Background: Recently, a new set of classification criteria for Giant Cells Arteritis (GCA) and Takayasu Arteritis (TA) has been developed by the DCVAS project and presented as draft criteria at the 19th International Vasculitis and ANCA Workshop held in Philadelphia in 2019.

Objectives: The purpose of the present study is to analyze the performance of the 2019 DCVAS Draft Classification Criteria in differentiating GCA and TA in a cohort of patients with Large Vessel Vasculitis (LVV), comparing their sensitivity and specificity to 1990 ACR Classification Criteria.

Methods: 2019 DCVAS Draft Criteria and 1990 ACR Criteria were retrospectively applied to a cohort of 130 consecutive patients with Large Vessel Vasculitis. In all patients the diagnosis of vasculitis was histologically and/or radiologically isolated.

Results: One-hundred patients had a clinical diagnosis of GCA, 25 patients of TA and 5 patients of other form of LVV, different from GCA and TA (idiopathic isolated aortitis n:2, aortitis with retroperitoneal fibrosis n:2, isolated pulmonary involvement n:2). Among the 100 patients clinically diagnosed as GCA (F:M: 68/32, age: 74 (60-83)) only 82 fulfilled the 1990 ACR Criteria for GCA, while all of them fulfilled the 2019 DCVAS Draft Criteria for GCA.

Instead, among the 25 patients with a clinical diagnosis of TA (F:M: 21/4, age: 34 (16-48), 22 (88%)) could be classified as TA according to the 1990 ACR Criteria, 25 (100%) according to 2019 DCVAS Draft Criteria.

In the group of patients diagnosed with other form of LVV (F:M: 2/3, age: 56 (38-71)) 4 patients (80%) fulfilled the 2019 DCVAS Draft Criteria for GCA, while none of them fulfilled the 2019 DCVAS Draft Criteria for TA or the 1990 ACR Criteria for GCA or TA. One of these patients did not fulfill any classification criteria.

On the contrary, one GCA patient could be classified both as GCA or TA according to the 2019 DCVAS Draft Criteria but didn’t fulfill the 1990 ACR Criteria for GCA or TA.

For GCA, 2019 DCVAS Draft Criteria showed a sensitivity of 100% and a specificity of 80%, compared to 82% and 100% of 1990 ACR Criteria. For TA, 2019 DCVAS Draft Criteria showed a sensitivity of 100% and a specificity of 99%, compared to 88% and 100% of 1990 ACR Criteria.

Table: The new draft classification criteria showed a lower specificity if compared to the older ones, but also a higher sensitivity: in particular 2019 DCVAS Draft Criteria can better identify GCA patients with extracranial involvement, historically excluded from the 1990 ACR criteria. Both GCA and TA 2019 DCVAS Draft Criteria, demonstrating that this classification well performs in differentiating GCA and TA.

Disclosure of Interests: None declared.

References:

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PREVALENCE, BURDEN OF DISEASE AND HEALTHCARE UTILIZATION AMONG PATIENTS WITH EOSINOPHILIC GRANULOMATOSES WITH POLYANGIITIS (EGPA) IN JAPAN 2005-2017

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Background: EGPA is a rare vasculitis condition with very limited data available worldwide. The purpose of this research was to describe HCU and treatment patterns among Japanese EGPA patients.

Methods: This was a retrospective descriptive cohort study using a large administrative claims database covering up to more than 5 million corporate employees and their dependents (MDM claim database) in Japan. Annual prevalence from 2005-2017 was estimated using two EGPA case definitions: a) patients with ≥1 ICD-10 code (2003 version) for EGPA (M30.1), b) patients with ≥2 ICD-10 codes for EGPA (M30.1) during the year in which prevalence was calculated. Among newly identified EGPA patients with no EGPA code in at least 12 months before, clinical burden, comorbidities, after hour visiting (AHV), all cause hospitalization, and treatment with drugs, including oral corticosteroid (OCS) use was described. OCS dose was expressed as prednisone equivalent.

Results: The total number of newly identified patients in 2006-2016 was 45 persons and the mean (SD) age was 42.3 years (SD 14.7 years). The prevalence (per 1,000,000 patients) of EGPA with case definition a) in Japan in 2017 was estimated to be 38.0. The stratified prevalence (per 1,000,000) by age was: 2.3 in the group aged <18 years, 34.0 in those aged 18-59 years, and 91.1 in those aged ≥60 years, respectively. The prevalence in females (50.0) was approximately 1.7-fold higher than that in male (28.7). The prevalence, including stratified results, with definition b) was similar to that with definition a).

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None of newly identified patients had at least one hospitalization and 55.6% had AHV in the year after the first observed EGPA code during the study period. Following index date, new patients were treated: 77.8% with OCS, 11.1% with Methotrexate, 8.9% with intravenous immunoglobulin, 6.7% with Cyclophosphamide, 4.4% with Methylprednisolone, and 2.2% with Rituximab (non mutually exclusive).

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