What You Should Know About Behcet's Disease and the Eye

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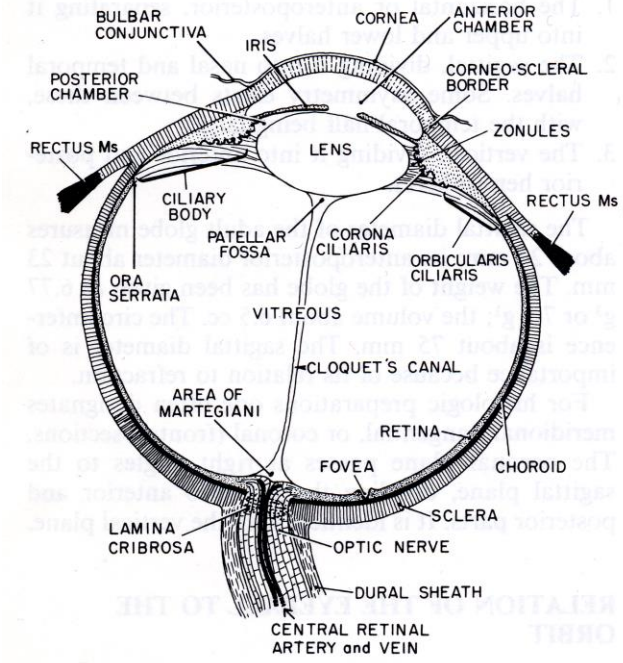
For purposes of explanation, the eye may be likened to a camera. Both contain a lens, which focuses incoming light and both have a thin film, which lines the back wall. In the case of the eye, this thin film is called the retina. It is nerve tissue capable of converting light into nerve signals, which travel to the brain via the optic nerve. The lens of the human eye is suspended by zonules, small cables located behind the iris, the colored tissue surrounding the black pupil. Figure 1 shows the structures of the human eye. This simplified explanation of the anatomy of the eye will be helpful as we turn now to Behcet's disease.

Figure 1. Anatomy of the Human Eye



What is Behcet's Disease?

Behcet's disease is an inflammatory disease that can affect many organ systems in the body. A typical case would cause oral and genital ulcers (see Figure 2) and inflammation of the eye, termed uveitis. A pooling of inflammatory white blood cells can accumulate in the anterior chamber, a clinical finding called hypopyon. Blood vessels in the retina may be inflamed, called vasculitis (see Figure 3). Behcet's disease can affect skin, joints, brain, heart, kidney, and gastrointestinal tract too. The prevalence of Behcet's disease varies with geographical region. In Turkey, the prevalence is 370 per 100,000 persons. In the United States, the prevalence is closer to 1 per 100,000 persons.[1]

