CASE REPORT

Oral ulcerations after placement of orthodontic braces and skin pustules after laser hair removal: novel inducers of pathergy reactions in new-onset Behçet's disease

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SUMMARY

A 25-year-old man was admitted to our hospital, with fever, severe ulceration of the oral and pharyngeal mucosa, and pustules on his skin. These pathergy-like lesions had emerged after placement of orthodontic braces and laser therapy for hair removal, respectively. The patient's clinical condition, together with his ethnic background, pointed to the diagnosis of Behçet's disease. Treatment with colchicine and prednisone resulted in rapid improvement of his symptoms. Pathergy reactions in response to placement of dental braces and laser hair removal are a rare first presentation of Behçet's disease.

BACKGROUND

Behçet's disease is a systemic vasculitis involving small and large sized vessels. Clinical manifestations most typically include recurrent oral and genital ulceration with uveitis, but may also involve the vascular system, central nervous system and gastrointestinal system. The exact aetiology is unknown, although there is a clear association with specific genetic backgrounds such as those carrying the human leucocyte antigen (HLA)-B51 allele.¹ Behçet's disease is most prevalent in countries along the Old Silk Road, which extends from eastern Asia to the Mediterranean.^{2 3} In the absence of a specific diagnostic test for Behçet's disease, the diagnosis relies on clinical pattern recognition. The International Study Group for Behçet's Disease composed a set of diagnostic criteria, which require the presence of oral ulceration plus any two of the minor criteria: genital ulceration, specific eye lesions, specific skin lesions, or a positive pathergy test.¹ A positive pathergy test shows a pustule-like lesion or papule 48 h after skin prick by a 20 gauge needle.⁴ Our case report is, to the best of our knowledge, the first description of a patient with new-onset Behçet's disease in whom the presenting symptoms reflect a classic pathergy reaction to non-classical modern-day traumas of the skin and oral mucosa.

CASE PRESENTATION

A 25-year-old man was admitted to our hospital, with fever, and severe ulceration of the oral and pharyngeal mucosa. His medical history was unremarkable and he did not take any medication. The

patient was born in Iraq and had migrated to the Netherlands when he was 5 years old. For 4 weeks prior to admission, he had been experiencing a persisting fever up to 40°C without obvious focal symptoms of infection. Empiric antibiotic treatment by his general practitioner had not affected the course of his febrile illness. During this period, he had received orthodontic braces. Within days after placement, painful oral ulcerations occurred. These lesions progressed in severity to the point where he had difficulty opening his mouth, was unable to take in solid foods and had substantially lost weight. One week prior to admission, the patient had also undergone laser treatment performed on his arms and torso for hair removal. Within days after the procedure, pustules occurred at the specific sites of laser treatment, but later also at distant sites such as the face.

On physical examination, we saw a young man in great pain due to extensive oral ulcerations (figure 1). On the skin, painless pustules were present on the head, torso and arms (figure 2). Besides multiple tender palpable cervical lymph nodes, there were no other remarkable findings during the general physical examination.

INVESTIGATIONS

Laboratory tests showed an increased erythrocyte sedimentation rate (36 mm/h, reference 0–15 mm/h) and a mildly elevated C reactive protein level (42 mg/L, reference 0–10 mg/L). A chest X-ray was normal. On ultrasound, the cervical lymph nodes were slightly enlarged but did not display malignant characteristics. A PCR for herpetic viruses on an oral swab was negative. Bacterial culture of the content of a skin pustule was negative.

DIFFERENTIAL DIAGNOSIS

- Herpes virus infection of the oral mucosa
- Bacterial folliculitis of the skin
- Type I hypersensitivity reaction

TREATMENT

Under suspicion of Behçet's disease, the patient was treated with prednisone and colchicine.

OUTCOME AND FOLLOW-UP

The patient experienced rapid improvement of oral and cutaneous symptoms within a day after

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Figure 1 Oral ulcers.

initiation of treatment. After 2 weeks of treatment, the prednisone dose could be tapered and was eventually stopped while continuing colchicine maintenance therapy. The patient has been free from relapse since.

DISCUSSION

The prevalence of Behçet's disease is highly variable across ethnic groups. In Northern European countries, the prevalence ranges from 1 case per 15 000 to 1 case per 500 000 people, while Behçet's disease is most common in Turkey and surrounding countries with prevalence of up to 370 cases per 100 000 people.^{2 3 5}

The pathergy phenomenon is defined as a state of altered tissue reactivity that occurs in response to minor trauma and is a very typical finding of Behçet's disease. The exact pathogenesis of the phenomenon is unknown, but minor trauma apparently triggers a much more extensive release of cytokines in the

epidermis than normal, resulting in perivascular infiltration, as observed in skin biopsies.^{4 6} A study conducted by Inaloz *et al*⁷ suggests that cell adhesion molecule interaction together with endothelial proliferation may play an important role in the formation of the pustules.

The mucosal and cutaneous lesions occurring in response to minor traumata described in this case are similar to those caused by a needle in a positive pathergy test. Therefore, together with the fact that the extensiveness of the lesions was out of proportion compared with the intensity of the traumata, we consider this a positive pathergy test. The differential diagnosis included infections and a classic type I IgE-mediated hypersensitivity reaction. We excluded a herpetic virus infection in the oral cavity and bacterial infection as a cause of the skin lesions. A classic type I hypersensitivity reaction was considered unlikely because the cutaneous symptoms would typically include an urticarial reaction rather than the papulopustular skin lesions observed in our patient. Also, the oral ulcerations are inconsistent with a type I hypersensitivity reaction. Based on the exclusion of other causes, the clinical symptom pattern and the rapid response to initiated therapy, we established the diagnosis of Behçet's disease.

We do not know of any other report of a patient with Behçet's disease having these presenting symptoms, although a study by Karacayli *et al*⁸ showed that dental and periodontal therapies may be associated with a flare-up of oral ulcers in patients with established Behçet's disease.

Learning points

- ▶ In Behçet's disease, pathergy reactions may occur as presenting symptom in reaction to various clinical triggers, here in response to placement of dental braces and laser hair removal.
- ▶ This emphasises that Behçet's disease should be considered when unusual mucosal or cutaneous phenomena occur after minor trauma.
- ▶ Clinicians should take the ethnic background into consideration when evaluating a patient with symptoms of diseases, such as Behçet's disease, associated with a specific genetic risk profile.

Competing interests None declared.

Patient consent Obtained.

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Figure 2 Folliculitis.

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