Discussion – Raised arterial stiffness was detected at baseline in GPA patients. Mean endothelial function improved at 6 months after rituximab in the three patients with available follow-up data.

Conclusion – Our preliminary data demonstrate a possible association between rituximab and improved endothelial function in GPA. A full report is awaited.

References

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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-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 hyperthyroidism, 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-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 reading

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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 (SGV) 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 SGV.

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 cholecistitis. 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. SGV is associated with high mortality.

Further readings

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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 lymphoedema. 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].