AB0701

COMPARISON OF THE NEW ACR/EULAR CLASSIFICATION CRITERIA OF ANCA-ASSOCIATED VASCULITIS WITH THE EMA ALGORITHM IN CLASSIFICATION OF VASCULITIS

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Objectives: A new set of classification criteria for ANCA-associated vasculitis (AAV) was presented in 2017's ACR annual scientific meeting. In order to evaluate this new set of classification, we conducted the current study to compare it with the EMA's consensus algorithm for classification of systemic vasculitis proposed by Watt et al. in our centre.

Methods: One hundred and twenty-two Chinese patients with clinically diagnosed as AAV in our centre during the past 15 years were retrospectively studied. We compared the new set of ACR/EULAR's classification criteria for AAV, with the EMA's consensus algorithm with surrogate parameters, in the same cohort of patients with primary systemic vasculitis.

Results: Applying the EMA's consensus algorithm with surrogate parameters, the diagnoses were EGPA (n.3), GPA (n.55), microscopic polyangiitis (MPA) (n.47), drug related AAV (n.2), and unclassified (n.5). Using the new ACR/EULAR's classification criteria for AAV, the diagnoses were EGPA (n.8), GPA (n.33), MPA (n.65), overlap with EGPA and GPA (n.2), overlap with GPA and MPA (n.8), and unclassified (n.7) (See the below picture).

Abstract AB0701 – Figure 1

Conclusions: The new 2017 ACR/EULAR classification criteria for AAV and Watts' algorithm were all useful methods to classify patients with systemic vasculitis. The Watts' algorithm can classify all patients into a single category, with more GPA patients, less unclassified patients and without overlapping diagnosis, in comparison, the new 2017 ACR/EULAR classification criteria classified more MPA patients, more unclassified and more overlapping patients.

Disclosure of Interest: None declared

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AB0702

ALKALINE PHOSPHATASE AS A PREDICTOR OF GIANT CELL ARTERITIS – A RETROSPECTIVE ANALYSIS OF CLINICAL FEATURES AND TEMPORAL ARTERY BIOPSY FINDINGS

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Background: Giant cell arteritis (GCA) is the most common large vessel vasculitis in the United Kingdom and Northern Europe. Inadequate treatment and delay in diagnosis can lead to a number of devastating consequences.

Objectives: This study looks at whether the presence of a raised alkaline phosphatase (ALP) may aid the diagnosis of GCA, improve the sensitivity of TAB, including in the presence of other proven serum markers and whether it has any correlation with severity of clinical presentation.

Methods: Retrospective multicenter cohort study.

Information was retrospectively gathered on patients who underwent TABs following a clinical working diagnosis of GCA. Only patients who fulfilled the American College of Rheumatology (ACR) classification criteria and had ALP measured within 4 weeks of undergoing TAB were included in the study. Once patients were identified, further information was extrapolated including the values of other serum markers taken, and presenting clinical features.

Results: Our 223 patients sample population who fulfilled the inclusion criteria reflected typical GCA patients: 147 (65.9%) were female and mean age was 73.1 years (SD 10.5). TAB was positive in 54 patients (24.2%). Two patients (3.7%) who had a positive TAB had completely normal serum markers. We were unable

to comment on variables, which could have contributed to this but they likely represented already administered aggressive glucocorticoid treatment at time of serum measurement or atypical presentation of GCA. Raised ALP sensitivity at the current cut off value was very low (14.8%) but with high specificity (90.5%), which was reinforced following ROC curve analysis. Pearson coefficient analysis suggested that there was a weakly associative relationship between raised ALP and degree of clinical suspicion (Correlation 0.346, Sig 0.01)

Conclusions: Patients with a higher level of clinical suspicion and TAB positivity were more likely to have a raised ALP. However, the association strength was weak. ALP is suggested to be highly specific for TAB positivity. The association of raised ALP to degree of clinical features and suspicion of GCA is weak and of low significance, likely a reflection of the limitations of this study. Further robust research may further evaluate this observed relationship.

