Authors' reply to 'Important role of corticosteroids in chronic granulomatous disease'

Roesler (2007) has contributed a correspondence article on the importance of steroids in the treatment of chronic granulomatous disease (CGD) in response to our paper (Arimura *et al.*, 2006). First, we would like to describe here in detail the clinical course of the case, which was not included in the original report on account of saving journal space.

In September 2004, a patient (aged 19 years) presented with bloody diarrhoea, which he had contracted for the first time; an unclassified colitis associated with CGD was diagnosed in the patient. The bloody diarrhoea was quickly resolved with intravenous prednisone (60 mg per day) drip infusion after 5-aminosalicylate failed to induce remission. Four months after the initial presentation of colitis, when the oral dose of prednisone was being tapered to 7.5 mg per day, the bloody diarrhoea recurred, remission was induced by administering 60 mg per day intravenous prednisone. The bloody diarrhoea flared up again during the tapering off of prednisone to 17.5 mg per day. The patient was referred to our hospital for admission 6 months after the second relapse. Clinical remission was achieved once again by administering 40 mg per day intravenous prednisone for 2 weeks; towards the end of this course, the patient developed a disseminated lethal varicella-zoster virus infection while receiving 35 mg per day prednisone.

As mentioned above, the patient experienced three relapses of colitis; on the first relapse, prednisone was tapered off to 7.5 mg per day, on the second to 17.5 mg per day and on the third to 35 mg per day under appropriate prophylaxis. This is a case of the so-called corticosteroid-dependent intractable colitis in which the remission maintenance period shortens after every relapse. Finally, the patient unfortunately developed severe varicella infection due to the adverse effects of the prednisone therapy.

Corticosteroids are extremely effective for symptomatic relief from CGD-related colitis in the short term; however, the side effects of the long-term usage of corticosteroid and dependency on it can be very serious. Undoubtedly, one of the most important therapeutic options is to use corticosteroids as a short-term rescue in the treatment of life-threatening opportunistic infections, such as the fatal *Aspergillus* pneumonia or fatal organopathy caused by CGD, as described by Roesler (2007), although this treatment is not specific for CGD.

However, unfortunately, corticosteroid therapy is different from the definitive therapy for CGD-related colitis. As discussed in the original report, we would like to stress that the determination of the optimal treatment of colitis secondary to CGD is a crucial goal to be achieved.

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Arimura, Y., Goto, A., Yamashita, K., Endo, T., Ikeda, H., Tanaka, K., Tsutsumi, H., Shinomura, Y. & Imai, K. (2006). Intractable colitis associated with chronic granulomatous disease. *J Med Microbiol* 55, 1587–1590.

Roesler, J. (2007). Important role of corticosteroids in chronic granulomatous disease. *J Med Microbiol* 56, 1253.

DOI 10.1099/jmm.0.47324-0