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A case of relapse of Churg-Strauss syndrome (CSS) with crescentic lesions
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Introduction.-- A 68-year-old female with weight loss, allergic rhinitis, bronchial asthma, hyper-eosinophilia (22 484/µL), a high titer of MPO-ANCA (over 640 EU/mL) and CRP (25.65 mg/dl), necrotizing vasculitis with crescentic lesions in a renal biopsy was diagnosed as CSS by Japanese Ministry of Health, Labor and Welfare criteria. She improved by oral corticosteroids (CS) and methylprednisolone pulses, oral cyclophosphamide (CPA). CPA was stopped after three month from start, CS slowly tapered from 40 mg/day and was off within 7 months, when she was in good condition and MPO-ANCA and CRP became within normal limits. But 16 days later from oral steroids discontinuation, fever and sinitusis reappeared. Blood test showed the reelevation of CRP 2.51 mg/dl and MPO-ANCA 17 EU/ml. Four months later sinusititis had worsened and the slight symptom of asthma appeared. Urinary test revealed urine protein (2+) and urine occult blood (3+). CRP elevated to 8.04 mg/dl, MPO-ANCA to 1000 EU/mL. Then two weeks later, she can almost eat nothing with fever and fatigue. Creatinine (Cr) elevated from 0.79 to 2.25 mg/dl and CRP to 10.58 mg/dL, eosinophil to 7280/µl, MPO-ANCA to 1570 EU/mL. She readmitted in 370 days from first visit. A renal rebiopsy was taken to find focal glomerulonephritis with fibro-cellular crescent. CS (40 mg/day) restarted and methylprednisolone pulse (500 mg/day x 3 days) was also given one time. She improved and CS was tapered to 2.5 mg/day in 524 days. Azathioprine (AZA) (50 mg/day) was also started from 405 days. In 635 days, she remains in good health and MPO-ANCA is lower to 10 EU/mL within normal limits. CS (2.5 mg/day) and AZA (25 mg/day).

Conclusion.-- It is important to notice earlier the sign of relapse for the good prognosis. In this case the symptom like common cold had not to be missed;
we need to recognize the utility of serial measurement of ANCA. Our case showed the elevation of ANCA titer from 10 EU/mL to 17 EU/mL before the appearance of serious symptoms.

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Copy number variations of FCGR3B in eosinophilic granulomatosis with polyangiitis (Churg-Strauss)
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Introduction.— FcRIIb (CD16b) is a low-affinity Fcγ receptor. It is encoded by the FCGR3B gene, acts as a stimulatory receptor, and is present almost exclusively on the surface of neutrophils. Copy number variations (CNVs) of FCGR3B gene confer susceptibility to different autoimmune conditions. They were also investigated in Microscopic Polyangiitis and Granulomatosis with Polyangiitis (Wegener’s) but not in Eosinophilic Granulomatosis with Polyangiitis (Churg-Strauss syndrome, EGPA). The aim of the present study was to explore the role of CNVs of the FCGR3B gene in EGPA.

Methods.— We studied 124 Italian patients with EGPA and 249 sex-matched Caucasian healthy controls. Genotyping was performed using Real-Time polymerase chain reaction with Taqman® custom CN assay (HS04211858; Applied Biosystems, Foster City, CA, USA). Differences in distribution of FCGR3B CNVs between patients and healthy controls were analyzed by Pearson χ2-square test. The strength of these differences was assessed by calculating P-values.

Results.— The frequencies of FCGR3B gene CN in EGPA patients and healthy controls were as follows: CN = 1 was found in 14.5% patients vs. 7.6% controls, CN = 2 in 72.6% patients vs. 80.3% controls, CN = 3 in 9.7% patients vs. 8.0% controls, and CN = 4 in 3.2% patients vs. 4.0% controls. A significant association was thus found between low FCGR3B gene CN (CN = 1 vs CN > 1) and EGPA (P = 0.036), although the overall distribution of the CNVs did not differ significantly between the two groups (P = 0.172). Clinical data were available for analysis in 90 patients. We observed a statistically significant association between low CNV and cutaneous involvement (P = 0.042), particularly with purpura (P = 0.034).

Conclusion.— Low copy number of FCGR3B gene may be a susceptibility factor for EGPA.

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Methotrexate vs cyclophosphamide as maintenance therapy in severe eosinophilic granulomatosis with polyangiitis (Churg Strauss): A subanalysis of the powercrime trial
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Introduction.— Standard therapy for severe EGPA with poor-prognosis factors (FFS ≥ 1) combines corticosteroids and cyclophosphamide (CYC) to induce remission, followed by a less toxic immunosuppressant. The randomized controlled trial Powercrime (Maritati et al. in preparation) showed that CYC and methotrexate (MTX) are equally effective and safe as maintenance therapy for AAV (MPA, GPA and EGPA). This subanalysis investigates the trial results in patients with EGPA.

Methods.— We studied patients with active EGPA and FFS ≥ 1, either at disease onset or relapse. All patients received the same remission-induction therapy, consisting of prednisone and CYC. At remission, patients were randomly assigned to continued CYC therapy (1.5–2 mg/kg/day) or to receive methotrexate (15 mg/week progressively titrated up to 25 mg/week) for 12 months. The primary end-point was relapse (time from remission to relapse).

Results.— Of the 30 EGPA patients who entered remission, 13 were randomly assigned to CYC and 17 to MTX. The median follow-up was 58.4 months (IQR, 29.4–86.2). The median time to relapse was similar in the two groups, 24 months (IQR 16–29) in the CYC group and 22.5 (IQR 19–24) in the MTX group (P = 0.95). Five patients had one or more relapses (38.4%) in the CYC group and four in the MTX group (23.6%, P = 0.51). During maintenance treatment, there was one death in the MTX group. No serious adverse events occurred in the CYC group while two occurred in the MTX group (P = 0.13).