SAT0525

AN UPDATE ON PULMONARY ARTERY INVOLVEMENT IN BEHCET'S SYNDROME: MORE PULMONARY ARTERY THROMBOTIC DISEASE AND A BETTER OUTCOME

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Background: Pulmonary artery involvement (PAI) is the most common form of arterial involvement in Behçet's syndrome (BS) and is a well-known cause of mortality and morbidity. A previous survey¹ by our group had analysed the clinical characteristics and outcome of 47 pts with PAI registered between 2000–2007 and shown that: 1. the overwhelming male predominance was decreasing; 2. 1/4th of the pts had isolated pulmonary artery thrombosis (PAT); and 3. the mortality rate was 26% after a mean follow-up of 7 years. Recently we had the impression that female/male ratio was perhaps increasing, we are becoming to see more pts with isolated PAT and that we started to use more biologics.

Objectives: This survey was done to look at these assumptions formally in a recent group of BS pts with PAI.

Methods: We reviewed the records of about 3390 pts with BD who were registered at our multidisciplinary BS clinic between Jan 2008 and Jan 2018. From this group we identified 47 (42 M/5 F) pts who were diagnosed with PAI and recorded all information regarding clinical characteristics, outcome, radiological studies and medical or surgical treatment.

Results: The prevalence of pts with PAI decreased from 1.9% to 1.4% in the recent cohort. The M/F ratio, the mean age at the onset of PAI were similar across 2 cohorts. The frequencies of other vascular involvement were almost similar to that observed in the previous cohort. However, there were more pts with neurological disease (parenchymal) in the recent cohort. As usual, PAT or PAA were mostly bilateral and involved descending lobar arteries. On the other hand, types of PAI involvement at presentation had changed substantially: those with isolated PAT reached a share of 45%. Forty-five (96%) pts received cyclophosphamide (Cy) pulses for a mean of 6±4 courses, which was significantly shorter compared to that observed in the previous cohort. A total of 23 (49%) pts received infliximab because of relapsing course, side effects or unresponsiveness to Cy for a mean follow-up of 8±4 mo while only 2 pts received anti-TNF's in the older cohort. 4 pts had lung surgery. These were lobectomies in 3 pts due to giant rapidly progressing aneurysms and cavectomy in 1. Bronchial artery embolization was done in 3 pts because of refractory hemoptysis. By Jan 2018 the outcome of information was available on 45/47 pts: 4 pts (all male) (8%) had died, 2 were lost to follow-up after 12 and 16 mo of follow-up and the remaining were alive after a median follow-up of 5 [IQR:3–9] years. The causes of deaths were massive hemoptysis in 3, severe pulmonary hypertension in 1. As shown in the figure, the survival has improved significantly in the recent yrs.

Conclusions: The surveys of 2 consecutive cohorts showed that the prevalence of PAI perhaps mildly decreased, isolated PAT type of involvement was with considerably higher frequency and the outcome was getting better. Cy is still the first agent in these pts however its duration of use became much shorter and anti-TNF's mainly infliximab was used in about half of the cohort. The survival seems to have improved significantly. This could have been due to a decreased severity of the type of PAI, with isolated PAT becoming the most frequent type, or a better management.

REFERENCE:

[1] Seyahi E. Medicine (Baltimore) 2012.

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SAT0526

IS RELAPSE RATE OF GIANT CELL ARTERITIS IN REAL-LIFE EXPERIENCE LOWER THAN IN THE CONTROLLED TRIALS? RESULTS OF A RETROSPECTIVE, MULTI-CENTRE COHORT STUDY

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Objectives: Corticosteroids (CS) are accepted as the standard first-line treatment for giant cell arteritis (GCA). However, controlled trials of tocilizumab and abatacept demonstrated relapse rates of up to 70%–80% in patients on CS-only protocols in 12–24 months. Though level of evidence is low and not suggested by guidelines (except for methotrexate), conventional immunosuppressives (ISs) are also commonly used. We aimed to assess the relapse rates in patients with GCA in routine practice, retrospectively.

Methods: We assembled a retrospective cohort of patients with GCA from Turkey. All data was abstracted from records. Relapse was defined as any new manifestation or increased acute-phase response leading to the change of the CS dose or use of a new therapeutic agent by the treating physician.

Results: The study included 156 (F/M: 95/61) patients with GCA (table 1). The mean age at disease onset was 67.8±9.1 years. Polymyalgia Rheumatica was also present in 48 (30.8%) patients. Diagnosis was proven histopathologically in 99 patients. All patients received 1 mg/kg/day CS for remission induction, additional CS pulses were given to 36 (23.1%) patients. Conventional ISs including methotrexate and azathioprine were used in 89 (56.1%) and 26 (16.6%) patients respectively, while 10 (6.4%) patients received biologic treatments (8 tocilizumab, 2 etanercept). Forty-four (28.2%) patients used only CS during follow-up. Follow-up of at least 6 months was available for 132 patients, and median follow-up duration was 35 (6–268) months. Relapses occurred in 27 (20.5%) patients during follow-up. Mortality rate was 7.5% (n=10) during follow-up. VDI score was 2.4±1.7. Main causes of damage were related to CS treatments such as cataract, osteoporosis and diabetes mellitus.

	Giant Cell Arteritis (n=156)
Manifestations of systemic inflammation, n/156 (%)	
Anemia (<12 mg/dl for female, <13 mg/dl for male)	94 (60.2 %)
Erythrocyte Sedimentation Rate (mm/hour) *	84.6 ± 30.1
C-reactive protein (mg/l) *	84.8 ± 65.3
Malaise	132 (84.6%)
Weight loss	72 (46.1%)
Fever	35 (22.4%)
Stiffness and/or pain muscles and joints	48 (30.8 %)
Manifestations of Vascular Ischemia, n /156 (%)	
Headache	144 (92.3%)
Scalp Tenderness	78 (50 %)
Jaw Claudication	57 (36.5 %)
Ocular Symptoms	72 (46.2 %)
Extremity Claudication	14 (8.9 %)
Neurological Manifestations	14 (9 %)

Conclusions: In this first multi-centre series of GCA from Turkey, we observed that only one fifth of patients had relapses during a mean follow-up of 35 months. This lower relapse frequency suggests a different clinical spectrum in routine