A 13-year-old girl presented with exacerbation of symptoms including photophobia, itchy eyes, and pain in her right eye. Slitlamp examination revealed giant papillae on the upper tarsal conjunctiva and superficial punctate keratitis in the right eye . Her best-corrected visual acuity was 20/63 OD and 20/16 OS. She had previously been diagnosed with VKC associated with AD at 9 years old and for 4 years was treated with eye drops containing tacrolimus, cyclosporine, steroids, and antiallergic agents. Despite topical treatment with eye drops, the giant papillae on the tarsal conjunctiva continued to worsen and resolve seasonally. Corneal lesions such as a shield ulcer and superficial punctate keratitis in her right eye persisted for 3 months. AD was observed on her skin, including the face, and was treated with tacrolimus ointments, steroid ointments, and a moisturizer. As the patient’s AD was severe with persistent skin rash refractory to treatment with topical medications, oral upadacitinib (15 mg/d) was administered for AD. Scores for the Eczema Area and Severity Index and the Investigator’s Global Assessment before and 2 weeks after upadacitinib treatment improved from 28.0 to 1.2 and 3 to 2, respectively. The pain and itching in the right eye improved within a month after upadacitinib administration. Additionally, the giant papillae gradually flattened, and corneal lesions improved after upadacitinib administration. After 3 months of oral upadacitinib administration, the giant papillae and punctate epithelial keratitis resolved completely , and vision improved to 20/16 OD. After 11 months of treatment, upadacitinib dosage was increased to 30 mg/d owing to the exacerbation of AD. After more than 14 months of upadacitinib treatment, no recurrence of giant papillae or corneal damage was observed with topical treatment of only antiallergic eye drops , and visual acuity was maintained at 20/16 OD and 20/20 OS.