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, hypeereosinophilia (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 CPA became within normal limits. But 16 days later from oral steroids discontinuation, fever and sinusitis reappeared. Blood test showed the reactivation of CRP 2.51 mg/dl and MPO-ANCA 17 EU/mL. Four months later sinusitis had worsened and the slight symptom of asthma appeared. Urinary test revealed urinary protein (2++) and urinary 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 3 days) was also given. In 635 days, she remains in good health and MPO-ANCA is lower to 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 in EGPA patients and healthy controls as 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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Copy number variations of FCGR3B in eosinophilic granulomatosis with polyangiitis (Churg-Strauss)

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Introduction.– FcγRIIa (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 chi-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).