EULAR update

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Abstract sessions

Angiogenesis and blood vessel stability in Giant Cell Arteritis

D. Molloy et al [OP0179] Ann Rheum Dis 2012;71(suppl 3):115

- Pathogenesis unknown but involves vascular remodeling, inflammatory infiltrate arterial wall
- Aim: assess blood vessel stability and oxidative damage in GCA patients and correlation with disease activity
- 20 patients with GCA included (16 positive temporal artery biopsy)
- Results: unstable inflamed vessels in GCA, associated with EC/pericyte interaction, expression of Ang2 and oxidative damage markers

TLR4 and VEGF polymorphism in chronic periaortitis

F. Atzeni et al. [OP0180] Ann rheum dis 2012;71(suppl3):115

- Chronic periaortitis (CP) is a rare disease
- TLR4 and VEGF are associated with inflammatory diseases
- 102 CP and 200 healthy controls genotyping
- No difference TLR4 allele frequency or genotype in CP or healthy controls
- Polymorphisms in (II+ID) VEGF associated with increased risk ureteral obstruction (heterozygote) and thrombosis (homozygote)

Retrospective monocentric cohort of orbital masses in granulomatosis

J. Holle, et al [OP0184] Ann Rheum Dis 2012;71(suppl 3):117

- 1142 GPA patients (1988-2011)
- 53 orbital mass, 40 complete dataset
- 1/3 bilateral involvement, 70% origin paranasal sinuses
- Clinical: diplopia, orbital pain, propoptosis, reduced eye motility, reduced vision
- Most frequent on patients without systemic vasculitis symptoms
- Refractory disease in 40%

Burden of childhood CNS vasculitis: identifying high risk factors for poor cognitive outcome

P Gowdie et al. [OP0277] Ann Rheum Dis 2012;71(suppl 3):150

- 111 children with childhood cns-vasculitis
- 70 completed neurocognitive evaluation
- 53% of small vessel disease and 27% of large vessel disease are cognitive impaired (FSIQ<85)
- Risk factors: seizures

General sessions

News in vasculitis

D. Jayne.

- Incidence AAV in Europe stable
- First GWAS in AAV (Chr 6, serpina 1, PRTN3)
- 2 new antibodies: hLAMP-2 and anti-plasminogen antibodies
- Role of complement in AAV (CLEAR study)
- NORAM 5 year results: more steroids in MTX than CYC arm
- Rituximab in AAV (still not clear maintenance/induction)
- Mepolizumab for EGPA (CSS)

Chapel Hill 2012 Classification

N. Rasmussen. [SP0065] Ann Rheum Dis 2012;71 (suppl3):16

- Large Vessel Vasculitis (LVV)
- Medium Vessel Vasculitis (MVV)
- Small Vessel Vasculitis (SVV)
- Variable Vessel Vasculitis (VVV)
- Single Organ Vasculitis (SOV)
- Vasculitis Associated with Systemic Disease
- Vasculitis with Probable Etiology

Chapel Hill 2012 Classification

- Large Vessel Vasculitis (LVV)
  - Takayasu Arteritis (TAK)
  - Giant Cell Arteritis (GCA)
- Medium Vessel Vasculitis (MVV)
- Small Vessel Vasculitis (SVV)
- Variable Vessel Vasculitis (VVV)
- Single Organ Vasculitis (SOV)
- Vasculitis Associated with Systemic Disease
- Vasculitis with Probable Etiology

Chapel Hill 2012 Classification

- Large Vessel Vasculitis (LVV)
- Medium Vessel Vasculitis (MVV)
  - Polyarteritis Nodosa (PAN)
  - Kawasaki Disease (KD)
- Small Vessel Vasculitis (SVV)
- Variable Vessel Vasculitis (VVV)
- Single Organ Vasculitis (SOV)
- Vasculitis Associated with Systemic Disease
- Vasculitis with Probable Etiology

Chapel Hill 2012 Classification

- Large Vessel Vasculitis (LVV)
- Medium Vessel Vasculitis (MVV)
- Small Vessel Vasculitis (SVV)
  - AAV
    - Microscopic Polyangiitis (MPA)
    - Granulomatosis with Polyangiitis (GPA)
    - Eosinophilic Granulomatosis with Polyangiitis (EGPA)
- Variable Vessel Vasculitis (VVV)
- Single Organ Vasculitis (SOV)
- Vasculitis Associated with Systemic Disease
- Vasculitis with Probable Etiology

Chapel Hill 2012 Classification

- Large Vessel Vasculitis (LVV)
- Medium Vessel Vasculitis (MVV)
- Small Vessel Vasculitis (SVV)
  - Immune complex SVV
    - Anti-GBM disease
    - Cryoglobulinemic Vasculitis
    - IgA Vasculitis (HSP)
    - Hypocomplementemic Urticarial Vasculitis
- Variable Vessel Vasculitis (VVV)
- Single Organ Vasculitis (SOV)
- Vasculitis Associated with Systemic Disease
- Vasculitis with Probable Etiology

