consensus in the final round of the Delphi, a round table discussion between the expert panel members.

Discussion – Until clinical studies provide evidence for all aspects of vasculitis patient care, use of expert opinion is an acceptable alternative. This study will provide guidance for the holistic care of these patients.

Conclusion – Using Delphi methodology we have achieved consensus where previously there was considerable variability in practice. We anticipate publication of these guidelines in summer 2013, routine use of which should assist in improving quality of care for the vasculitis patient and subsequent reduction in morbidity and mortality.

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Estimating renal survival using the ANCA-associated glomerulonephritis classification: A validation study

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Introduction – Recently, a classification for renal biopsies of patients with anti-neutrophil cytoplasmic antibodies associated vasculitis (AAV) was published (i.e. the ANCA-associated glomerulonephritis or AGN classification) [1]. We decided to evaluate this classification.

Patients – ANCA positive patients with a renal biopsy were included. Biopsies were classified using the EUVAS classification into four groups: focal (> 50% glomeruli on the slide appear normal), crescentic (> 50% of glomeruli contain cellular crescents), sclerotic (> 50% of glomeruli are sclerotic) and mixed [1].

Results – Five year renal survival rate according to the AGN classification scheme was: 91% for the focal group, 69% for the mixed group and 64% for the crescentic group (P < 0.0001). One patient was classified as ‘sclerotic’. Importantly, a worse renal survival was found in patients classified as either crescentic or mixed when the percentage of normal glomeruli in the renal biopsy was < 25% (P = 0.04) (figure 1).
Introduction

Granulomatosis with polyangiitis (Wegener’s) (GPA) is a primary systemic vasculitis mainly affecting the ENT system, kidneys and lungs. Patients with ENT disease have a higher incidence of relapse and of a refractory course. There are no accepted serological markers for disease activity and grumbling disease can be difficult to differentiate from infection and damage. Therapies targeting ENT disease require a specific scoring system for ENT disease in GPA. The aim of this study is to explore the reliability of two scoring systems for ENT/GPA disease activity.

Methods—The first instrument, ENTAS [1], designed by experienced ENT clinicians, was tested for reliability by a group of experienced and inexperienced clinicians. The instrument is comprehensive and has a 5 step grading system for disease activity.

The second instrument, ENT/GPA DAS [2] was designed using recognised validity steps in a cohort of 144 patients with GPA. The scoring system is dichotomised and was only designed to measure disease activity.

Cohen’s kappa was used to measure agreement between two assessors and Fleiss’ kappa was used to measure agreement among multiple assessors.

Results—ENTAS was assessed using 50 still images of nasal endoscopies and 12 assessors. In an experienced approach intra- and inter-rater reliability κ values were 0.58/0.72 and 0.62/0.50 (inexperienced/experienced clinicians).

Intra-rater reliability on ENT/GPA DAS was done using 50 video clips and the κ value was 0.48. Inter-rater reliability tests were done using 20 video clips and four assessors and 17 simultaneous assessments by two assessors and the κ values were 0.67 and 0.68 respectively.

Conclusion—The scoring systems showed moderate to substantial agreement and were feasible to score. They will further be amalgamated to produce a single ENT disease activity and damage-scoring instrument.

References


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Towards an international standardized ENT disease activity score in GPA

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Discussion—We confirmed the study by Berden et al. by showing that patients with a renal biopsy classified as focal had the best renal survival. Only one patient could be classified in the “sclerotic” group. Patients who were classified in the “crescentic” group had a worse renal survival compared to patients classified in the “mixed” group. This finding is in contrast to the study by Berden et al. who found a better renal survival in patients classified in the “crescentic” group as compared to the “mixed” group.

Most importantly, patients classified “crescentic” and “mixed” in our study had a worse renal survival when the amount of normal glomeruli was < 25%.

Conclusion—We conclude that the AGN classification for renal biopsies is a practical and informative scheme to classify renal biopsies of AAV patients. In addition, we believe it is important to add the specific percentage of normal glomeruli to the AGN classification group to optimize the predictive value of the classification scheme.

Reference


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Cardiovascular outcomes and prognostic predictors in ANCA-associated vasculitis

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Introduction—ANCA-associated vasculitis (AAV) [granulomatosis with polyangiitis (GPA), and microscopic polyangiitis (MPA)] is associated with an increased frequency of cardiovascular events (CVE).

Objectives—To characterize cardiovascular outcomes and predictors, including the role of vasculitis therapies, for CVE/death in AAV.

Methods—A single center retrospective review of 307 AAV patients [173 GPA, 134 MPA, 47% male, 12% diabetic, mean age 53 (+/-7) years with follow-up 6.1 (+/-5.3) years] The primary end-point was CVE (defined as acute coronary syndrome, new onset angina, symptomatic peripheral vascular disease, stroke or transient ischaemic attack), or death.

Results—Fifty-one CVE occurred in 42 patients (13.6%) with 28 (9%) deaths; 15.7% CVE occurred at the onset of AAV, 57.4% CVE/death occurred within first year of AAV diagnosis, and 27.8% between 1–5 years. Independent predictors for the end-point (CVE/death) were: maintenance prednisolone dose [hazard ratio (HR) 169.6

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