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Article type : Letter to the Editor

Efficacy of tofacitinib in Kimura's disease in a patient with concomitant atopic dermatitis

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Funding Sources: This work was supported by the Zhejiang Medical and Health Science and Technology Project (2015KYA092, 2020KY558).

Conflict of Interest: None to declare.

Dear editor, Kimura's disease (KD) is a rare chronic inflammatory disorder that mainly affects young Asian men. It is characterized by subcutaneous mass or nodule in the head and neck region¹. There is no standard treatment for KD.²

Here, we report Janus kinase (JAK) inhibitor tofacitinib was effective for treating a case of KD. This article has been accepted for publication and undergone full peer review but has not been through the copyediting, typesetting, pagination and proofreading process, which may lead to differences between this version and the [Version of Record](#). Please cite this article as [doi: 10.1111/CED.14671](#)

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KD associated with severe atopic dermatitis (AD). Institutional Review Board approval was granted (ID: IIT20200554A).

A 46-year-old male presented to our department of dermatology for treating his severe chronic AD suffering from childhood. He complained he had a history of KD for 20 years that presented with recurrent painless masses behind both ears and in the inguinal regions. Ultrasonographic examination revealed hypoechoic masses in the dermis in these areas. Excision biopsies showed nodular lymphocytic infiltrate with germinal centers involving the dermis, and reactive germinal centers were surrounded by lymphocytes and eosinophils. Laboratory investigations were unremarkable besides increased serum IgE level of 5676.9 IU/mL (reference range: ≤ 158.0 IU/mL). A diagnosis of KD was established. Masses reoccurred in the inguinal regions 3 years after surgery. Although his eosinophil count was elevated to $0.75 \times 10^9/L$ 6 year ago, this had decreased to normal.

Physical examination revealed generalized eczema, with lichenification mainly in the flexures. Postoperative scars were found behind both ears and right groin. Subcutaneous masses with diameters of 2cm and 4cm were palpable on both sides of inguinal region.

Topical corticosteroids and oral antihistamines were not effective for controlling the symptoms of AD. He received intermittent systemic corticosteroids and the eruption relapse shortly after withdrawal of the corticosteroids.

After discussion of the treatment options for AD, tofacitinib 5mg twice daily was administered, according to the patient's preference. After 3 months, the total Scoring Atopic Dermatitis was reduced from 57.3 to 16.2, and the Eczema Area and Severity Index was reduced from 25.5 to 3.6.³ The itch was also significantly improved. The bilateral inguinal masses subsided completely (Fig. 1). Total serum IgE level decreased to normal. The patient continued to receive tofacitinib treatment in the following 1 year. Except for slight drowsiness and xerophthalmia in the first 3 months of receiving tofacitinib, no other side-effects were found. The lesions of KD did not relapse whilst on tofacitinib.

To our knowledge, this is the first case of KD concomitant with AD who was successfully treated with JAK inhibitors. The etiology and pathogenesis of KD are still currently unknown. The

proliferation of CD4⁺ T cells, specifically the CD4 Th2 cells leads to overproduction of cytokines, including granulocyte macrophage colony-stimulating factor, tumor necrosis factor- α , IL-4, IL-5, eotaxin, and RANTES trigger the production of lymphoid follicle and high IgE of KD patients.⁴ It is suggested that Th2 pathway may also involve in the pathogenesis of KD. This may explain the effectiveness of tofacitinib for treating KD.

