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FRI0321 CIRCULATING IL-6 AND OTHER METABOLIC BIOMARKERS COMPARING EFFECTS OF MODIFIED-RELEASE PREDNISONE (MR) AND IMMEDIATE-RELEASE PREDNISOLONE (IR) IN NEW GCA

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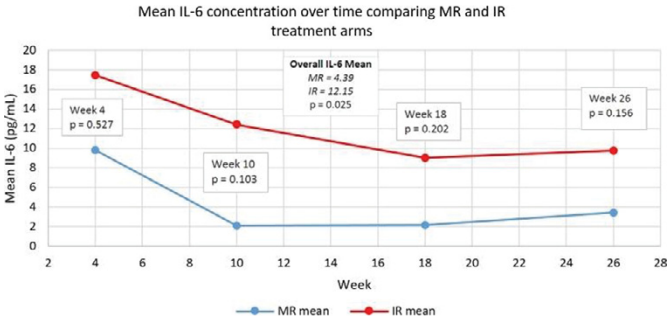
Background: GCA may be an interleukin-6 (IL-6) driven disease and IL-6 blockade is emerging as an exciting therapy of IL-6.[1] We measured serial IL-6 levels in new patients with GCA treated in an RCT of modified-release prednisone (MR) versus immediate-release prednisolone (IR) used in a tapering regimen conforming to BSR guidelines.[2,3]
Methods: Patients (n=12) were randomised into two treatment arms (7 MR, 5 IR) and followed up over 26 weeks. IL-6 samples were collected at 9am at weeks 4, 10, 18 and 26 and were measured using Beckman Coulter IL-6 immunoassay, validated in a controlled study according to ACB criteria. We also measured bone markers (CTX, P1NP, vitamin D), HbA1c, cortisol, ACTH and PTH.
Results: Significantly higher overall mean IL-6 levels were seen in the IR arm (n=5) compared to MR (n=7) [unpaired two-tailed Student's t test]. IL-6 levels in both arms were lowest between weeks 4–10 and continued to decrease in the IR arm to week 26, whereas lower but constant levels were seen in the MR arm (Figure)
Mean CTX concentration was significantly higher at week 4 (M =0.29, SE =0.04) compared to Week 26 (M =0.13, SE =0.02) p=0.002. No significant difference was seen between treatment arms.
Patients on MR had complete suppression of ACTH compared to IR ($p<0.05$) without a significant difference between groups in 9 am cortisol levels (p=0.3412). No significant differences were seen in levels of vitamin D, calcium, PTH, ESR, CRP, or HbA1c.



Conclusions: Our study suggests that elevated levels of Il-6 are better suppressed by MR prednisone therapy compared to IR prednisolone in new GCA. Bone resorption marker CTX was significantly reduced in both treatment arms. ACTH suppression with MR prednisone may reflect a greater impact on the HPA axis although cortisol levels were not affected.
Our findings suggest that MR prednisone may warrant further clinical trial investigation in GCA.
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FRI0322 ASSESSMENT OF DAMAGE AND PROGNOSIS IN PATIENTS WITH ADULT IGA VASCULITIS: RETROSPECTIVE MULTICENTERED COHORT STUDY

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Background: IgA Vasculitis is a leukocytoclastic vasculitis involving small vessels with depositions of immune complexes containing IgA. There is limited data for the prognosis of adult IgA Vasculitis, with also no damage assessment.
Objectives: We aimed to evaluate the clinical characteristics, treatment, outcome and damage of patients with adult IgA Vasculitis.
Methods: We assembled a retrospective cohort of patients with adult IgA Vasculitis, from tertiary Rheumatology Centers in Turkey. All data were abstracted from medical records. Birmingham Vasculitis Activity Score (BVAS),prognostic Five Factor Score (FFS) and vasculitis damage index (VDI) were calculated.
Results: The study included 52 (male/female:40/12) patients with adult IgA Vasculitis. The mean age was 42.2±17 years. Infection history within 6 weeks before presentation was present in 22 (42.3%) patients (18 upper respiratory tract,3 gastrointestinal and 1 urinary tract). Cutaneous manifestations were the most common clinical manifestations (Table 1). All patients were treated with oral glucocorticoids (GC).As additional immunosuppressive agents, azathiopirine was given to 21 (40.4%) and pulse cyclophosphamide to 11 (21.2%) patients. Twenty-eight patients (53.9%) had follow-up of 28.6 months. Five (17.8%) patients relapsed during follow-up. While 3 relapses were major, 2 of them were minor relapses. At the last visit, disease status was evaluated as active or treatment failure by the treating physician in 6 (21.4%) patients. Mortality was 3.6% (n=1) during follow-up, due to pneumonia. The mean VDI score was 0.6 in the last visit. Nine (32.1%) patients had at least one damage item at the end of follow-up period.

