

中文題目：類風溼性關節炎合併腦部血管炎和嗜酸粒細胞增多症

英文題目：Hypereosinophilia with cerebral vasculitis in a case of Rheumatoid Arthritis

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Case

A 41 year-old man with rheumatoid arthritis presented with acute onset cognitive dysfunction for two weeks.

The patient has been diagnosed with seropositive RA 3 years ago, with swelling of MCP, PIP, wrist and shoulder joints. Four months before admission, patient was regularly treated with Etanercept twice a week. During these periods, the patient had been in clinical remission without any extraarticular involvement. Etanercept was changed to Tofacitinib due to well performance and convenience of oral form medication.

Three weeks before admission, symptoms began insidiously with bilateral temporal headache without nausea or vomiting, progressive encephalopathy characterized with loss of cooperativeness and decreased spontaneous speaking in addition of bilateral upper limb weakness. Disorientation to person and place, difficulty in naming and gait imbalance were noted by his family, which affect his driving and daily activities. Comprehensive review of symptoms was essentially negative, including no upper respiratory tract symptoms, fever, night sweating, arthralgias or skin rash. Patient has no significant past medical history, and he denied recent travel history, no insect bite, nor raw food intake. He also denied any food supplement or chinese herbs used.

On admission, patient was confused, but able to perform simple commands. Laboratory data revealed marked eosinophilia, 2306/uL, elevated erythrocyte sedimentation rate 52mm/Hr, normal complement level (C3 120mg/dL, C4 43.71mg/dL), negative anticardiolipin and lupus anticoagulant, negative ANCA, with significant RF elevation. Review past history, patient's eosinophil count throughout the disease course prior to commencement of Tofacitinib was within normal range. Brain computed tomography scan revealed multiple ill-defined low attenuated foci of bilateral brain parenchyma, predominantly involving bilateral cerebral cortex and subcortical white matter. No evidence of intracranial hemorrhage. Lumbar puncture showed CSF protein 58.2mg/dL, other survey including Herpes Simplex Virus polymerase chain reaction (PCR), Venereal disease research laboratory (VDRL), cryptococcal antigen, gram stain, bacterial and fungal cultures, tuberculosis were all negative. Brain magnetic resonance imaging (MRI) was compatible with vasculitis, presented with multifocal, patchy lesions (mainly of cortical-subcortical region, with small focal hemorrhages, mild gyriform enhancement, and restricted diffusion) of bilateral cerebral hemispheres is noted, infarction of varying stage is considered. Imaging and clinical presentation highly suggest vasculitis or embolic lesions. Cardiac echocardiography showed no vegetation nor valvular abnormalities. Brain biopsy was not performed due to lack of technique expertise in our hospital.

Cerebral vasculitis was highly impressed, either extra-articular manifestation of RA or sequela of hypereosinophilia. IV form Methylprednisolone 500mg was prescribed for three consecutive days, accompanied with one single dose of Cyclophosphamide 250mg. Eosinophil counts and symptoms improved drastically. During outpatient follow up, symptoms improved gradually under medication and rehabilitation.

Discussion

RA is a chronic progressive systemic inflammatory disorder involving joints. It is associated with many extra-articular manifestations, including systemic vasculitis but rarely isolated cerebral vasculitis. Hypereosinophilia is an abnormal condition defined by the presence of more than 500 eosinophils per microliter of blood. It occurs in allergic reaction, helminthic infestation, collagen vascular disease and malignancies.(1) Recent study showed that eosinophilia is quite common in RA patients but does not have any relation with disease activity. (2) Besides, a single case of drug-induced eosinophilia related to Tocilizumab therapy was reported.(3)

In our case, marked eosinophilia was noted before admission. It could be related to the JAK-inhibitor Tofacitinib, RA disease itself, or idiopathic hypereosinophilia which may lead to cerebral vasculitis(4, 5) or embolic infarction. Kim et. al. measured TNF level in a case of hypereosinophilia, which revealed marked TNF increased and suggested that TNF could play a role in the inflammatory processes.(6)

References

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