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SHORT COMMUNICATION

Isolated conjunctival ulcerations as the first sign of Behçet's disease

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PURPOSE. *To report an unusual incident of isolated conjunctival ulcerations which preceded all other signs of Behçet's disease.*
DESIGN. *Observational case report.*
METHOD. *Description of a 34-year-old woman of West Indian origin with an unremarkable medical history presenting with bilateral bulbar conjunctival ulcerations.*
RESULTS. *The authors noted the presence of a diffuse bilateral conjunctival hyperemia which was more pronounced at the temporal bulbar conjunctiva of the right eye and the nasal conjunctiva of the left eye, each having one ulceration. The rest of the ophthalmologic examination was normal. One month later, the appearance of buccogenital aphthosis led to the diagnosis of Behçet's disease.*
CONCLUSIONS. *This observation is unusual, as the conjunctival ulcerations are isolated and precede the onset of all other symptoms of Behçet's disease. Conjunctival ulcerations are rarely seen with Behçet's disease, but are characteristic enough to be included among the diagnostic criteria. (Eur J Ophthalmol 2006; 16: 751-2)*

KEY WORDS. *Behçet's disease, Conjunctival aphthosis, Conjunctival ulceration, Uveitis*

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INTRODUCTION

Behçet's disease is a relapsing systemic vasculitis that primarily affects small veins. Ocular impairment is very common and most often presents as an anterior or posterior uveitis or an optic neuropathy (1). Isolated conjunctival ulcerations are rare, especially when they are the initial sign of disease, as in our observation.

Case report

A 34-year-old woman of West Indian origin had complained of bilateral ocular pain for several months. Her medical and surgical histories were unremarkable. The visual acuity in both eyes was 20/20. We noticed a diffuse bilateral conjunctival hyperemia, more pronounced at the temporal bulbar conjunctiva of the right eye and the nasal conjunctiva of the left eye, each having one ulceration. Both ulcerations were round and measured approximately 2 mm in diameter (Fig. 1). They were located distal to the limbus, their borders distinct and surrounded by an edematous and erythematous halo. The fundus was a yellowish color and retained fluorescein. There was no keratoconjunctivitis sicca or signs of anterior or posterior uveitis. Fluorescein angiography did not reveal subclinical retinal vasculitis. Treatment with dexamethasone drops was initiated at a rate of six times per day. Full recovery with total resolution of ulcerations without scarring was obtained after 10 days of treatment.

One month later, the patient was hospitalized for a complete workup of diarrhea and buccogenital aphthosis associated with a marked decline in overall health. Endoscopic exploration of the esophagus and colon revealed numerous round, disseminated, superficial ulcerations accompanied by vascular lesions and separated by zones of intact mucosa. The HLA B12 allele was identified. The cu-

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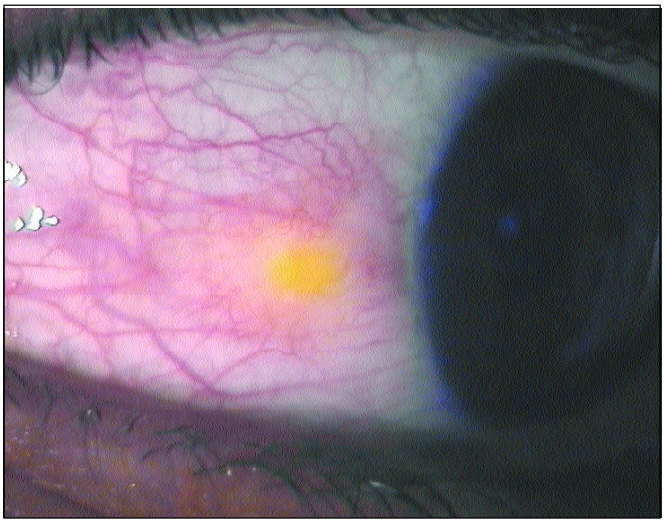

Fig. 1 - *Right eye. Temporal conjunctival ulceration approximately 2 mm in diameter situated distal to the limbus and surrounded by an edematous and erythematous halo. The fundus is yellowish and retains fluorescein.*

taneous pathergic test was positive. A diagnosis of Behçet's disease was made according to the criteria of the International Study Group for Behçet's disease (1). The digestive symptoms were treated acutely with corticotherapy followed by colchicine. In 3 years of follow-up, several relapsing buccal or genital aphthoses were identified, one being accompanied by digestive lesions. No ocular relapse was observed.

DISCUSSION

Except for a few isolated observations, four references describe more than one observation of conjunctival ulcerations (1-4). Zamir et al report six cases, three of which have a history of uveitis (2). Matsuo et al describe four cases observed among 152 (2.6%) patients in Japan (3). In Turkey, the incidence of conjunctival ulcerations would be 5/540 (0.9%) (4). Paradoxically, in India the conjunctival ulcerations would be more frequent than uveal damage: 5/19 (26.3%) versus 2/19 (10.5%) (5). In all these cases, the conjunctival ulcerations were preceded by another mucocutaneous or ocular sign of Behçet's disease. They arose during an exacerbation of the disease and were always accompanied by other symptoms. The histologic study showed an ulceration of the conjunctival epithelium in which the fundus was infiltrated by polymorphonuclear leukocytes, lymphocytes, and macrophages particularly around the vessels (2, 3). In the majority of cases, conjunctival ulcerations appeared at the same time as ulcerations of the mouth, digestive tract, or genitals, and rarely at the same time as an inflammatory attack of the uvea, which suggests a common mechanism for the incidence of all the mucosal ulcerations.

This observation is unusual, as the conjunctival ulcerations are isolated and precede the appearance of all other signs of Behçet's disease (MEDLINE, National Library of Medicine). Conjunctival ulcerations are rarely seen in Behçet's disease, but are characteristic enough to be included among the diagnostic criteria.

No authors have any proprietary interest.

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