REFERENCES:

- [1] Yates M, Graham K, Watts RA, MacGregor AJ. The prevalence of giant cell arteritis and polymyalgia rheumatica in a UK primary care population. *BMC Musculoskelet Disord*. 172016.
- [2] Ness T, Bley TA, Schmidt WA, Lamprecht P. The Diagnosis and Treatment of Giant Cell Arteritis. *Dtsch Arztebl Int*. 2013;110(21):376–86.
- [3] Nordborg C, Nordborg E, Petrusdottir V. Giant cell arteritis. *Epidemiology, etiology and pathogenesis*. *Appl*. 2000;108(11):713–24
- [4] Hunder GG, Bloch DA, Michel BA, Stevens MB, Arend WP, Calabrese LH, et al. The American College of Rheumatology 1990 criteria for the classification of giant cell arteritis. *Arthritis Rheum*. 1990;33(8):1122–8.
- [5] Walvick MD, Walvick MP. Giant cell arteritis: laboratory predictors of a positive temporal artery biopsy. *Ophthalmology*. 2011;118(6):1201–4.
- [7] Sheehan NJ. Alkaline phosphatase and gamma-glutamyltransferase in polymyalgia rheumatica and giant cell arteritis. *Ann Rheum Dis*. 1985;44(9):644–5.

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AB0703

LONG TERM FOLLOW-UP OF BEHÇET'S SYNDROME PATIENTS TREATED WITH CYCLOPHOSPHAMIDE

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Background: Cyclophosphamide (CYC) remains an important treatment option for Behçet's syndrome (BS) patients with life-threatening conditions such as arterial aneurysms. However, several adverse events may occur with CYC and this has led to increased use of biologic agents such as rituximab in other vasculitides.

Objectives: The aim of this study is to delineate the outcome and short and long-term adverse events with CYC use among BS patients.

Methods: We conducted a retrospective chart review of all BS patients treated with oral or intravenous CYC between 1976 and 2006. Patients were called and a standard form was used for collecting demographic characteristics, CYC indication, cumulative dose of CYC and short-term serious adverse events necessitating the cessation of therapy and/or requiring hospitalisation and long-term adverse events (malignancy and infertility), and outcome.

Results: We identified 198 (M/W: 184/14) BS patients who had received CYC. After a median follow up of 17 (IQR: 9–26) years after the initiation of CYC therapy, 52 (26%) patients had died within a median duration of 4^{1–12} years, 33 (17%) were lost after a median follow-up of 9 (3.5–14) years, and 113 (57%) were contacted. CYC was prescribed for vascular involvement in 132 (67%) patients, eye involvement in 52 (26%), central nervous system involvement in 5, both vascular and eye involvement in 7 and both vascular and central nervous system involvement in 2 patients. The median duration of CYC use was 12 (IQR: 4–24) months and median cumulative dose was 13.5 (IQR: 6–49) gr. Among the 52 patients who died, reasons for death were vascular involvement in 26, malignancies in 7, infections in 5 (5 bacterial infections, 1 additional tuberculosis), neurologic involvement in 2, ischaemic stroke in 1, traffic accident in 1, and secondary amyloidosis in 1, esophageal variceal bleeding in 1, and unknown in 5 patients. Sixteen (8%) patients experienced serious adverse events associated with short-term CYC use and 1 of them died due to infection. Among these adverse events, haemorrhagic cystitis occurred in 7 patients, infections in 4 (1/4 died), leukopenia, acute myocardial

infarction, anaphylactic reaction, azoospermia, liver toxicity, and severe nausea in 1 patient each. Overall, 16 malignancies were observed in 14 (7%) patients after a median follow up of 25 (IQR:15–26) years. The malignancies were bladder carcinoma (n=4), lung adenocarcinoma (n=3), prostate adenocarcinoma (n=2), carcinoma of unknown primary origin, pancreas adenocarcinoma, t-MDS-AML, lymphoma, colon adenocarcinoma, squamous cell carcinoma and thyroid papillary carcinoma. Among the 113 patients, we were able to question regarding infertility, 67 patients (59%) had children, 22 (19.5%) did not wish to have a child and 24 (21.5%) tried to have a child, but was not able to.