Table 1. Clinical characteristics of patients with adult IgA Vasculitis

	Adult IgA Vasculitis (n=52)
Laboratory parameters	
Erythrocyte Sedimentation Rate (mm/hour)*	32.7±22
C-reactive protein (mg/l)†	25.2 (1–94.9)
Proteinuria (>300mg/24 hours)	28 (53.9%)
Creatinine (mg/dl)*	0.9±0.4
Clinical Manifestations, n/52 (%)	
Fever	7 (13.5%)
Weight loss	14 (26.9%)
Myalgia/Weakness/Leg tenderness	24 (46.2%)
Arthritis and/or arthralgia	46 (88.5%)
Neurologic manifestations	1 (1.9%)
Testicular pain or tenderness	3 (5.8%)
Recent onset or severe hypertension	2 (3.8%)
Cutaneous Manifestations	48 (92.3%)
Gastrointestinal manifestations	39 (75%)
FFS=0	29 (55.8%)
FFS=1	15 (28.8%)
FFS≥2	8 (15.4%)
BVAS score at diagnosis*	4.1±1.7

FFS: Five Factor Score, BVAS: Birmingham Vasculitis Activity score. *Mean ± SD; †Median (Minimum–maximum.)

Conclusions: Our results showed that approximately one fifth of patients with adult IgA Vasculitis had relapses during follow-up. At the end of follow-up, one third of patients had at least one damage item. Although, 45% of patients had FFS≥1, the mortality rate was observed to be low in the present study.
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FRI0323 COMPARISON OF BIOPSY PROVEN GIANT CELL ARTERITIS IN NORTH AMERICA AND SOUTHERN EUROPE: A POPULATION-BASED STUDY

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Objectives: To compare clinical characteristics, treatment, long-term follow-up and prognosis of two population-based cohorts of patients with biopsy-proven giant cell arteritis (GCA) from Olmsted County, Minnesota, USA (Olmsted cohort) and the Reggio Emilia area, Northern Italy (Reggio cohort).
Methods: All patients residing in Olmsted County and the Reggio Emilia area

with a new diagnosis of biopsy-proven GCA in 1986–2007 were retrospectively identified. Patients were followed from GCA diagnosis to death, migration or September 2011. Comparisons were performed using Chi-square and rank sum tests, Kaplan-Meier methods and Cox models.
Results: The study included 110 patients in the Olmsted and 144 in the Reggio cohort. Compared with the Olmsted cohort, patients from the Reggio cohort were younger (mean±SD age 74.6±7.4 years vs 77.8±7.6, p=0.002), more likely to have cranial symptoms (93% vs 86%, p=0.048), temporal artery abnormalities at physical examination (68% vs 42%, p<0.001), partial or complete unilateral or bilateral permanent vision loss (21% vs 6%, p=0.001), systemic symptoms (67% vs 46%, p=0.001) and polymyalgia rheumatica at or before GCA diagnosis (47% vs 26%, p<0.001). Scalp tenderness was less common in the Reggio cohort (36% vs 49%, p=0.033). ESR and CRP were higher (mean 88±29 mm/h vs 73±77, p<0.001 and mean 89.0±60.2 mg/L vs 35.2±43.4, p<0.001 respectively) and hemoglobin lower (mean 11.2±1.4 g/dl vs 11.8±1.4, p=0.004) in Reggio than in the Olmsted cohort. Patients from the Olmsted cohort received a higher initial prednisone dose (mean 53.6±15.3 mg/day vs 49.5±12.8, p=0.001). There were no differences in relapse rates, cumulative glucocorticoid (GC) dosages at 1, 2 and 5 years, and time to first GC discontinuation. However, the Reggio cohort reached a prednisone dose <10 mg/day sooner (median 4.9 months vs 7.9, p=0.012) and had a first relapse later (median 13.6 months vs 7.9, p=0.003) than the Olmsted cohort. Patients from the Reggio cohort had a significantly higher mortality compared to those from the Olmsted cohort (HR 1.72, 95% CI 1.12–2.65 adjusted for age and sex).
Conclusions: Genetic and/or environmental factors may contribute to the differences in clinical characteristics and disease outcomes observed in this study comparing patients with GCA from North America and Southern Europe.
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FRI0324 SMALL VESSEL VASCULITIS SURROUNDING A PRESERVED TEMPORAL ARTERY: A DIAGNOSTIC ALGORITHM TO ASSESS CLINICAL SIGNIFICANCE

