

Supplementary file

An atypical case of spontaneous regression.

Case: A 41-year-old man had initially bilateral LG enlargement (Supplement figure) and high serum levels of IgG4 (1530 mg/dL, normal; <104 mg/dL) and soluble IL-2 receptor (1461 U/mL, normal; 135-483 U/mL), as well as blood eosinophilia. The patient presented with schizophrenia and refused the corticosteroid treatment. After 35 months of follow-up, the bilateral LG enlargement decreased spontaneously (supplement figure), and the serum levels of IgG4 and soluble IL-2 receptor decreased to 473 mg/dL and 376 U/mL, respectively.

Discussion

While we could not determine the reason for the spontaneous regression, interestingly, the patient had a concurrent illness of schizophrenia with a worse condition. The clinical activity of schizophrenia has been linked to the level of soluble IL-2 receptor,[1, 2] which may be correlated with systemic T cell activation *in vivo*. [3] Thus, the patient may have had T cell-related systemic immune activation. In our study, patients with EOM/CN V enlargements had a significantly greater incidence of blood eosinophilia and elevated serum level of

IgG4 which might be correlated with systemic immune activation. Thus, the patient may paradoxically support our idea that systemic immune activation might underlie the activity of IgG4-ROD.

Supplementary figure legend

Left: Bilateral lacrimal gland enlargement were observed at the initial visit.

Right: Right lacrimal gland enlargement spontaneously regressed at 36 months after the initial visit.

References

1. Igue R, Potvin S, Bah S, et al. Soluble interleukin-2 receptor levels correlated with positive symptoms during quetiapine treatment in schizophrenia-spectrum disorder. *Prog Neuropsychopharmacol Biol Psychiatry* 2011;35:1695-8.
2. Steiner J, Bernstein HG, Schiltz K, et al. Immune system and glucose metabolism interaction in schizophrenia: a chicken-egg dilemma. *Prog Neuropsychopharmacol Biol Psychiatry* 2012;48:287-94.
3. Rubin LA, Nelson DL. The soluble interleukin-2 receptor: biology, function, and clinical application. *Ann Intern Med* 1990;113:619-27.