Conclusions: Short term serious adverse events occurred in 8% of the patients during CYC treatment. During long term follow-up malignancies occurred in 7% and infertility in 21.5% of the patients. These results underline the need for safer and effective alternatives to CYC for serious organ involvement in BS, similar to that in other vasculitides.

Disclosure of Interest: None declared

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AB0704

CLINICAL-ANALYTICAL CHARACTERISATION OF 52 DIAGNOSED PATIENTS OF BEHÇET DISEASE WITH INCLUSION OF PAEDIATRIC CASES IN A SPANISH TERTIARY HOSPITAL

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Background: Behçet's disease (BD) is a chronic and recurrent inflammatory disease of unknown etiology, classified into polygenic autoinflammatory diseases or variable vessel vasculitis. It has a wide spectrum of symptoms with a very variable range of severity, from mucocutaneous involvement to neurological manifestations, systemic vascular or severe ocular manifestations. About 5.4%–7.6% of Behçet's cases have a paediatric debut.

Objectives: To evaluate and compare the clinical and laboratory manifestations of a series of 52 patients, adults and children, diagnosed with BD according to the classification criteria of the International Study Group of BD (ISGBD-1990).

Methods: Retrospective cross-sectional observational study, which included 43 adult patients and 9 paediatric patients diagnosed with EB in the Rheumatology Department of a Madrid tertiary hospital. The clinical-analytical characteristics of both groups were evaluated, as well as the correlation of HLA-B51 with the described symptomatology.

Results: The mean age at diagnosis of BD was 36.9±11.8 years in adults and 11.4±5.1 years in children. 27.3% of adults and 11.1% of children with BD were male, with oral ulcers close to 90% in both groups. Contrary to what was reported in other series, genital ulcers were more frequent in children (77.8% versus 65.9% of adults), as was the presence of uveitis (44.4% in children compared to 22.7% in adults) and neurological manifestations (22.2% in children versus 6.8% in adults). Joint involvement was also more frequent in children (88.9% versus 52.3% in adults), as well as fever (44% in children versus 14% in adults); being these two manifestations the only parameters that were associated in a statistically significant way with their presentation in the paediatric age in BD. In contrast, skin involvement and vascular manifestations were more frequent in adults. The positivity of HLA-B51 did not correlate statistically with any clinical manifestation, but those who had it had a mean age at diagnosis of 26.5 years compared to a mean of 39 years in those who did not present this genetic marker.

Conclusions: Behçet's disease presents with a wide spectrum of clinical manifestations, potentially serious, ranging from skin lesions to neurological or vascular manifestations. In our series, patients diagnosed at paediatric age most frequently had systemic manifestations (fever), arthritis or severe clinical manifestations such as neurological involvement or uveitis. **Limitations:** a small number of paediatric cases included in our study.

Disclosure of Interest: None declared

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AB0705

LONG-TERM OUTCOMES AND PROGNOSTIC FACTORS ASSOCIATED WITH AORTIC VALVE SURGERY IN PATIENTS WITH TAKAYASU ARTERITIS AND AORTIC VALVEREGURGITATION

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Background: Some patients with Takayasu arteritis (TA) have aortic valve (AV) involvement, which can lead to aortic regurgitation (AR). However, data on the long-term outcomes, including survival of TA patients with AR, are lacking. Moreover, previous studies were limited to patients who underwent AV surgery.

Objectives: This study aimed to characterise the long-term outcomes and clinical characteristics of TA patients with AR regardless of whether they underwent surgical intervention.

Methods: Medical records of patients with TA between January 1995 and December 2015 were retrospectively reviewed. AR was diagnosed using transthoracic echocardiography. Poor outcomes were defined as all-cause death and major adverse cardiac and cerebrovascular events (MACCE). Multivariate analysis was performed to determine the factors affecting poor prognosis in the surgical group.

