A52

Large vessel vasculitis: Estimating disease activity in patients with thoracic aortic aneurysms

A. Clifford1, T. Clark1, D. Johnston2, G. Petterson2, E. Roses2, E. Soltesz2, L. Svensson2, G. Hoffman1

1. Center for Vasculitis Care and Research, Cleveland Clinic Foundation, Cleveland, USA
2. Thoracic and Cardiovascular Surgery, Cleveland Clinic Foundation, Cleveland, USA

Introduction.– Thoracic aortic aneurysms due to large vessel vasculitis may be a sign of ongoing inflammation or the sequelae of previous damage to the arterial wall. Ongoing inflammation in the aorta is usually estimated indirectly using clinical features, inflammatory markers and imaging characteristics. Whether these surrogates accurately reflect inflammation within the vessel wall is not known.

Methods.– We retrospectively reviewed the clinical, laboratory and imaging data for patients in whom aortitis was historically demonstrated at the time of reconstruction for thoracic aortic aneurysms (May 2011–December 2012). ACR classification criteria were used to define vasculitis subtype. Pre- and postoperative assessments of disease activity were estimated over time using predefined criteria.

Results.– Among 126 consenting study patients who underwent repair of thoracic aortic aneurysms, 14 (11%) had histological features of aortitis. Thirteen specimens had active inflammatory infiltrates, and one had features of healed aortitis. Seven patients met criteria for Giant Cell Arteritis, two for Takayasu’s arteritis, and five for focal isolated aortitis. Median duration of disease prior to surgery was 35.5 months (range 0–180). At the time of surgery, median ESR and CRP were 11 mm/hr (range 2–38) and 1.0 mg/dl (range 0.2–1.7), respectively, and angiograms revealed new lesions in 3/14 patients. Nine patients (64%) were judged to be in clinical remission, in three the degree of disease activity was uncertain, and two met criteria for active disease. Seven patients were treated with prednisone within 12 weeks of surgery [median dose = 20 mg/d (range 2–55 mg)].

Discussion.– About 2/3 of patients found to have aortitis do not have clinical, laboratory or imaging indications of active disease at the time of surgery.

Conclusion.– Indirect estimates of thoracic aortic inflammation are insensitive in patients with aortitis.

Further reading

http://dx.doi.org/10.1016/j.jrpm.2013.02.054

A53

Assessment of disease activity in large vessel vasculitis: Initial results of an international Delphi exercise

S.Z. Aydin1, H. Direshkeneli1, E. Matteson2, P.A. Merkel3

1. Istanbul, Turkey
2. Rochester, USA
3. Pennsylvania, USA

Introduction – Inflammatory vasculitides may be systemic or limited to large or medium-sized vessels. Large vessel vasculitis affects patients mainly older than 50 years. The aorta is most frequently involved, and may be symptomatic in the majority of cases. Cardiovascular events are common in large vasculitis, and are due to aortic insufficiency, ischemia, or mural thrombus. Several studies show that the risk of cardiovascular events increases with disease activity. However, there is no widely accepted method for disease activity assessment in large vessel vasculitis.

Objective.– This study aimed to develop an international consensus-based disease activity (DA) assessment tool in large vessel vasculitis.

Methods.– An international Delphi process was conducted to assess the existing evidence, and to develop a consensus tool for DA assessment in large vessel vasculitis. The study was divided into two rounds. In round one, all experts evaluated the evidence regarding current markers of disease activity and the ideal DA assessment tool. In round two, experts were asked to rate the importance of developing a consensus tool for DA assessment.

Results.– In round one, experts assessed that markers of disease activity were not validated, and that an ideal DA assessment tool did not exist. In round two, the importance of developing such a tool was widely supported. A consensus tool for DA assessment in large vessel vasculitis was developed. The tool included clinical, laboratory and imaging markers of disease activity.

Conclusion.– The consensus tool for DA assessment in large vessel vasculitis was developed. This tool may help to improve the management of patients with large vessel vasculitis.

http://dx.doi.org/10.1016/j.jrpm.2013.02.053
Introduction. Assessment of disease activity in large vessel vasculitis (LVV) is challenging. To develop well-accepted and validated outcome measures for use in clinical research in LVV, a Delphi exercise has been launched as part of an international initiative to systemically arrive at consensus expert opinion and identify candidate outcomes for assessment of disease activity in LVV.

Methods. The Delphi survey was sent to > 300 experts in LVV from different specialties. The first round included 99 items on a 5-point scale aiming to cover potential disease manifestations. Items accepted or rejected by > 70% of voters are not advanced to subsequent rounds.

Results. One hundred and sixteen experts from 23 countries completed the survey. Most vascular/cardiovascular items were accepted by > 70% of experts for Takayasu’s (TAK); ocular findings were considered high-priority outcomes for GCA. Vascular imaging was accepted for both TAK and giant cell arteritis (GCA). SF36 and patient global assessment were widely accepted in both diseases. Del. Tak was the only composite index accepted for TAK. Only ESR and CRP were suggested as biomarkers in TAK; hemoglobin level was also supported in GCA. Findings rejected by > 70% of the experts included erythema nodosum for TAK and pulmonary assessments for GCA. Additional items proposed by participants (e.g. IL-6 levels and novel PRO measures), and the items that did not reach the 70% threshold, will be considered in subsequent rounds. 63% of experts voted to have a common approach for both TAK and GCA but to also develop additional specific instruments for each disease; 25% felt the two diseases were unsuitable for common outcome measures.

