THU0318 CORONARY ARTERY DISEASE IN PATIENTS WITH BEHÇET'S DISEASE: A RETROSPECTIVE, SINGLE CENTER STUDY

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Background: Behçet's disease (BD) is a chronic inflammatory disease affecting various size of arteries and veins. Coronary artery disease (CAD), a life-threatening condition, is rarely reported in patients BD.
Objectives: To investigate the clinical characteristics of BD patients complicated with CAD, and to elucidate the potential risk factors of CAD in BD patients.
Methods: We retrospectively reviewed all the medical records of BD patients who were admitted to our institute from 2001 to 2016. CAD was defined as aneurysm, stenosis and (or) occlusion of coronary arteries confirmed by angiography or contrast-enhanced computer tomography. BD patients with CAD and age- and gender-matched BD patients without CAD (at 1:3 ratio) were enrolled. Demographic, clinical and laboratory data were systemically collected, analyzed and compared between two groups.
Results: In total, 19 patients, including 17 male and 2 female, were complicated with CAD. The mean onset age of BD was 34 and the mean duration from the onset of BD to the diagnosis of CAD was 4.1 year. Angina pectoris (8/19) and acute myocardial infarction (8/19) were the most common cardiac symptoms, arrhythmia was presented in one patient, and three patient remained asymptomatic. Coronary artery aneurysm, stenosis and occlusion were presented in 9, 13 and 3 patients, respectively. Smoking (7/19) was frequently observed, while hypertension (3/16), diabetes mellitus (2/19), obesity (1/19) and alcohol consumption (1/19) were rarely present. Additionally, seven arterial and two venous extra-cardiac vasculopathies were presented. Oral ulceration (19/19) and skin lesions (16/19) were the most common BD-associated symptoms. Comparing with BD patients without CAD, patients with BD presented with higher ESR (mean, 34.4 vs 16.3 mm/hr, p=0.0018) and CRP (mean, 36.4 vs 12.2 mg/L, p=0.002), more frequency of skin lesions (84% vs 55%, p=0.0334) and pathergy reactions (37% vs 26%, p=0.0103). Furthermore, multivariate analysis confirmed that elevated CRP was a independent risk factor of CAD (OR 1.032, 95% CI 1.011–1.053, p=0.003).
Conclusions: CAD, a rare complication of BD, predominately affect male patients. BD patients with CAD presented with active BD disease symptoms and elevated inflammatory markers, which implicated aberrant vascular inflammation was the key mechanism of CAD in BD patients. CRP, but not traditional CAD risk factors, was the risk factor of CAD in BD patients.
Disclosure of Interest: None declared
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THU0319 MR ANGIOGRAPHY FOR EVALUATION OF VASCULAR INFLAMMATION IN ELDER PATIENTS WITH LARGE VESSEL VASCULITIS

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Objectives: The aim of the study was to evaluate the feasibility of MR angiography (MRA) for evaluation of vascular inflammation in elder patients with large vessel vasculitis.
Methods: 16 patients with established on PET with 18F-FDG large vessel vasculitis (14 female and 2 male; average age 66 years) were enrolled in our study. 14 patients had got giant cell arteritis (just only 3 biopsy-proven cases), 2 patients with isolated aortitis of thorax aorta and 2 cases of PMR-associated arteritis. The average duration of any disease was 8 months. All patients underwent MRA with or without contrast enhancement of aorta and its branches at 1.5 Tesla (Siemens Magnetom Essenza). All patients had repeated MRA at 6 and 12 months. All images were studied by one specialist. We evaluated the role of mural oedema as a sign of activity of vasculitis. The results of MR-angiography were compared with clinical and laboratory data, ultrasound and PET with 18F-FDG.
Results: A total of 42 MRA were obtained in 16 patients. Significant mural oedema of thorax aorta or large arteries was shown by imaging in 20 of 42 cases (48.0%) and correlated with clinical and laboratory signs of vasculitis activity, progressive arterial stenosis detected by ultrasound and increased uptake of 18F-FDG on PET. 12 patients with high degree of inflammation on MRA were streroid-naïve. In 22 cases (52%), MRA had showed mild or moderate oedema of the arteries' wall. Low-moderate activity of vascular inflammation in our patients was associated with moderate or high of immunosuppressive therapy (prednisolone 40–60 mg/day). However, gradual reduction in the intensity of immunosuppression in 4 patients with mild mural oedema was associated with development of relapse of large vessel vasculitis. Notably, contrast enhancement did not improve significantly edema imaging.
Conclusions: Visualisation of artery wall oedema by MRA may be a usefull approach to detect persisting inflammation in elder patients with large vessel vasculitis.
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THU0320 DIFFERENT PATTERNS OF VASCULAR INVOLVEMENT IN PET/CT ACCORDING TO CRANIAL SYMPTOMS IN BIOPSY PROVEN GIANT CELL ARTERITIS, A PRELIMINARY STUDY

