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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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.

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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