017 was done. We looked specifically for the prevalence of arterial aneurysms. Demographic characteristics of patients with aneurysms, clinical presentation, and associated clinical features.

Results: Twenty-seven (16.8%) patients had arterial aneurysms. All of them were males and the onset of development of the aneurysm is usually under the age of 40 years, 74% of the patients developed aneurysm 3.2±3.0 years after their disease onset. Apart from the oro-genital ulcerations; deep venous thrombosis was the most common associated manifestation. Pulmonary artery was the most common artery involved in 12 (44.4%) of patients, followed by the abdominal aorta in 4 (14.8%). Surgical intervention was done for 11 (40.7%) patients; all of them received cyclophosphamide pulses before surgery except one. Four (14.8%) patients in this study died.

Conclusions: Arterial aneurysms are common in Egyptian patients with BD. The profile of Egyptian BD patient that is susceptible for development of arterial aneurysm is a male patient, under the age of 40 years, smoker with relatively short disease duration.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.3058

AB0687

DOES ANTI-GLOMERULAR BASEMENT MEMBRANE (ANTI-GBM) ANTIBODY POSITIVITY CORRELATE WITH RELAPSE IN PATIENTS WITH ANTI-GBM DISEASE?

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Background: Anti-GBM disease is characterised by rapidly progressive glomerular nephritis with or without pulmonary haemorrhage. It is usually monophasic in nature and disease severity correlates with antibody titer. The disease is mediated by pathogenic antibodies directed against the non-collagenous region of the $\alpha 3$ chain of type IV collagen.

Despite the known pathogenicity of anti-GBM antibodies, and the correlation of disease severity with their titers, there is conflicting reports on whether anti-GBM antibody positivity correlates with disease relapse on long term follow up.

Objectives: To assess for correlation of anti-GBM antibody positivity and disease relapse in patients with anti-GBM disease.

Methods: Patients seen in one single academic centre between 1997 and 2017 were initially screened for the presence of anti-GBM disease by ICD 9/10 code for anti-GBM disease or Goodpasture's syndrome. 435 patients were identified. Patients were then included in the study if the diagnosis was confirmed by a board certified rheumatologist or nephrologist at our institution and had positive anti-GBM antibodies and/or biopsy results consistent with a diagnosis of anti-GBM

disease. The primary endpoint of this study was anti-GBM antibody positivity at the time of relapse. All charts were reviewed for baseline demographics, clinical manifestations, anti-GBM antibody and anti-neutrophil cytoplasmic antibody (ANCA) positivity at the time of initial presentation; these were compared between those with relapsing and non-relapsing disease. Results were analysed using a two tailed standard t-test. These same characteristics were also examined the relapsing cohort at the time of relapse.

Results: 40 patients were confirmed as having anti-GBM disease at our institution. Mean follow up from disease onset to the date of last follow up was 56.2 months. 8 patients had relapsing disease and 32 patients had non-relapsing disease. Baseline characteristics and clinical manifestations were similar between groups (table 1). Patients with relapsing disease had a statistically higher incidence of ANCA co-positivity as compared to non-relapsing patients (62.5% vs. 21.7% respectively p value- 0.03).

In patients with relapsing disease, only 14.7% (1/7 tested patients) had positive anti-GBM antibodies at the time of their relapse.

Abstract AB0687 – Table 1. Clinical features and laboratory values at baseline and at the time of relapse of Anti-GBM disease

	Non-relapsing Anti-GBM Disease (n=32)	Relapsing Anti-GBM Disease (n=8)	p-Value
Baseline characteristics			
Length of follow up, mos (range)	62.79 (2-228)	26.29 (3-78)	0.16
Time to diagnosis, days (range)	30 (1-182)	62 (5-243)	0.33
Age, Yr (range)	46.06 (17-78)	41 (14-62)	0.52
Male Gender (%)	53.1	37.5	0.44
Non-Smoking Status (%)	43.8	50.0	0.61
Renal involvement (%)	93.8	100	0.48
Peak creatinine mg/dL (range)	7.24 (0.8-17.6)	5.37 (1.6-13.7)	0.33
Pulmonary hemorrhage (%)	30.0	37.5	0.69
Positive Anti-GBM antibodies (%)	73.9 (17/23)*	37.5 (3/8)	0.07
Positive ANCA (%)	21.7 (5/23)*	62.5 (5/8)	0.03
Characteristics at time of relapse			
Time to relapse, weeks (range)		77.14 (6-480)	
Positive Anti-GBM antibodies (%)		14.3 (1/7)*	
Positive ANCA (%)		75 (3/4)*	
Renal involvement (%)		83.3	
Peak creatinine mg/dL (range)		3.42 (2.7-4.85)	
Pulmonary hemorrhage (%)		71.4	

Values are expressed as mean (range) or percent

*Incomplete data available. Number of positive patients out of total tested represented in parentheses

Conclusions: In this study, anti-GBM positivity did not correlate with disease relapse in patients with anti-GBM disease. Patients with relapsing disease had a higher incidence of ANCA positivity, consistent with previous investigations. In patients with newly diagnosed anti-GBM disease, ANCAs should be obtained to assess for the risk of relapse. Larger studies are needed to validate our results.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.1671

AB0688

PULMONARY AND THORACIC VASCULAR FINDINGS OF BEHCET'S DISEASE AT COMPUTED TOMOGRAPHY

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Background: Behcet disease is a multisystemic and chronic inflammatory disorder with unknown etiology. Pulmonary involvement is relatively infrequent.