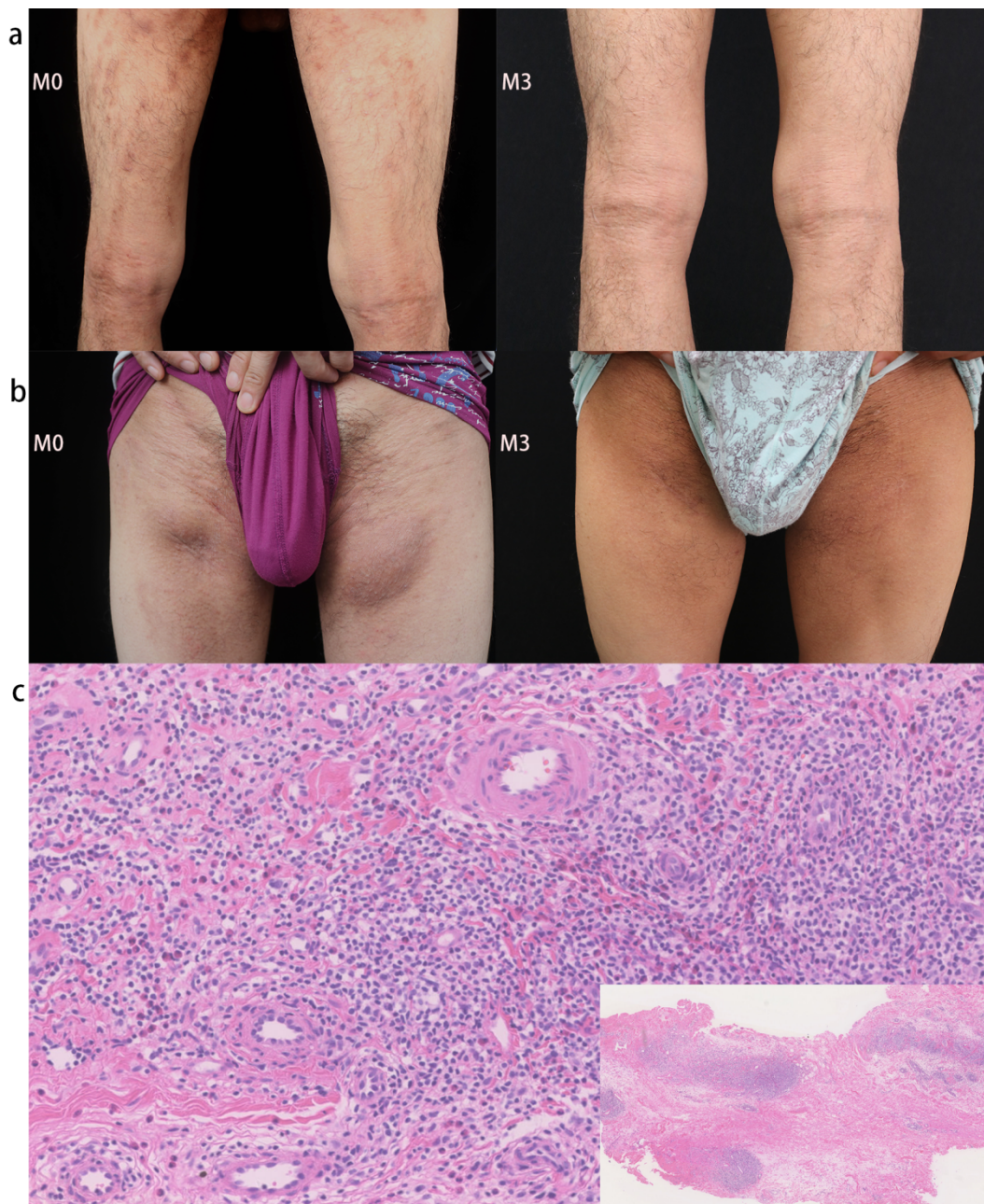
In conclusion, our case report indicates that tofacitinib may be an effective and safe therapy for patients with KD. More clinical evidence is needed to support this observation.

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Figure legend:

Fig. 1 Representative photographs of the patient with concomitant atopic dermatitis (AD) and Kimura's disease (KD) and histopathological image of KD. (a) Improvement of AD from month 0 to month 3. (b) Improvement of KD from month 0 to month 3. (c) The histological features of KD: Nodular lymphocytic infiltrate with germinal centers involving the dermis, and reactive germinal centers surrounded by lymphocytes and eosinophils. (HE $\times 15$, $\times 2.5$)



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