and ANCA-positive patients, whereas lower percentages of $\text{AST}_\text{Reg}$ were observed in ANCA-positive patients as compared to ANCA-negative patients and HC. Importantly, a significant increase in the percentage of IL-17 and IL-21 cells was seen within the NONTReg from ANCA-positive patients and HC. Importantly, a significant increase in the percentage of IL-17 and IL-21 cells was observed in ANCA-positive patients as compared to ANCA-negative patients and HC.

Conclusion

Increased FoxP3 expression in Th-cells from GPA-patients is related to an increase in a subset of non-suppressive Th-cells. Increased production of IL-17 and IL-21 cytokines in NONTReg cells from ANCA-positive patients, in conjunction with a decrease in their suppressive capacity, points towards their effector function in ANCA production.

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A15  
Performance of the ACR classification criteria as diagnostic criteria for granulomatosis with polyangiitis (GPA) (Wegener’s) in a respiratory referral centre

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Introduction

As no diagnostic criteria for granulomatosis with polyangiitis (GPA) exist, the ACR classification criteria have been used as surrogate for diagnosis. They have a 88% sensitivity (Se) and 92% specificity (Sp). In spite of the high frequency of respiratory manifestations of GPA, and the presence of two items of the respiratory system in the ACR criteria, their evaluation as diagnosis surrogate in a respiratory centre has not, to our knowledge, been reported.

Patients

The ACR classification criteria were applied to consecutive, prospective (in- and outpatients) subjects with suspicion of GPA attending a nationwide respiratory referral centre. Diagnosis was based on clinical, serological and histopathological data. Sample size estimation was calculated for Se of 50–90% and Sp of 75–90% [1]. Se, Sp, positive and negative predictive values (PPV and NPV), positive and negative likelihood ratios (PLR and NLR, respectively) and area under the curve (AUC) were estimated.

Results

Ninety-three patients with clinical suspicion of GPA were included. Thirteen (14%) had the disease. With 2/4 ACR criteria 5e was 31%, Sp 56%, PPV 10%, NPV 83%, PLR 0.69, NLR 1.23 and AUC 0.5.

Discussion

ACR classification criteria for GPA derived from data of cohorts in where patients had already an established diagnosis. Their performance as surrogate for diagnosis is questionable. Most validation studies have been done in rheumatological or nephrological units. Determining their value as diagnostic surrogates in defined settings, with GPA being a multisystemic disease, is useful. We have explored the performance respiratory diseases referral centre prospectively, by including all consecutive patients with GPA suspicion.

Conclusion

In our setting, ACR classification criteria have poor performance when used as diagnostic surrogates. This highlights the relevance of the ongoing DCVAS project for classification and diagnostic criteria of the vasculitides.

Reference


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A16

Use of the CHCC 2012 definitions in the classification of AAV using the EMA algorithm

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Introduction

The classification of the ANCA-associated vasculitides (AAV) (GPA, MPA, EGPA) and PAN remains controversial. The ACR classification criteria whilst validated do not include MPA, whilst the Chapel Hill Consensus Conference (CHCC) definitions promulgated in 1994 were not intended to be used for classification. In 2005, an international consensus group produced the European Medicines Agency (EMA) algorithm for classification of AAV [1]. It combines the ACR and CHCC definitions. In 2012 the CHCC definitions were revised to include updated information in particular the role of ANCA [2]. The EMA algorithm was validated using a cohort of 99 patients with systemic vasculitis from a single-centre [1].

The aim of this study was to reclassify the same 99 patients using the CHCC 2012 definitions to determine whether the new definitions changed the classification of patients with AAV.

Methods

The original dataset from the 99 patients used in the validation of the algorithm was used to reclassify the patients. The reclassification was performed by an independent observer (RA) who was not involved in the original study and was blinded to the diagnosis/previous classification of the patients.

Results

Using the 2012, definitions 59/99 patients were classified with GPA, 22 MPA and 18 EGPA. There were no unclassified patients. Using the 1994 definitions there were 59 GPA, 22 MPA and 18 EGPA. However, one patient changed from MPA to GPA and one GPA to MPA. The kappa statistic was 0.96. The patient who changed from MPA to GPA reflected a difference in the interpretation of the significance of renal involvement. The patient who switched from GPA to MPA reflected a change in the interpretation of infiltrates on chest X-ray.

Discussion

The revision of the CHCC definitions makes no difference to the performance of the EMA algorithm for the classification of AAV.

Conclusion

The EMA algorithm for the classification of AAV is a valid algorithm for classifying patients with AAV using the CHCC 2012 definitions.

References


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