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 criteria, 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 = 15), 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).

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


APR0701

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 severe 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: 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 4–12 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 52, 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) g. 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