Discussion. This exercise points out to similarities and differences from experts’ perspective for assessing the clinical activity in TAK and GCA.

Conclusion. Based on the Delphi, it is anticipated that a consensus-driven set of outcomes will include many data elements common to both diseases, supplemented by disease-specific items for TAK and GCA.

http://dx.doi.org/10.1016/j.lpm.2013.02.055

A54

Outcome and prognostic factors in elderly patients with ANCA-associated vasculitis

M. Weiner1, S.M. Goh2, A. Mohammadi3, Z. Hrusková4, A. Tanna5, A. Bruchfeld6, D. Selga7, Q. Ko7, K. Westman7, P. Eriksson8, C. Pusey9, V. Tesar4, A. Salama2, M. Segelmark1
1. Department of Medical and Health Sciences, Linköping University, Linköping, Sweden
2. UCI Centre for Nephrology, Royal Free Hospital, London, United Kingdom
3. Department of Rheumatology, Skåne University Hospital, Lund, Sweden
4. Department of Nephrology, 1st Faculty of Medicine, Charles University, Prague, Czech Republic
5. Imperial College Renal and Transplant Centre, Hammersmith Hospital, London, United Kingdom
6. Department of Renal Medicine, Karolinska University Hospital, Huddinge, Sweden
7. Department of Nephrology and Transplantation, Skåne University Hospital and Lund University, Lund, Sweden
8. Department of Rheumatology, County Council of Östergötland and Linköping University, Linköping, Sweden
9. Imperial College Renal and Transplant Centre, Hammersmith Hospital, Linköping, United Kingdom

Introduction. Outcome among elderly patients with ANCA-associated vasculitis (AAV) is largely unknown. In this multicenter study we investigated prognostic factors for outcome of patients ≥ 75 years of age.

Patients. Consecutive patients from six centers in Sweden (n = 78), England (n = 37) and Czech Republic (n = 25) diagnosed 1997-2009 were included. Inclusion criteria were a clinical diagnosis of AAV and age ≥ 75 years at diagnosis. The patients were followed for up to two years from diagnosis. Sustained remission was counted starting on day 90. The grade of comorbidity was assessed using a modified version of the Davies index.

Results. One hundred and fifty patients (M:F; 75:75) were included. Median age at diagnosis was 79 years (IQR77-82). 69% were diagnosed with MPA and 31% with GPA. Sixty-two percent were positive for MPO-ANCA, 36% for PR3-ANCA and 2% were ANCA-negative. Mean value for BVAS at diagnosis was 16 (SD 5.7) and median plasma creatinine 283 μmol/L (IQR152-483). Data on renal function and survival were available for 137 patients and regarding sustained remission for 114 patients. One-year patient survival was 70% and 2-year survival was 63%. In multivariable Cox regression analysis including sex, age, comorbidity grade, ANCA, BVAS and creatinine we found that older age (P = 0.02) and low BVAS score (P = 0.01) were associated with increased mortality. Twenty-five percent of the patients started chronic renal replacement therapy, all during the first year. Renal survival censored for death was 74% after 2 years. Nine percent of the patients suffered a relapse during follow-up.

Conclusion. In this large retrospective study of elderly patients with AAV we found that 30% died and 25% had started dialysis after 1 year. Among the 60 patients who were alive in dialysis-free remission after 1 year, prognosis was good with no development of end-stage renal disease and only five relapses and three deaths during the second year. The results raise questions regarding the need for and extent of long-term maintenance therapy in elderly patients.

http://dx.doi.org/10.1016/j.lpm.2013.02.056

A55

Factors associated with major cardiovascular events in patients with primary systemic necrotizing vasculitides: Results of a longitudinal long-term follow-up study

B. Terrier1, C. Pagnoux1, G. Chironi2, A. Simon2, L. Mouchon1, L. Guillemin1
1. Cochin, Paris, France
2. HEGP, Paris, France

Introduction. Primary systemic necrotizing vasculitides (SNV) were shown to be associated with more frequent subclinical atherosclerosis, independently of cardiovascular (CV) risk factors and C-reactive protein (CRP) level, suggesting that SNV might be associated with a higher risk of major CV events (MCVE).

Objective. To identify factors predictive of MCVE in SNV patients.

Methods. Consecutive patients in SNV remission were assessed for CV risk factors and subclinical atherosclerosis, and prospectively followed in the same center. High-risk status, defined according to the NCEP/ATP III, was a known history of CV disease, diabetes or 10-yr Framingham Risk Score ≥ 20%. MCVE, defined as myocardial infarction, stroke, arterial revascularization, hospitalization for unstable angina and/or death from CV causes, were recorded. Kaplan–Meier MCVE-free survival curves were plotted and compared with the log-rank test.

Results. Thirty-seven patients (24 males, age 54 ± 15 yr) were followed for 7.0 ± 2.6 yr. SNV diagnoses were: GPA, 19; EGPA, eight; MPA, seven; and PAN, three. Seven (18.9%) patients suffered MCVE. The respective 5- and 10-yr MCVE rates were 10.8% and 25.7%.

Univariate analysis selected NCEP/ATP III-defined high-risk status (HR 5.02, P = 0.03), BMI > 30 kg/m² (HR 4.84, P = 0.02) and plaque...