Chapel Hill 2012 Classification

- Large Vessel Vasculitis (LVV)
- Medium Vessel Vasculitis (MVV)
- Small Vessel Vasculitis (SVV)
- Variable Vessel Vasculitis (VVV)
  - Behçet Disease (BD)
  - Cogan's Syndrome
- Single Organ Vasculitis (SOV)
- Vasculitis Associated with Systemic Disease
- Vasculitis with Probable Etiology

Chapel Hill 2012 Classification

- Large Vessel Vasculitis (LVV)
- Medium Vessel Vasculitis (MVV)
- Small Vessel Vasculitis (SVV)
- Variable Vessel Vasculitis (VVV)
- Single Organ Vasculitis (SOV)
  - Cutaneous Leukocytoclastic Angiitis
  - Cutaneous Arteritis
  - Primary Angiitis of the CNS (PACNS)
  - Isolated Aortitis
- Vasculitis Associated with Systemic Disease
- Vasculitis with Probable Etiology

Chapel Hill 2012 Classification

- Large Vessel Vasculitis (LVV)
- Medium Vessel Vasculitis (MVV)
- Small Vessel Vasculitis (SVV)
- Variable Vessel Vasculitis (VVV)
- Single Organ Vasculitis (SOV)
- Vasculitis Associated with Systemic Disease
  - Lupus vasculitis
  - Rheumatoid vasculitis
  - Sarcoid vasculitis
- Vasculitis with Probable Etiology

Chapel Hill 2012 Classification

- Large Vessel Vasculitis (LVV)
- Medium Vessel Vasculitis (MVV)
- Small Vessel Vasculitis (SVV)
- Variable Vessel Vasculitis (VVV)
- Single Organ Vasculitis (SOV)
- Vasculitis Associated with Systemic Disease
- Vasculitis with Probable Etiology
  - HCV-associated cryoglobulinemic vasculitis
  - Drug-associated immune complex vasculitis
  - Drug-associated ANCA-associated Vasculitis
  - Cancer associated vasculitis

Classification and diagnosis of vasculitis

R. Luqmani. [SP0066] Ann Rheum Dis 2012;71 (suppl3):17

- DCVAS study undertaken at the moment in > 40 centers and plan to include > 2000 patients
- Currently 900 included
- ongoing

Long-term outcomes in systemic vasculitis

K. Westman. [SP0067] Ann Rheum Dis 2012;71 (suppl 3): 17

- Long-term follow-up of the first 4 RCTs of EUVAS
- 535 patients (281 GPA, 254 MPA)
- 46% female, 53% PR3-ANCA, median BVAS 17
- Overall mortality higher than age matched population (2.6)
- Highest death risk in 1st year (active vasculitis or infection)
- Predictors mortality: older age at onset, renal insufficienty, higher BVAS

Poster sessions selection

Serum angiopoietin-2 level strongly reflects the disease activity and renal function in AAV

Y Wada et al [THU0198] Ann Rheum Dis 2012;71 (suppl3):222

- Ang-2 key mediator of endothelial cell activation
- 59 AAV patients (MPA 27, GPA 15, EGPA 14, other 3)
- ANG-2 correlated with high BVAS, CRP, serum creatinine, urinary protein excretion and negatively correlated with est GFR

Cross-sectional assessment of damage in Takayasu arteritis with a validated tool

A. Omma et al. [THU0203] Ann Rheum Dis 2012;71 (suppl3):224

- 103 TAK patients f-up > 6 months
- Vascular damage index used for damage of TAK
- VDI scores in TAK patients identical to systemic necrotizing vasculitis patients
- Longer disease duration, higher GC and CY exposure correlated with VDI

Efficacy of anti-TNF therapy in 15 patients with refractory takayasu's arteritis: long term unicentric follow-up

E. Thombetti et al. [THU0209] Ann Rheum Dis 2012;71 (suppl3):226

- 15 TAK patients (2004-2011)
- Mean follow-up 46 months
- 13 infliximab, 5 adalimumab, 1 golimumab
- Reduction of prednisone, CRP, ESR during follow-up
- Revascularisation procedure lesions worsened during treatment while naïve lesions improved
- Possible different pathophysiology revascularization treated and naïve lesions

Differences in clinical presentation and outcome in patients with early versus late onset Giant cell arteritis: analysis of 94 patients

M. Alba et al. [THU0222] Ann Rheum Dis 2012;71 (suppl3): 230

- 170 patients with GCA (1995-2005) biopsy proven
- 94 met inclusion criteria
- 3 groups < 67 (n=16), 68-80 (n=57), > 81 (n=21)
- Early onset: more fever, high ESR
- Late onset: high frequency amaurosis fugax and blindness
- Less relapse in late onset group

Successful maintenance treatment of granulomatosis with polyangiitis with rituximab- a case series

A. Knight et al. [AB0751] Ann Rheum Dis 2012;71 (suppl 3):681

- Retrospective study of 11 patients
- All PR3 positive, previously treated with different immunosuppressant including CYC and GC.
- Median of 4 (2-11) Rituximab infusion for relapse
- Median follow-up 18 months after 1 infusion (range 9-96)
- No relapses during follow-up 7 pt in remission (BVAS 0 and GC < 7.5)
- Well-tolerated, but 7 infections requiring antibiotics during follow-up (1 *Pneumocystis Jiroveci*)