

CLINICAL VISTAS

Behçet's disease

A 27-year-old man of Middle Eastern descent presented to the hospital after waking with severely reduced visual acuity in his left eye. Several months before presentation he had experienced an episode of reduced visual acuity in his right eye that had slowly resolved over several weeks. The patient had a history of long-standing recurrent oral-labial aphthae, intermittent painful swelling of his knees and ankles, low-grade fevers and night sweats.

On examination, the patient was afebrile and had normal vital signs. An oral-labial aphtha was present (Fig. 1). His visual acuity was 6/60 in the left eye

and 6/6 in the right. Funduscopy revealed disc edema and retinal vasculitis, shown by superficial retinal hemorrhages with pale centres (Fig. 2). Anterior uveitis was noted on examination with a slit lamp. The patient also had erythematous plantar maculae (Fig. 3), effusion in his right knee and scattered pseudofolliculitis.

The results of blood counts and serum tests were normal. The C-reactive protein level was elevated at 123 (normal < 5) mg/L, as was the erythrocyte sedimentation rate at 35 (normal < 10) mm/h. Results of serologic tests, including those for collagen vascular diseases, were normal. Thoracic, lumbar and sacroiliac radiographs were unremarkable. Results of arthrocentesis were consistent with an inflammatory arthropathy. Results of an

echocardiogram were normal. Fungal, viral (including HIV), bacterial and spirochete cultures were negative. Tuberculin purified protein derivative and pathergy skin testing also yielded negative results. The patient was positive for the HLA-B51 antigen. A skin biopsy revealed evidence of vasculopathy, including swelling of endothelial cells and perivascular inflammatory cell infiltrate (Fig. 4). Behçet's disease was diagnosed.

The patient received no specific therapy for Behçet's disease, and after 4 weeks all symptoms had completely resolved.

Behçet's disease is an inflammatory disorder, with recurrent attacks of acute multisystem vasculitis.¹ Prevalence is

DOI:10.1503/cmaj.061136



Fig. 1: Oral-labial aphtha.

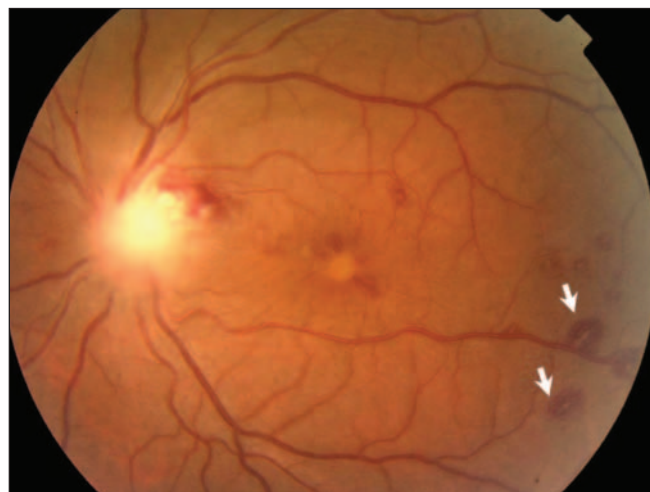


Fig. 2: Retinal hemorrhages with pale centres.



Fig. 3: Erythematous plantar maculae.

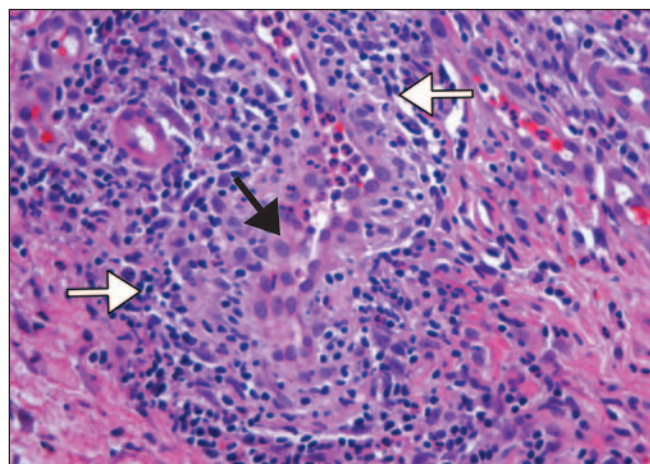


Fig. 4: Endothelial cell swelling (black arrow) and perivascular inflammatory cell infiltrate (white arrows).