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Background: Systemic vasculitides are complex and heterogeneous diseases with overlapping features that frequently pose a diagnostic challenge to clinicians. The temporal artery biopsy (TAB) is the gold standard for the diagnosis of giant cell arteritis (GCA) but, occasionally, TAB show inflammation of small vessels surrounding a spared temporal artery (SVV) as the only pathologic finding. Ultimate diagnosis and, consequently, optimal treatment remain uncertain in these patient.
Objectives: To analyze the final diagnosis of patients with SVV surrounding a spared temporal artery after a pre-established diagnostic algorithm and to identify clinical and laboratory findings with potential usefulness in predicting the ultimate diagnosis.
Methods: Patients with TAB showing SVV were subjected to the diagnostic algorithm displayed in figure 1, completed by at least 1 year follow-up. Clinical and laboratory features at the time of diagnosis were recorded. The algorithm led to the following final diagnosis: GCA, other systemic vasculitis and undetermined condition. Chi-squared and ANOVA tests were used for statistical comparison using IBM SPSS Statistics 20.
Results: From 1998 to 2007, 380 TAB were performed in our institution. Biopsy disclosing small vessel inflammation surrounding a normal temporal artery (SVV) was described in 47 (12%) patients. In all patients TAB was selected as the first invasive procedure because GCA was initially considered the most likely diagnosis. Accordingly, 24 (51%) fulfilled at least 3 ACR classification criteria for GCA. 7 patients declined to undergo subsequent work-up to complete the diagnostic algorithm, 10 died or were lost to follow –up before completing 1 year. All of them were excluded. The study cohort consisted of 30 patients (19 women and 11 men) aged 77±10.4 years followed for 55.16±55.20 months. In 13 patients the final diagnosis was consistent with GCA based on the absence of SVV in other territories, large-vessel inflammation by imaging or subsequent development of aortic aneurysm; in 12 SVV was subsequently demonstrated in other territories and were diagnosed with other systemic vasculitis (7 AAV, 0 cryo, 3 PAN, 0 vasculitis associated to autoimmune diseases, 2 unclassified small vessel vasculitis), and in 5, diagnosis remained undetermined. No significant differences in clinical or routine laboratory abnormalities were found among patient subgroups stratified according to the final diagnosis.
Conclusions: Inflammation of small vessels surrounding a spared temporal artery in a TAB conveys a substantial diagnostic uncertainly. After a detailed diagnostic work-up most of patients can be diagnosed with GCA. However other systemic vasculitis requiring more aggressive treatment may disclose similar histopathologic findings and are also frequent and diagnosis remains uncertain in a substantial proportion of cases. Search for more accurate molecular biomarkers is necessary for a better interpretation of these findings
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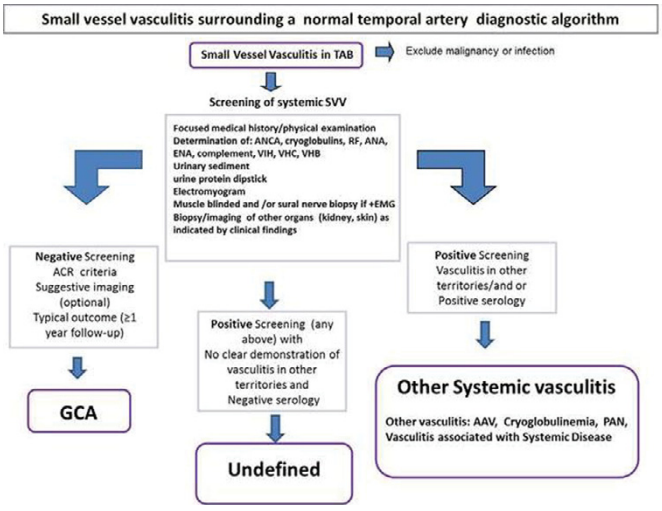

Figure 1

FRI0325 PREVALENCE OF TAKAYASU ARTERITIS IN YOUNG WOMEN WITH ACUTE ISCHEMIC HEART DISEASE

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Background: Takayasu arteritis (TA), a systemic vasculitis typically occurring in female patients aged ≤40, can affect the coronary arteries and cause ischemic heart disease. The prevalence of TA among young females with acute ischemic heart disease is undetermined.
Objectives: In this study, we investigated the prevalence of TA in young women presenting with ischemic heart disease in the Emergency Department.
Methods: We conducted a retrospective evaluation of the hospital records of 172,790 consecutive female patients aged <45, who accessed the Emergency Department of our institution over 8 consecutive years (2007–2015). The prevalence of TA and of other etiologies of ischemic heart disease was determined. Diagnosis of TA was established based on the 1990 American College of Rheumatology criteria.
Results: Overall, 2,090 women aged <45 presented to the Emergency Department with chest pain, dyspnea, palpitations, angina, heart failure, or cardiac arrest; 40 had confirmed acute ischemic heart disease. The etiology was "classic" atherosclerosis in 24 cases (60%), TA in 4 cases (10%), vasospasm and sympathomimetic drug abuse in 3 cases each (7.5%), coronary artery dissection and microvascular angina in 2 cases each (5%), Takotsubo and radiation-induced cardiomyopathy in 1 case each (2.5%).
Conclusions: Although a diagnosis of TA is likely to be overlooked, TA is not infrequent in younger females presenting with acute ischemic heart disease. Specifically, TA accounted for 10% of cases of acute ischemic heart disease in female patients aged <45, a finding relevant to the diagnosis and management of these young patients.
Disclosure of Interest: None declared
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FRI0326 RECOMMENDATIONS FOR THE MANAGEMENT OF NEURO-BEHÇET DISEASE BY JAPANESE RESEARCH COMMITTEE FOR BEHÇET DISEASE

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Background: Central nervous system involvement is one of the most serious complications in Behçet's disease (BD). This condition is referred to as neuro-