

Extranodal NK/T cell lymphoma mimicking Behçet's syndrome

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An 18-year-old man presented to our department with recurrent oral aphthous ulceration and a 1-month history of low-grade fever. On physical examination, there was painless ulceration of his glans penis (Figure 1A) and scrotum (Figure 1B) and erythematous plaques on his lower extremities. Behçet's syndrome was suspected and treated with low-dose glucocorticoids and thalidomide. However, his skin lesions failed to resolve and quickly progressed (Figure 1C and D). His erythrocyte sedimentation rate was normal. HIV, rapid plasma regain, and pathergy tests were all negative. Histopathological examination of the skin biopsy revealed a diagnosis of extranodal NK/T cell lymphoma (ENKTL), nasal type, for which he received chemotherapy.

ENKTL usually arises in midline facial structures and presents as localized disease. The clinical symptoms of the primary lesion are sometimes unremarkable, and penile metastases have only very rarely been reported.^[1] Behçet's syndrome is a multisystem vasculitis that can cause oral aphthous ulceration, genital ulceration, eye and skin lesions, and a positive pathergy test.^[2, 3] Genital ulcers, often appearing on the scrotum in men, are highly specific for Behçet's syndrome; however, in our case, ENKTL with penile metastasis was an exceedingly rare mimic. The defined skin manifestations of Behçet's syndrome include erythema nodosum, pseudofolliculitis, acneiform nodules, papulopustular lesions, superficial thrombophlebitis, and palpable purpura^[4] but not

the erythematous plaques seen in our case. The presence of atypical, progressive skin lesions should prompt further investigations to avoid diagnostic delay.



Figure 1. Ulceration of the patient's glans penis (A), scrotum (arrow in B), and erythematous plaques or infiltrated papules (arrow in C) on the patient's lower extremities (C) and trunk (D).

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Ethical statement

This study was granted an exemption from review by the Institutional Review Board of Peking Union Medical College Hospital.

Conflict of interest

All authors declare that no conflict of interest exists.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and the accompanying images.

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