

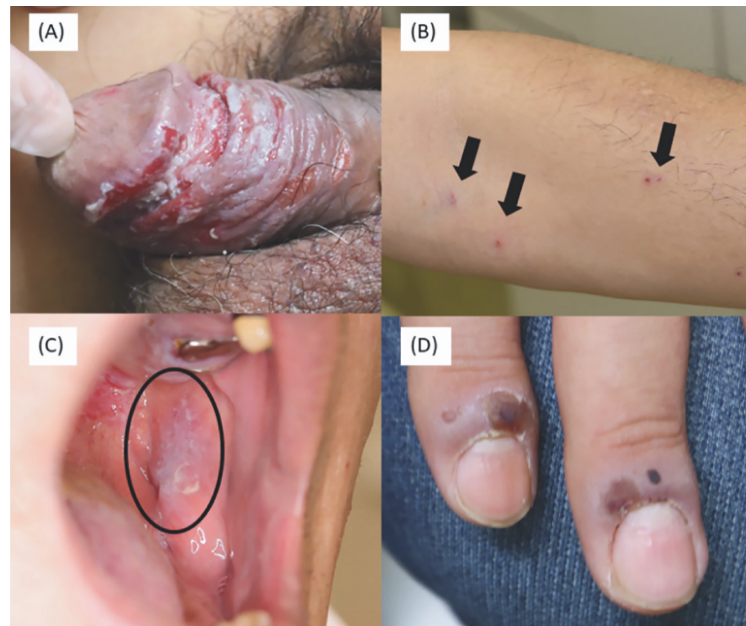
## A case of pemphigus vulgaris with folliculitis-like nodules, genital and oral ulcers difficult to differentiate from Behçet's disease

Sirs,

Pemphigus vulgaris (PV) is an autoimmune mucocutaneous vesiculobullous disease (1). The condition typically begins with mucous lesions that, after several weeks or months, start to affect the skin. Among mucous membranes, the most commonly involved is the oral mucosa. Although nail involvement is occasionally detected in PV patients, it is underappreciated and viewed as a sign of the severity of PV (2, 3). Behçet's disease (BD) is a complex multisystem disease characterised by the presence of oral ulcers, genital ulcers, synovitis, erythema nodosum, folliculitis or posterior uveitis (4). For most patients with BD, oral ulcers are the first and the most frequent symptoms (5) and are usually recurrent. Since both PV and BD present with oral mucosal disorders and genital ulcers, they are sometimes challenging to distinguish. We describe a patient diagnosed with PV by histopathological findings of nail folds lesions who presented with oral, genital ulcers, and folliculitis-like nodules.

A 45-year-old man who had no medical or family history of the disease reported symmetric excruciating swelling of the proximal nail folds of all fingers of both hands (Fig. 1) and both first toes. Additionally, he had folliculitis-like lesions on his upper arm, oral ulcers on both sides of the buccal mucosa, and genital ulcers in his penis (Fig. 1). He had been treated with valaciclovir and topical antibiotics for two weeks without any enhancement. One month later, he was referred to our centre for suspecting BD. During the physical examination, suppurative inflammation was observed over the proximal and lateral nail folds of all fingers of both hands and both first toes. Confluent ulcers on both sides of the buccal mucosa, genital ulcers on the sulcus coronarius, and folliculitis-like nodules in the upper arms were also noted. These lesions were painful. An ophthalmologic examination showed no abnormalities. Laboratory tests revealed a white blood cell count, of 10500/mm<sup>3</sup> (neutrophils 60%, lymphocytes 30.7%, eosinophiles 2.5%, basophiles 1.0%, and monocytes 5.7%); and C reactive protein 0.48 mg/dL. Negative autoantibody titers included antinuclear, antineutrophil cytoplasmic, antidesmoglein1, and antibullous pemphigoid 180 antibodies. Antidesmoglein3 antibodies were 4.2 IU/mL (normal range, <19.9).

Written informed consent was obtained from the patient prior to participation in the study. This study was performed according to the principles of the Declaration of Helsinki. Ethical Approval Statement was not required for this manuscript



**Fig. 1.** A: Genital ulcers in the penis. B: Folliculitis-like nodules (arrows) in the upper arm. C: Oral ulcer (red circle) on the left side of the buccal mucosa. D: Suppurative inflammation over the proximal nail folds of the fingers.

We believe that the oral and genital ulcers and folliculitis-like nodules might be associated with BD. However, the nail fold lesions were rare for BD, and therefore we conducted a skin biopsy of the nail fold. The biopsy specimens showed hyperkeratotic skin tissue with lymphocyte-dominated inflammatory cell infiltration seen just below the epidermis, and acantholysis was observed just above the basal layer in the epidermis. Direct immunofluorescence of the perilesional skin revealed epidermal intercellular deposition of IgG and C3. He was identified with PV and he was treated with oral prednisolone at an initial dose of 1 mg/kg/day. All cutaneous and mucosal lesions were improved by these therapies within two months.

In our patient, oral ulcers were not recurrent and not the first manifestation. But we could not rule out BD because he also complicated with genital ulcers and folliculitis-like nodules. Although nail changes are seen in 47% of PV patients, it is underestimated (6). Paronychia was the most common finding and was seen in 60% of PV patients followed by onychomadesis (2).

We herein report a patient with PV who describe oral and genital ulcers and folliculitis-like nodules which are also presented in BD. The patient was identified as having PV based on histological evidence of nail fold lesions, which are occasionally observed but underappreciated in PV. Since BD is identified by a combination of symptoms in absence of specific findings, it is essential to concentrate on symptoms that are atypical for BD and to make a differential diagnosis.

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