Results: Of the total 105 patients with TA and AR, 41 (39.0%) underwent AV surgery. Among patients who underwent AV surgery, inflammation values (Erythrocyte sedimentation rate, 62.07±31.8 mm/hr vs. 39.16±28.4 mm/hr; C-reactive protein, 3.66±4.1 mg/dL vs. 0.92±1.7 mg/dL), AR degree (3.56±0.7 grade vs. 2.08±1.0 grade), and sinus diameter (37.24±5.7 mm vs. 33.22±4.5 mm) were significantly higher than in those who did not undergo AV surgery. Long-term survival and freedom from MACCE were not significantly different between the groups (10 year survival, 84.3% vs. 79.4%; p=0.827; 10 year event-free survival, 51.8% vs. 71.2%; p=0.29). Twelve of the 41 patients who underwent AV surgery had a poor outcome during follow-up (median, 92.5 months; IQR, 54.5–183.5), and eight of them had a recurrence of AR requiring reoperation. Multivariate Cox analysis revealed that coronary disease [hazard ratio (HR), 4.234; 95% confidence interval (CI), 1.381–12.979; p=0.012], LV dysfunction (HR, 3.387; 95% CI, 1.143–10.042; p=0.028), and impaired renal function (HR, 19.983; 95% CI, 3.480–114.731; p=0.001) were significant risk factors associated with poor outcomes at follow-up (table 1).

Abstract AB0705 – Table 1. Multivariate analysis of predictive factors of poor outcomes in patients with Takayasu arteritis who underwent aortic valve surgery.

Variables	HR	95% CI	p-value
Coronary disease*	4.234	1.381–12.979	0.012
LV dysfunction**	3.387	1.143–10.042	0.028
Impaired renal function	19.983	3.480–114.731	0.001

*coronary disease: severity is more than moderate stenosis

**LV dysfunction: EF <50%

Conclusions: In patients with TA with AV involvement, there were no significant differences between long-term survival rate and event-free survival (MACCE) among those who had or had not undergone AV surgery. In the surgical group, the prognosis was poor when coronary artery disease, LV dysfunction, and renal impairment were present at the time of surgery.

Disclosure of Interest: None declared

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AB0706

OCULAR PRESENTATION IN GRANULOMATOSIS WITH POLYANGIITIS (GPA) PATIENTS: RELATION TO AUTOANTIBODIES AND DISEASE ACTIVITY

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Objectives: To study the disease characteristics, autoantibodies and activity in granulomatosis with polyangiitis (GPA) patients with ocular manifestations.

Methods: 46 GPA patients visiting the ophthalmology clinic were included. Ocular manifestations, clinical and slit lamp examination were performed. The Birmingham Vasculitis Activity Score (BVAS) was recorded. Laboratory investigations were recorded and the antineutrophil cytoplasmic antibody (ANCA) performed.

Results: The median age of the patients was 44.5 (32–63) years, 22 males:24 females and disease duration 6.5 (1–16) years. Ocular manifestations were present in all patients; 12 (26.1%) had proptosis, 40 (87%) scleritis/episcleritis with perforation in 3 (6.5%), keratoconjunctivitis in 33 (71.7%) – acute infiltrative stromal keratitis in 11, peripheral ulcerative keratitis in 15 and sclerosing keratitis in 11 patients. Uveitis was present in 11 (23.9%) and retinal changes included vasculitis, exudates and haemorrhage was present in 7 (15.2%). 43 (93.5%) of the patients had blurring of vision and vision loss was present in 2 (4.3%). Glaucoma was present in 4 (8.7%) and hypotony in 2 (4.3%). Involvement was bilateral in 32 (69.6%) patients. Rheumatoid factor was positive in 56.5% and significantly associated with uveitis (p=0.04) while ANA was positive in 45.7% and significantly associated with keratoconjunctivitis (p=0.04). BVAS tended to be higher in those with uveitis (p=0.05).

Conclusions: Ocular involvement must be considered in all GPA patients and referral to an experienced ophthalmologist is mandatory for proper management and improved outcome of such a rare systemic disease. ANA and RF positivity may raise suspicion for KC or uveitis respectively. There was a remarkable association between uveitis and disease activity.