**Conclusion.**—Increasing awareness of this condition amongst clinicians will help with prompt diagnosis and therapy preventing thus complications related to fibrotic phase of the disease.

**Reference**


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**P140**

Silica binds to LL-37 and CpG DNA complexes

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**Introduction.**—Exposure to silica has been implicated in a number of inflammatory conditions affecting the respiratory system including the development of ANCA associated vasculitis. However, the exact mechanism behind the immunostimulatory properties of Silica is not totally understood. It is known that in the presence of highly cationic molecules, silica is capable of binding DNA oligonucleotides. Given that activated neutrophils are capable of generating the cationic peptide LL-37, which can complex with DNA, we wondered if these complexes could bind to silica.

**Methods.**—The effect of silica on immune cells was studied using Whole Blood and/or isolated PBMC cultured in the presence of silica particles (50 μg mL⁻¹) of 10–20 nm (Sigma Aldrich), previously treated with different combinations of LL-37 (4 μg mL⁻¹) and CpGB-FITC (1.6 μg mL⁻¹) i.e. alone or forming LL-37-CpGB-FITC complexes. The expression of activation markers were measured by flow cytometry and gene expression by RT-PCR. Silica particle-CpG-FITC complex were visualized study by confocal microscopy.

**Results.**—We found that immunostimulatory CpG oligonucleotides alone do not bind silica, but LL37-CpG complexes do. Furthermore we show that LL37 alone binds to silica particles, resulting in a LL37-Silica complex that is capable of binding CpG oligonucleotides. While silica particles are normally endocytosed by phagocytic cells, we found that silica coated with CpGB complexes are recognized in addition by B cells, leading to an increase in their expression of MHC-Class II and co-stimulatory molecules CD40 and CD86. We also found that silica can also induce de novo synthesis of ANCA antigens such as PR3 and MPO in PBMCs and therefore our results suggest that exposure to silica particles during a bacterial infection could potentially contribute to an overall amplification of the inflammatory response and production of ANCA autoantibodies.

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**P141**

Aneurysms in the view of rheumatology perspective

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**Introduction.**—An aneurysm is an abnormal widening or ballooning of a portion of an artery due to weakness in the wall of the blood vessel. The aneurysms might a presenting finding or occur during the course of a disease. Although it is not clear exactly what causes aneurysms, in case of rheumatic symptoms and signs, patients should be evaluated regarding Behçet’s disease (BD) and large vessel vasculitis etc. This study is aimed to investigate the rheumatologic diagnosis and vessel involvements of the patients consulted to rheumatology department with probable diagnosis of aneurysms.

**Methods.**—Database of our university hospital between 2000–2012 was used for this study. The patients had been consulted to rheumatology department with a probable diagnosis of aneurysm regarding rheumatic disease were evaluated.

**Results.**—Totally 651 patients were included to this study. In 128 (19.7%) of the patients, a rheumatic diagnosis was found and an aneurysm was found in 56 (43.8%) of them. The most frequent rheumatic diseases were BD (n = 27), Takayasu arteritis (n = 10), polyarteritis nodosa (PAN) (n = 4) and giant cell arteritis (n = 2). Regarding these, 43 patients distribution of the vessel involvements were aorcs aorta and branches 27.9%, pulmonary artery 18.6%, abdominal aorta, 27.9%, cerebral artery 11.6%, peripheral artery 9.3%, renal and/or hepatic and/or splenic artery 9.3%. Fourteen patients had other rheumatic diseases (six SLE, four RA). There were four RA patients with aneurisms and one patient uncontrolled hypertension was accepted as the cause of aneurism. In 2 SLE patients with infective endocarditis, mycotic aneurisms were found.

**Discussion.**—This study highlights the presence of aneurysms in inflammatory rheumatic diseases and emphasizes the Behçet’s disease, large vessel vasculitis and PAN as a cause in Turkish population. It should be kept in mind than aneurysms might occur during other rheumatic diseases such as RA and SLE because of hypertension and infectious complications.

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**P142**

IgA associated vasculitis (IgAV) – a 5-year retrospective study

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**Introduction.**—To review the clinical presentation, potential precipitating factors and outcome of patients with leukocytoclastic vasculitis (LCV) and IgA deposition (IgAV).

**Methods.**—Retrospective search of all cases of LCV diagnosed by skin biopsy in 2007–2012. Of 213 cases, 100 had direct immunofluorescence (DIF), 32% being positive for IgA. We reviewed the charts of all 32 patients and recorded demographics, clinical and laboratory data, duration of follow-up, treatment and outcome.