highest among people of Middle Eastern or Far Eastern ancestry. Diagnosis of Behçet's disease is based on clinical criteria in the absence of alternative clinical explanations. Several sets of criteria have been proposed, but the most widely accepted are those of the International Study Group for Behçet's Disease.² According to their criteria, recurrent oral ulceration must be present as well as at least 2 of the following: recurrent genital ulceration, eye lesions, skin lesions or a positive pathergy test result. (In pathergy testing, a sterile 20–22-gauge needle is used to obliquely penetrate a vascular skin to a depth of 5 mm; the result is considered positive if an erythematous papule develops at the test site after 48 hours).

The features of Behçet's disease, however, are not limited to the diagnostic criteria: the syndrome can present in a myriad of ways and can involve nearly

every organ system. In fact, morbidity and mortality predominantly result from vasculitis in large vessels, cerebrovascular disease, gastrointestinal involvement (often difficult to differentiate from inflammatory bowel disease) and ocular involvement (can lead to blindness). Therapy is tailored to the specific organ systems involved but typically involves treatment with corticosteroids and other immunomodulatory agents. However, evidence for the effectiveness of many of the available treatments is scant or conflicting.³

Although Behçet's disease is perhaps less familiar to North American physicians than to those practising in the Middle or Far East, it is often included in the differential diagnosis for cases that are difficult to solve and involve multiple organ systems. Our patient's case illustrates several classic findings of Behçet's disease as well as

the importance of a thorough history and physical examination.

Devon R. McDonald

Christie Lee

Department of Medicine

Khaled Abuhaleeqa

Department of Ophthalmology

Robert A. Fowler

Department of Medicine

Sunnybrook Health Sciences Centre

University of Toronto

Toronto, Ont.

Competing interests: None declared.

REFERENCES

1. Sakane T, Takeno M, Suzuki N, et al. Behçet's disease. *N Engl J Med* 1999;341:1284-91.
2. International Study Group for Behçet's Disease. Criteria for diagnosis of Behçet's disease. *Lancet* 1990;335:1078-80.
3. Saenz A, Ausejo M, Shea B, et al. Pharmacotherapy for Behçet's syndrome. *Cochrane Database Syst Rev* 2000; (2):CD001084.

How you can get involved in the CMA!

The CMA is committed to providing leadership for physicians and promoting the highest standard of health and health care for Canadians. To strengthen the Association and be truly representative of all Canadian physicians, the CMA needs to hear from members interested in serving in elected positions and on appointed committees and advisory groups.

The CMA structure comprises both governing bodies and advisory bodies either elected by General Council or appointed by the CMA Board of Directors. The Board of Directors — elected by General Council — has divisional, affiliate, resident and student representation, is responsible for the overall operation of the CMA and reports to General Council on issues of governance. CMA councils and committees advise the Board of Directors and make recommendations on specific issues of concern to physicians and the public. Five core councils and committees consist of either divisional or regional representation, while other statutory and special committees, and task forces consist of individuals with interest and expertise in subject-specific fields. Positions on one or more of these committees may become available in the coming year.

For further information on how you can get involved, please contact:

Paula Wilson
Corporate Affairs
Canadian Medical Association
 1867 Alta Vista Drive
 Ottawa ON K1G 3Y6
 Fax 613 526-7570
 Tel 800 663-7336 x2047
involved@cma.ca

By getting involved, you will have an opportunity to make a difference.

We hope to hear from you!

ASSOCIATION
MÉDICALE
CANADIENNE



CANADIAN
MEDICAL
ASSOCIATION