Objectives: To investigate the pulmonary parenchymal changes and thoracic vascular abnormalities due to Behcet's disease at computed tomography.

Methods: 21 patients diagnosed with a diagnosis of Behcet's disease between 2004–2017 were evaluated. Clinical, laboratory, and thoracic computed tomography findings were retrospectively evaluated. Also, the immunosuppressive treatments of the patients were documented.

Results: 18 of the patients (85.7%) were male. The mean age of the patients was 44.42±11.7 years, onset age of the disease was 32.4±6.4 years, length of follow-up was 8.1±2.3 years. 13 of the patients (61.9%) had hemoptysis at the time of initial diagnosis of Behcet's disease. 14 of the patients (66.6%) showed vascular involvement except pulmonary system. Pulmonary artery aneurysm was observed in 9 patients (42.8%). Pulmonary artery thromboembolism was observed in 15 patients (71.4%). Thoracic computed tomography demonstrated that 5 patients (23.8%) with pulmonary infarct, 4 patients (19%) with pulmonary consolidation, 5 patients (23.8%) with ground-glass opacity in the lung representing pulmonary haemorrhage, 2 patients (9.5%) with pleural effusion, 7 patients (33.3%) with non-specific parenchymal changes, and one patient with focal atelectasis. Also, one of the patients had intracardiac thrombus. Five of the patients were smokers.

Conclusions: Aneurysm of the pulmonary artery with or without thrombosis is the most common manifestation of Behcet's disease. Pulmonary vasculitis and thrombosis of pulmonary vessels result in infarction, hemorrhage, focal

atelectasis. Thoracic computed tomography can clearly demonstrate the pulmonary and vascular abnormalities of Behçet's disease.

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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.6682

AB0689 EXTRAVASCULAR MANIFESTATIONS OF TAKAYASU ARTERITIS: HISTORICAL COHORT STUDY IN KOREA

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Background: Takayasu arteritis (TAK) is systemic disease characterised by large vessel involvement. Although the vascular characteristics of TAK are well characterised, there is no well-organised study demonstrating the extravascular manifestations of TAK.

Objectives: To evaluate the characteristics of extravascular manifestations of TAK, and to identify the association between vascular and extravascular manifestations of TAK.

Methods: TAK patients from two independent cohorts who fulfilled the 1990 ACR classification and encoded M314 according to ICD-10 code between January 2012 and October 2017 were included in the study. Characteristics of the patients were retrospectively collected from the electronic dataset. A radiologist reviewed CT scans of all included patients to evaluate the pattern of vascular involvement and presence of sacroiliitis. Clinical findings including uveitis, skin lesion, oral ulcer, arthritis, and inflammatory bowel disease (IBD) were reviewed. Logistic regression analysis was performed to evaluate the association between vascular and extravascular manifestation.

Results: A total of 268 TAK patients were included. Mean age at diagnosis was 41.2±14.2 years and 236 (88.1%) were female. The most commonly involved vessel was common carotid artery (176 [65.7%]), and the most common type of vascular involvement was type V (120 [44.8%]). Extravascular manifestation of TAK was observed in 51 (19.0%) patients (table 1). The most common extravascular manifestation was arthritis (axial arthritis [sacroiliitis] [7.1%] and/or peripheral arthritis [6.0%]) (11.9%) followed by recurrent aphthous stomatitis (8.6%) and IBD (2.6%). In multivariable logistic regression analysis, the following factors were significantly associated with presence of arthritis (axial and/or peripheral arthritis): type IIB vascular involvement (adjusted OR 2.956, 95% CI 1.337–6.537, p=0.007) and erythrocyte sedimentation rate (ESR) (adjusted OR 1.014 95% CI 1.003–1.025, p=0.012).

Abstract AB0689 – Table 1. Extravascular manifestations of Takayasu arteritis

	n=268
Any extravascular manifestation	51 (19.0%)
Arthritis (axial arthritis [sacroiliitis] and/or peripheral arthritis)	32 (11.9%)
Recurrent aphthous stomatitis	23 (8.6%)
Inflammatory bowel disease	7 (2.6%)
Erythema nodosum	4 (1.5%)
Uveitis	2 (0.7%)

Conclusions: Extravascular manifestations of TAK are not rare and observed in up to one-fifth of patients. The most common extravascular manifestation was arthritis including sacroiliitis (11.9%). Type IIB vascular involvement pattern and high ESR were significantly associated with arthritis in TAK.

