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The association of granulomatosis with polyangiitis and IgG4-related disease: A case series

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Introduction.— IgG4-related disease is characterized by tumefactive lesions, fibrosis and an infiltrate of IgG4 positive plasma cells. Increased IgG4 plasma cells is nonspecific and can be seen in many inflammatory diseases but biopsies should lack other features of lymphoplasmacytic infiltrate, storiform fibrosis and obliterator phlebitis. A 53-year-old female presented with severe periaortitis, peri-carditis and positive PR3-ANCA. Pericardial biopsy revealed swirling fibrosis, a lymphoplasmacytic infiltrate and an IgG4/IgG ratio of 63%. IgG4-related periaortitis was diagnosed. One month later she developed mononeuritis multiplex; sural nerve biopsy demonstrated necrotiling vasculitis with an IgG4/IgG ratio of 33%. Granulomatosis with polyangiitis (GPA) was diagnosed. This prompted us to examine biopsies of patients with confirmed GPA for evidence of IgG4-related disease.

Patients.– A case series selecting patients with “Wegener’s” listed as a reason for biopsy and clinically confirmed GPA.

Results.– Eleven biopsies were examined from 10 patients. Five were eliminated as they did not demonstrate active GPA. Of the six included biopsies, two had increased numbers of IgG4 positive plasma cells. A sinus biopsy revealed 90 IgG4 cells per hpf with an IgG4/IgG ratio of 26%. A lung biopsy revealed 30 IgG4 cells per hpf with an IgG4/IgG ratio of 66%. Neither met criteria for IgG4-related disease as they lacked the other characteristic pathologic features and clinical signs consistent with this disease.

Discussion.– It does not appear that biopsies of GPA consistently meet criteria for IgG4-related disease. However, the index case may indicate a relationship between GPA-associated periaortitis and IgG4-related disease. Aortitis associated with GPA is more likely to be a periaortitis than in other forms of large vessel vasculitis which adds strength to this association.

Conclusion.– Granulomatous with polyangiitis is not consistently associated with IgG4-related disease but may be associated with GPA-related periaortitis.

Further readings

Multiple infections in a Filipino with polyangiitis overlap syndrome (POS)

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Introduction.– POS refers to primary vasculitis not classifiable into a well-defined syndrome. Infections may play etiologic roles in primary vasculitis. Our objective is to present POS with features of two ANCA-associated vasculitides (AAVs), granulomatosis with polyangiitis (GPA) and Churg-Strauss syndrome (CSS), and infections.

Patients.– A 25-year-old Filipino had 6 years of recurrent purpura and debilitating arthralgia, hemoptysis, epistaxis, eye redness, dyspnea. She had cachexia, oral ulcers, rhinorrhea, cracks, and polyparthritis. She had anemia, elevated ESR and CRP, c-ANCA and anti-proteinase 3, chronic sinusitis on rhinoscopy and lung vasculitis on chest CT. Skin biopsy showed leukocytoclastic vasculitis. There was eosinophilia without parasitism, but negative p-ANCA and anti-myeloperoxidase, also ANA and anti-dsDNA. She had high ASO titers, chronic hepatitis B, bronchial P. aeruginosa and sinus MRSA infections. HIV infection was ruled out.

Results.– There were incomplete features of limited GPA and CSS with multiple infections. She was given naproxen and culture-guided antimicrobials with partial resolution of symptoms. Prednisone was started 2 weeks after hepatitis B reactivation prophylaxis. Complete resolution of skin lesions, joint pains, and airway symptoms, and improvement of inflammatory markers were seen after 12 weeks. She is maintained on prednisone 10 mg OD.

Discussion.– There have been no more than 30 reported POS cases worldwide, and none in Southeast Asia. Our patient’s case is unique–POS as a combination of two AAVs, further compounded by multiple infections that may have triggered or perpetuated vasculitis, necessitating use of both antimicrobial and anti-inflammatory drugs to achieve remission.

Conclusion.– Knowledge of existence and early recognition of POS are essential to avoid treatment delay and prevention of irreversible organ damage.

Further reading
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Cardiovascular risk in different variants of granulomatosis with polyangiitis (Wegener’s)

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Introduction.– Persistent inflammation and treatment predispose patients (pts) with rheumatic diseases to early atherosclerosis. The risk of CV events in GPA is not well studied. The aim of our study was to evaluate the prevalence of CV risk factors, subclinical carotid atherosclerosis and CV outcomes in pts with GPA.

Patients.– Ninety-four pts with GPA were enrolled (53 females, 41 males; mean age 54.0 ± 14.9). Diagnosis was established according to ACR criteria and CHCC2012. Localized, early systemic and generalized disease was in 29, 24 and 41 pts, respectively. ANCA, mainly to PR3, were positive in 69 pts. We studied the traditional CV risk factors and outcomes (stroke, myocardial infarction [MI], revascularization procedures). Subclinical atherosclerosis was evaluated using carotid US (TIM and plaques).

Results.– We revealed high prevalence of CV risk factors in GPA: dyslipidemia – 68%, hypertension – 74%, overweight – 63%. Carotid US showed increased TIM and plaques in 63 and 27%, respectively. 10 pts (11%) had history of CV events: MI (n = 4), stroke (n = 3) and revascularization procedures (n = 3). The prevalence of dyslipidemia and overweight was similar in pts with different forms of GPA while hypertension was more frequent in generalized disease. The prevalence of carotid plaques was higher in generalized GPA but the % of pts with increased TIM was almost identical in groups. The risk of CV events tended to increase in generalized GPA. The prevalence of CV risk factors and subclinical atherosclerosis was similar in ANCA(+) and ANCA(–) pts. The rate of CV events was higher in ANCA(+) pts though P = 0.19 (Table I).

Conclusion.– The pts with GPA have high prevalence of CV risk factors, subclinical carotid atherosclerosis and increased risk of CV events. The rate of CV events tended to increase in pts with more severe forms of GPA(-) pts.

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<th>CV risk factors, subclinical carotid atherosclerosis and CV events in pts with different forms of GPA(%) K.</th>
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<td>Clinical forms</td>
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*P < 0.05 compared to generalized disease.