**Results.**—Of the 32 patients with IgAV: 91% were white, 60.6%-male, all but one patient (aged 16) were adults, median age 57 (16–76). On DIF 59% had IgA deposition alone, with IgM 25%, with IgG 6%, all three 9% in various combinations with complement and fibrinogen. With one exception, all patients had vascular or perivascular IgA; one patient with dermoepithelial IgA, M and G deposition was later diagnosed with systemic lupus erythematosus. Clinical presentation: 16% purpura, 35.5% purpura and arthritis, 29.5% combinations of purpura, arthritis, renal and gastrointestinal (GI) manifestations. Renal involvement was noted in 35% of IgA only and 41.6% of IgA + IgM patients. In the month prior to diagnosis, a bacterial/viral infection was reported in 61%, 48% received antibiotic therapy, a specific organism being identified in 25.8%. Serum IgA was elevated in 45% of 11 patients, cryoglobulins in 42% of 19 patients, median 87 (54–642). Median follow up was 22 (0–66) months, 29% received no treatment, 71% glucocorticoid taper ± other drugs. Outcome: 68% resolved, 16% persistent course, 9.7% unknown, 3% end stage renal disease secondary to IgAV, 3% died due to IgAV related GI complications.
Discussion.— We found IgAV present in one third of all DIF+ LCV cases, often in the context of an infection. The prognosis is favourable in most cases.

Conclusion.— The high frequency of bacterial infections seen before disease onset in this cohort support that further investigation may be warranted into whether a link exists between infection and adult IgAV.

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P143
Macrophage activation syndrome in vasculitis: An underrecognised complication?
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Introduction.— Macrophage activation syndrome (MAS) is potentially fatal in rheumatic diseases. We retrospectively studied the clinical picture, treatment and outcome of MAS in vasculitis patients seen in a referral rheumatology unit in Transylvania, Romania.

Methods.— The charts of vasculitis patients admitted or seen ambulatorily between 2000 and 2012 were reviewed. MAS was defined according to Ravelli preliminary criteria for juvenile idiopathic arthritis.

Results.— We identified four MAS cases, three in previously diagnosed vasculitis (microscopic polyangiitis with polychondritis, granulomatosis with polyangiitis, undifferentiated vasculitis) and one inaugural of Behçet’s disease. In all cases, high fever, lethargy, pancytopenia, elevated transaminases and hyposodemia were noted. Ferritin was elevated in the three cases available. Hepatosplenomegaly and/or adenomegaly were seen in all cases. Bone marrow showed hemophagocytosis in two of three cases performed. Coagulation abnormalities were noted in three patients, along with antiphospholipid syndrome and transverse myelitis in the granulomatosis case. The trigger was infectious in two patients (pneumonia with S. aureus and E. coli).

Methylprednisolone pulse therapy, cyclosporin, broad spectrum antibiotics, life support, transfusions, anticoagulation were employed, successful in two cases.

Discussion.— MAS can be inaugural in vasculitis and may mimick a drug-induced complication or a hematologic malignancy. Therapy cessation, surgery and infections are precipitating factors. Low ESR can be misleading.

Conclusion.— MAS is rare, but life-threatening in vasculitis. A high index of suspicion and prompt therapy are necessary for outcome improvement.

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P144
Retroperitoneal fibrosis at the onset of ANCA associated vasculitis: The risk of a delay in the diagnosis of vasculitis
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Introduction.— Retroperitoneal fibrosis (RPF) is characterised by the development of fibroserotic tissue in the retroperitoneum which leads to encasement of ureters and blood vessels. It is usually “idiopathic”, rarely it’s due to drugs, malignancies, infections, or autoimmune diseases including ANCA associated vasculitis (AAV).

Results.— We describe three cases of AAV with RPF as initial manifestation misinterpreted as idiopathic.

Three male patients (Pt1 42 yo; Pt2 49 yo, Pt3 59 yo) were admitted with back pain, deep vein thromboses (DVT) and monolateral hydronephrosis (Pt1); with DVT, bilateral hydronephrosis and acute renal failure (ARF) (Pt2); monolateral hydronephrosis and ARF (Pt3).

In Pt1 and 2, abdominal CT scan and MRI revealed homogeneous dense fibrous tissue around the aorta, with inferior vena cava and iliac veins compression and thrombosis and monolateral/bilateral ureteral involvement. In Pt3 renal US revealed monolateral hydronephrosis, he was submitted to ureteral stenting, no further evaluations were planned.

On the basis of diagnosis of idiopathic RPF, all pts started oral glucocorticoid therapy at mean doses of 0.5 mg/kg/day with symptomatic relief; in Pt2 obstructive ARF resolved. After 2 and 6 months respectively, Pts1 and 2 developed AAV overt manifestations. Pt 1: RPGN, multineuritis and a wide lung consolidation; Pt2 developed RPGN, multineuritis, cutaneous purpura and episcleritis. Pt1 died in acute phase because of diffuse lung hemorrhage favoured by chronic anticoagulation prescribed at time of DVT.

In Pt3, ARF didn’t improve and he started chronic hemodialysis. He was maintained on low dose steroids therapy for long time. Seven years later, while still on chronic dialysis, he developed persistent fever, malaise, arthralgias. CT scan and MRI revealed a RPF with DVT.

In all three pts, ANCA resulted highly positive with anti PR3 specificity.

Conclusion.— RPF can be the initial manifestation of AAV. Clinical manifestation of AAV (RPGN and other vasculitis related signs) can be hidden by RPF manifestations and by steroids therapy given to treat “idiopathic RPF”. RPF at the onset of AAV entails the risk of a delay in AAV diagnosis and treatment.

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Further readings