**Results.**—Thyroid disease was found in 44 of 181 patients with AAV (24.3%) and 15 of 202 controls (7.4%) \( (P = 0.0001, \chi^2\)-squared). 35/44 (79.5%) had treated hypothyroidism, 5/44 (11.4%) were treated for hyperthyroidism with either propylthiouracil or radio-iodine and 4/44 (9.1%) had transiently abnormal thyroid function tests thought to be thyroiditis. In the control group 8/15 (53.3%) had treated hypothyroidism, 2/15 (13.3%) were treated for hyperthyroidism, 2/15 had multinodular goitre and 1/15 had thyroid malignancy. More female patients had a diagnosis of thyroid disease in both AAV (72.7%) and control groups (66.7%). AAV patients with thyroid disease more commonly had anti myeloperoxidase (MPO) antibodies compared to the group without thyroid disease (56.8% vs. 40.1%, \( P = 0.0527, \chi^2\)-squared). Thyroid peroxidase (TPO) antibodies were positive in 5/19 (26.3%) of AAV patients and 0/3 control patients.

**Discussion.**—There is an association between thyroid disease and AAV with greater incidence of thyroid disease in those with anti-MPO antibodies. This could be due to cross-reactivity between anti-MPO and anti-TPO antibodies.

**Conclusion.**—In view of this association we suggest that patients diagnosed with AAV should be tested for concomitant thyroid disease.

**Further readings**

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P138

**Single-organ gallbladder vasculitis: Characterization and distinction from systemic vasculitis involving the gallbladder. An analysis of 57 patients**

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**Introduction.**—Systemic vasculitis involving abdominal structures usually has a poor prognosis. Gallbladder (GB) vasculitis (GV) has been reported in systemic vasculitis (SVGV) and as focal single-organ/isolated GV (IGV). We analyzed clinical and histologic characteristics of patients (pts) with GV in order to identify features that differentiate IGV from SBV.

**Methods.**—Pathology databases from our institution and a PubMed search were used to identify pts with GV. Clinical, laboratory, histologic features, therapies and outcomes were recorded. Patients were divided in IGV and SGV. IGV required isolated extent confirmation after a follow-up period of at least 6 ms.

**Results.**—Fifty-seven pts with GV were included (29F/28 M), 6 from our institution. 44% presented with gall stone associated cholecystitis (GSAC) or chronic cholecystitis and 44% with acalculous cholecystitis. GV was found in 20 (35%) and SGV in 37 (65%) of pts. No age or sex differences were observed. GSAC tended to occur more frequent in IGV pts, who also suffer more often from recurrent abdominal pain (53% vs. 17%; \( P = 0.01 \)). Fever was present in 20% of IGV pts and constitutional/musculoskeletal symptoms occurred only in SGV pts (in 50%). ESR was higher in SGV, without differences in Hgb or leucocyte count. Only 3 IGV pts received steroids, whereas all SGV pts were treated and 50% also received cytotoxic agents. 2 IGV pts died from unrelated conditions, and nine SGV pts died from disease activity complications or infections. Non-granulomatous inflammation with fibrinoid necrosis of medium-sized vessels occurred in 93% of both groups. SBV most often reported were PAN (\( n = 9 \)), HBV-associated vasculitis (7), cryoglobulinemic (essential or HCV-associated) vasculitis (6), EGPA (4) and MPA (4).

**Conclusion.**—IGV is uncommon and most often presents after recurrent episodes of abdominal pain, without systemic symptoms, normal ESR, and does not require systemic therapy. PAN and HBV and HCV-associated vasculitis are the most frequent SGV forms. GV is associated with high mortality.

**Further readings**

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P139

**Weight loss – a common presentation in a rare disease. IgG4 related disease (IgG4 RD) is an emerging diagnosis not to miss**

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**Introduction.**—IgG4 RD remains difficult to diagnose not only because of the continued lack of familiarity with this disease by many physicians but also because the presenting symptoms are highly variable. We report a case of IgG4 RD presented with simultaneous multiorgan involvement that took 12 months to diagnose from the onset of the disease.

**Methods.**—A 26-years-old man lost 10 kg in weight over a period of 6 months. He also complained of a cough associated with night sweats. The X-ray chest showed reticular shadowing in keeping with pneumonitis. Furthermore, a tender cervical lymph node, smooth splenomegaly and tenderness over the left renal angle were detected.

**Results.**—Blood tests revealed eosinophilia 1.48, Cr 161, eGFR 45 ml/min with negative immunology but raised ESR 132, CRP 21 and raised IgG 45 and IgE 284. Further tests excluded underlying TB, HIV, sarcoidosis and hepatitis B &C. Staging CT scan revealed enlarged left kidney 21 cm, splenomegaly 15 cm, and coeliac axis lymphadenopathy. Initial biopsies of lymph node, bone marrow and left kidney were all inconclusive but subsequent immunostaining of renal biopsy confirmed a large number of IgG4 (> 50/HPF). IgG subtype analysis showed high IgG4 50.3 (< 1.30) supportive of IgG4 RD diagnosis. A tapering regimen of oral glucocorticosteroids (GC) was introduced; within 2 weeks from starting GC the size of left kidney reduced to 15.3 cm and spleen to 11.4 cm. Furthermore, ESR, eosinophils, IgG all normalised and Cr stayed at 112. In view of hypertension, ECHO was performed and showed a degree of left ventricular hypertrophy; it is unclear whether this is due to possible underlying lymphoplasmatocytic heart infiltration or renal impairment.

**Discussion.**—The diagnosis of IgG4 RD continues to be challenging as it requires an integration of all clinical, serological, and histopathological findings [1].

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 cultured PBMC 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 CpG-FITC (1.6 µg mL⁻¹) i.e. alone or forming LL-37-CpG-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 by confocal microscopy.

Results.— We found that immunostimulatory CpG oligonucleotides alone do not bind silica, but LL-37-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 CpG 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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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 immunoflorescence (Diff), 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 Diff 59% had IgA deposition alone, with IgM 25%, with IgG 6%, all but one patient (aged 16) were adults, median age 57 (16–76). Of 213 cases, 100 had direct immunoflorescence (DI). 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.

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 Diff 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 deromphelial 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.

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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), poliarteritis nodosa (PAN) (n = 4) and giant cell arteritis (n = 2). Regarding these, 43 patients distribution of the vessel involvements were arcus 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 aneurysms and one patient uncontrolled hypertension was accepted as the cause of aneurysm. In 2 SLE patients with infective endocarditis, myotic aneurysms 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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