Siltuximab for Multicentric Castleman Disease—Letter

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We read with great interest the article by Deisseroth and colleagues (1) focusing on the recent global approval of the monoclonal IL6 antibody siltuximab for the treatment of patients with HHV-8-, HIV-negative multicentric Castleman disease (MCD). Up to now, treatment options for this rare lymphoproliferative disorder are very limited.

Lately, the phase III, double-blind, randomized trial of van Rhee and colleagues (2) reported substantial and durable response rates for siltuximab in the treatment of MCD patients. MCD is known to present with various clinical symptoms (e.g., fever, fluid retention, and skin rashes) related to an excessive secretion of IL6 (3). Furthermore, there are some reports of paraneoplastic skin diseases secondary associated with the disease (4, 5).

Here, we report the case of a 25-year-old patient, presenting with steroid-refractory MCD and severe paraneoplastic blistery skin lesions, showing a remarkable response to single-agent siltuximab. The patient was first diagnosed idiopathic MCD in 2011. Subsequently, he was treated several times with steroids and rituximab, each time with a shorter duration of response. In 2014, he presented again with signs of an active lymphoid disease (fever, lymphadenopathy, and fatigue), but this time with additional polymorphic skin lesions. Clinical examination showed severe confluent blistery skin changes at the forearms, wrists, and ankles (Fig. 1A and C), together with multiple purulent papules disseminated over the whole trunk. Blood counts and clinical chemistry revealed an excessive inflammatory response (CRP, 225 mg/L; normal, <5 mg/L; IL6, 309 pg/mL; normal, 2–3.5 pg/mL). Serological diagnostics and smear tests from the blisters were negative for viral or bacterial infections. ELISA tests for autoantibodies against desmoglein 1 and 3, and BP 180-NC 16A were negative. No specific antibodies for pemphigoid and pemphigus or antiendomysial antibodies could be detected by indirect immunofluorescence. Taken together, we suspected a severe paraneoplastic skin disease in an active MCD.

Because of the clinical nonresponse to steroids, single-agent siltuximab (11 mg/kg body weight) was started. Treatment showed an immediate response of both the cutaneous lesions and the patient’s general condition (Fig. 1B and D; after 2 doses of siltuximab) and was continued every 3 weeks. CRP and IL6 levels normalized rapidly within 4 days. Today, the patient is still in an ongoing remission.

The course of the patient demonstrates that siltuximab can provide a capable and well-tolerated option for a sustained remission, not only for the lymphoproliferative disease itself but as well for paraneoplastic phenomena secondary associated with MCD.

Disclosure of Potential Conflicts of Interest
No potential conflicts of interest were disclosed.

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