Figure 2 Oral Ulcer in Behcet's Disease

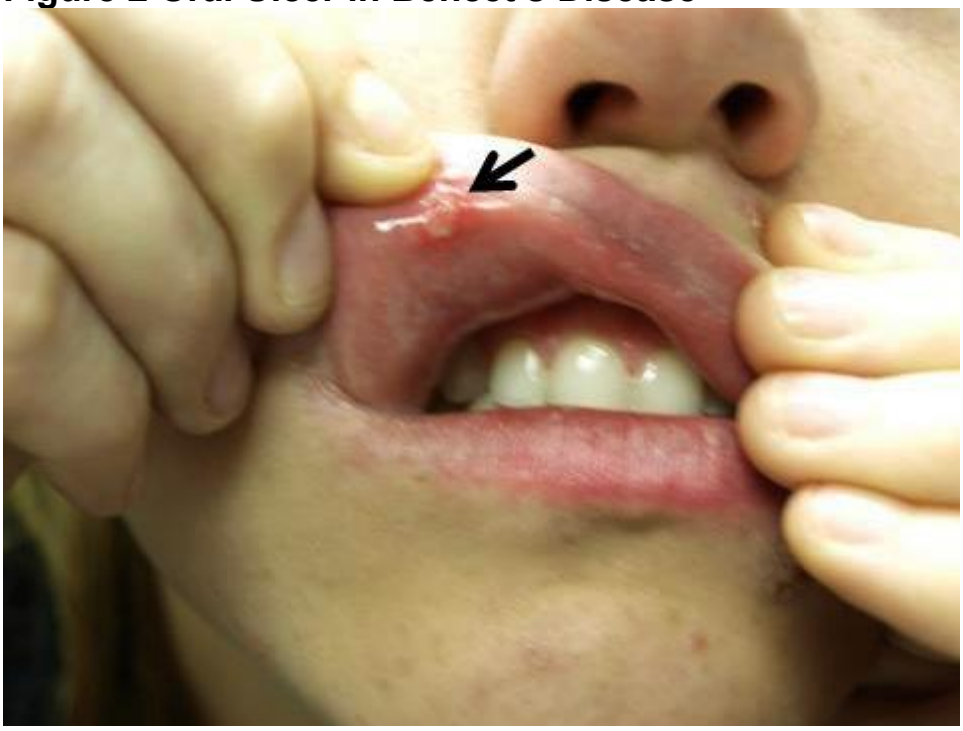
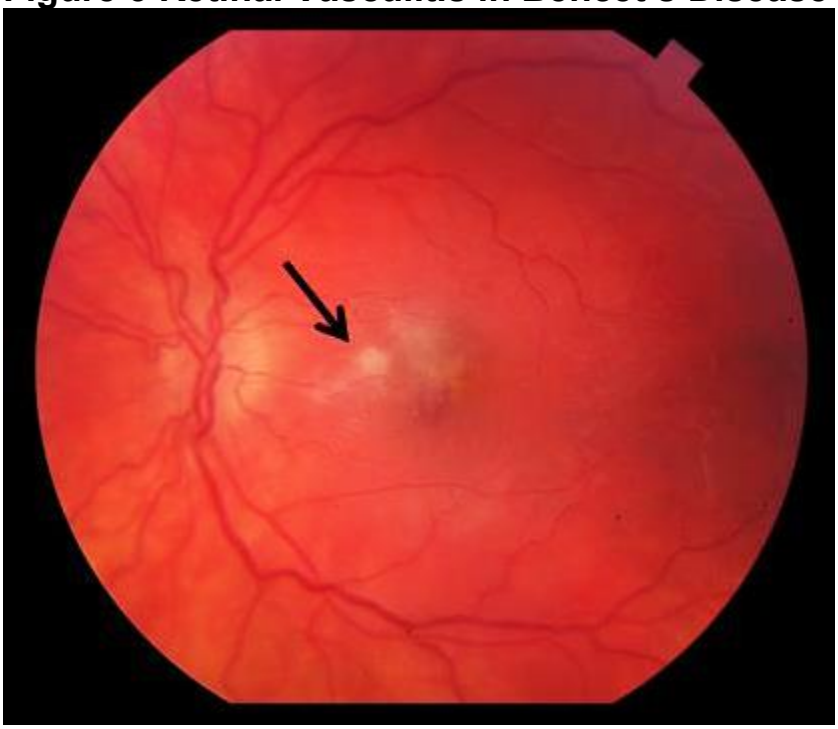


Figure 3 Retinal Vasculitis in Behcet's Disease



What Causes Behcet's Disease?

The cause of Behcet's disease is loss of tolerance of the body's immune system to its own cells. Why this happens is unknown, but the consequence is that the immune system attacks the body's tissues. There is a genetic predisposition to Behcet's disease. Patients with eye disease have a higher prevalence of the HLA B51 antigen subtype.[2]

What is the Treatment of Behcet's Disease?

The treatment of Behcet's disease involves corticosteroids (e.g. prednisone), antibodies to tumor necrosis factor α (e.g. infliximab), antimetabolite drugs (e.g.azathioprine), and alkylating drugs (e.g. cyclophosphamide). Prednisone is started immediately for its rapid onset of action. The other immune modulating drugs take longer to work, but have fewer side effects than prednisone. Dilating and steroid drops are often used for eye inflammation.[3]

What Is the Prognosis?

The visual outcome in Behcet's disease depends on how rapidly the diagnosis is made and effective therapy begun. Because Behcet's disease is rare in the United States, it may not be suspected initially. Treatment requires cooperation between the ophthalmologist and the internist who manages the systemic drugs. As a rule of thumb, with coordinated and timely care there is a 60-70% probablility of inducing remission and reducing prednisone to < 10 mg/ day within one year of onset of treatment. This 10 mg/day threshold is important for minimizing the adverse side effects of prednisone. [4]

Final Comments

Once you have read this brochure, if you would like to find more in-depth information about Behcet's disease, an excellent resource is the National Library of Medicine website section called PubMed at www.pubmed.com. It includes an extensive database of articles published in peer-reviewed medical journals from all over the world.

Reference List

1. James DG, Spiteri MA. Behcet's Disease. Ophthalmology 1982;89:1279-84.

2. Numaga J, Matsuki K, Mochizuki M, et al. An HLA-D region restriction fragment associated with refractory Behcet's disease. Am J Ophthalmol 2012;105:528-33.

3. Takeuchi M, Hokama H, Tsukahara R, et al. Risk and prognostic factors of poor visual outcome in Behcet's disease with ocular involvement. Graefes Arch Clin Exp Ophthalmol 2005;243:1147-52.

4. Lee RWJ, Dick AD. Current concepts and future directions in the pathogeneis and treatment of non-infectious intraocular inflammation. Eye 2012;26:17-28.

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