Acknowledgements: None

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4386

AB0690 DIAGNOSTIC VALUES OF ENDOTHELIN-1 IN PATIENTS WITH SYSTEMIC NECROTIZING VASCULITIS

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Background: Systemic necrotizing vasculitis (SNV) is characterised by destructive and inflammatory changes in the vessels. Binding of autoantibodies and immune complexes on the surface of endothelial cells stimulates the synthesis of endothelin-1 (ET-1), which leads to activation of macrophages and adhesion of neutrophils, remodelling of the vascular wall and its damage.

Objectives: To evaluate the serum level of ET-1 in patients with SNV and the possibility of its using for the diagnosis of SNV and involvement of individual organs.

Methods: The study included 36 patients with SNV (polyarthritis nodosa – 8, ANCA – associated vasculitis – 28) and healthy controls (n=26). Clinical activities of patients were calculated according to the Birmingham Vasculitis Activity Score (BVAS). All patients had active disease (BVAS >11). The serum levels of ET-1 (pmol/L) were determined by immunoassay analysis using the kits of Biomedica. The outcomes of this study were the differences in marker levels between patients with active SNV and healthy controls, patients with different forms of vasculitis, with varying degrees of BVAS activity, with involvement different organs and systems estimated by analysis of the absolute changes in marker levels and the areas under receiver operating characteristic (ROC) curves (AUC).

Results: The level of ET-1 (M±σ) in the general group of patients with SNV was 0.31±0.24 and did not differ significantly from the control group (0.27±0.10, p>0.05). At the same time, in patients who did not receive at screening glucocorticoids and cytotoxic agents (n=9), it was significantly elevated (0.62±0.58, p=0.03). However, ROC analysis indicated the moderate sensitivity (67%) and the low specificity (48%) of ET-1 for diagnosis of SNV. There were no significant differences in the levels of ET-1 between patients with different forms of vasculitis and with varying degrees of BVAS activity. In the analysis of the values of the ET-1 depending on the involvement of different organs and systems, it was found that only in patients with kidney involvement (n=15) its level (0.40±0.33) was significantly higher compared with patients without kidney involvement (0.28±0.22, p=0.04) and control group (p<0.01). ROC analysis showed that the AUC for ET-1 is 0.75±0.10 (p=0.004), which indicates acceptable capacity for ET-1 differentiate groups of patients with kidney involvement and patients without kidney involvement (sensitivity – 80.0%, specificity – 78.3%).

Conclusions: The serum levels of ET-1 were elevated in patients with SNV with kidney involvement (48% compared to healthy controls and 43% compared with patients without kidney involvement), which can be used for diagnostic purposes.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4570

AB0691 INTERSTITIAL LUNG DISEASE AND MICROSCOPIC POLYANGIITIS IN CHILEAN PATIENTS

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Background: Microscopic Polyangiitis (MPA) is an ANCA associated vasculitis (AAV), associated with p-ANCA (perinuclear) fluorescence pattern and anti-myeloperoxidase (MPO) specificity. Most frequently involved organs are kidney (80%–100%), peripheral nervous system and skin (30%). There is Pulmonary involvement in 25%–35% of patients, being alveolar haemorrhage frequently described. Interstitial lung disease (ILD) has also been recognised.

Objectives: The aim of our study is to report the characteristics of MPA Chilean patients with ILD and to compare it with other series.

Methods: Retrospective study. Patient diagnosed between 2007 and 2016 at the Hospital Clínico Universidad de Chile, with ILD, defined as interstitial lung disease on CT scan with Usual Interstitial Pneumonia (UIP) or Non Specific Interstitial Pneumonia (NSIP) pattern, and MPA were included. Demographic, clinical, laboratory and mortality data were plotted. Data from other series were compared with our results. Other causes that could explain the pulmonary involvement were excluded.

Results: From 94 patients with AAV, 36,1% were MPA, being 16 patients with ILD. All were Hispanic, median age 65.3 years, 32–84 female 62,5% (table 1). Common manifestations were constitutional symptoms (100%), weight loss (68,7%) and fever (68,7%). All patients had anaemia, high ESR (mean 84 mm/hr. range 33–120) and CRP (8–22 times above upper normal limit). All patients were ANCA-p and MPO positive. In 10 cases ILD was diagnosed concomitantly with MPA and in 6 was 0.5 to 15 years before. 4 patients developed pulmonary haemorrhage. Images patterns were 10 UIP and 5 NSIP. All patients received corticosteroid as induction therapy, 15 also received cyclophosphamide. One patient plasmapheresis, and one received Rituximab after a relapsed. Azathioprine was used as