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Background: GCA is the most frequent vasculitis in patients over 50 years old. It involves large vessels (LV) such as aorta, carotid, vertebral and subclavian arteries. After the development of new imaging techniques, extracranial LV involvement has been increasingly described. Past studies showed a trend in worse prognosis in those patients with aortitis at diagnosis but larger studies are needed to determine the clinical implications of these results. Correlation between symptoms at diagnosis and PET evaluation has not been done.
Objectives: To evaluate aortic and supraaortic involvement in a new onset GCA cohort through PET/CT. To compare aortic and supraaortic involvement in patients with and without cranial ischemic symptoms.
Methods: Prospective study including all newly diagnosed biopsy-proven GCA patients in 2016 according to ACR criteria. PET/CT was performed during initial evaluation. Clinical data, blood tests and PET/CT results were recorded. Ischemic symptoms included jaw claudication (JC) and visual symptoms. Analysis was made using Stata IC/14. Patients were compared according to clinical presentation (presence vs absence of ischemic symptoms) with Fisher exact test.
Results: 18 patients were included (63.6% were women). At diagnosis age was 77.4±8.7 years (mean ± SD). Patients presented headache (78.6%), polymyalgia rheumatica (14.3%), constitutional syndrome (42.8%), JC (42.9%) and visual symptoms (35.7%). On physical examination pulse was decreased in 78.6% of patients. Blood tests showed anemia in 71.4% with an ESR 90.41±33mm/h (mean ± SD). PET/CT was performed after (median, intercuartilic range) 6.5 days (2 - 10 days). Analysis of vascular uptake was semiquantitative. Aortic involvement was present in 42.9% of patients. Supraaortic and vertebral arteries were involved in 42.8%. Baseline characteristics were similar to other cohorts. At the time of diagnosis, proportion of aortitis was significantly lower (p=0.049) in patients with ischemic symptoms when compared to those without. We could not find any differences between these two groups when analyzing the presence of increased vascular uptake in carotid and vertebral arteries. Limitations of the study are the small size of the cohort and the variability of the PET/CT protocols among literature. However, we think this study is important because it shows the differences between these two subgroups of GCA.
Conclusions: In this study, patients with cranial ischemic symptoms showed less involvement of large vessels when compared to those GCA patients that presented with systemic symptoms. These results suggest that there may be different physiopathological subsets leading to GCA. Further studies are needed to better understand these mechanisms.
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THU0321 ALTERATIONS IN THE PERIPHERAL B CELL COMPARTMENT IN PATIENTS WITH EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS (CHURG-STRAUSS SYNDROME)

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Background: Eosinophilic granulomatosis with polyangiitis (EGPA) belongs to the group of ANCA associated vasculitides. While the combination of asthmatic symptoms and vasculitis characterize the disease clinically, eosinophilia and increased serum IgE concentrations are serologic hallmarks. The role of B lymphocytes in EGPA has not been defined so far, but therapeutic response to rituximab in EGPA points towards a role of B-cells in the pathogenesis of EGPA.
Objectives: To characterize the peripheral B cell compartment in patients with EGPA, to analyze the in vivo potential of B lymphocytes to class-switch to IgE, and to assess in vitro the differentiation of naïve B cells to IgE-secreting plasmablasts.
Methods: Laboratory work-up included ANCA-status, eosinophils, IgE, IgG, IgA, IgM, and peripheral $CD19^+$ B-cell count. B cell subpopulations (naïve, marginal zone, class-switched B cells and plasmablasts) were analyzed by staining PBMCs with fluorescent-labeled monoclonal antibodies against: CD27, CD20, CD38, IgD, IgG, IgA, IgE, CD21, and BAFF-R. For in vitro differentiation assays magnetically isolated B lymphocytes from EGPA patients and aged matched healthy controls were stimulated with CD40L and IL-21 and IL-4 in enriched Iscoves' medium supplemented with 10% FCS, 1 μg/mL insulin, 2.5 μg/mL apo-transferrin, 1% nonessential amino acids, 2 mmol/L glutamine, and 1 μg/mL reduced glutathione. Starting the culture with equal number of B cells, the absolute number of plasmablasts, and IgE class switched cells after 9 days was determined by counting the events in the $CD27^{high}CD38^{high}$ gate or the $IgG/A/D^-IgE^+$ gate by flow cytometry. IgE secretion in the supernatant was measured by ELISA.
Results: 18 patients (8 females, median age 59 years) with EGPA diagnosed according to ACR and CHC-criteria were included into the study. 22% of patients were ANCA-positive. Immunosuppressive therapy was azathioprine in 11 patients, methotrexate in 3 patients, and leflunomid or mycophenolate in one patient each and two patients received no immunosuppressive treatment. 7 patients had a history of a prior cyclophosphamide therapy. Median lymphocyte count was