Behçet's syndrome, a major cause of posterior uveitis and visual loss in the Third World, has probably existed for more than 4000 years. It is a multisystem, inflammatory disorder whose principal manifestations are oral and genital aphthosis as well as inflammation of the eye, skin, and joints. The principal causes of death are from vascular and neurological involvement. No etiological agent has yet been identified for the disease; current evidence suggests that the normal flora of mucosal tracts induce immunological hyper-reactivity in genetically predisposed individuals. The visual prognosis in patients with Behçet's disease is poor, the principal cause of visual loss being consecutive inflammatory ischemic retinal vessel occlusions and macular edema, scar and optic atrophy. Treatment is directed at suppressing the inflammatory response using corticosteroids and a variety of second line immunosuppressive agents. Despite different treatment regimens used in different countries the visual prognosis is much the same, with a hard core of 15–20% of patients seemingly resistant to therapy and progressing relentlessly to blindness.¹

In this issue of the Iranian Journal of Ophthalmology Davatchi and coworkers have presented an important review on "Longitudinal Study of Chlorambucil in Ocular Manifestations of Behçet's Disease". The visual acuity of their 89 patients was improved in 68.4% of the eyes and the mean visual acuity was increased from 2.6 to 3.3 of Snellen chart after mean duration of treatment of 26.2±2.6 months by Chlorambucil.

In the past mamo JG² has reported that untreated ocular Behçet's disease ends up in blindness in few years of time.

Mamo and Azzam³ were the pioneers to treat the ocular Behçet's disease by immunosuppressors and particularly by Chlorambucil and they reported successful results. Ever since, several immunosuppressors and new generations of drugs have been applied in treating this blinding disease and in many cases positive results have been reported. Despite all the efforts and progresses in treating ocular Behçet's disease in a recent international report by Kitaichi et al.⁴ The visual outcome of ocular Behçet's disease has been reported to be unfavorable. This report has been collected from 25 Behçet's disease centers including 1465 ocular Behçet's disease patients and the final visual acuity of 23% of the cases has been reported to be less than 0.1 despite the extensive treatments.

Looking more carefully at the registry of Shariati hospital of Tehran (Behçet's unit) in a survey of August 2008 among 6021 Behçet's disease patients only 4.3% of the eyes were legally blind unilaterally (vision=0.1 or less) and 3.8% were blind bilaterally and after 10.9±7.8 years of evolution of Behçet's disease which make us to postulate that the response of the Behçet's disease patients to the treatments is different at different regions of the world or the severity of the disease is variable at different regions and in different areas.

In this present report of Davatchi et al although the visual acuity worsened in 31.6% of the eyes and among them 7.1% became blind (No-light-perception) the authors claim that the complications of the disease such as cataract, optic atrophy, macular scar, macular edema etc are responsible for this regression of visual acuity but the total adjusted disease activity index improved from 21.5 to 16.2 (P=0.0002) and 75.3% of patients had improvement of ocular inflammations.

It is of interest to emphasize that 7.8% of the eyes in this presentation have had no-light-perception and they have gained some useful vision which could be explained by the very severe disc and optic edema and prevention of passage of axonic flow.

Thanks to the effort and long time investigation of our colleagues nowadays the satisfactory results of cytotoxic drugs such as Chlorambucil have been proven and many new horizons are being opened in the treatment of ocular Behçet such as TNF, Interferon, Infliximab and Rituximab.
Despite many advances in the understanding of the immunoregulatory mechanisms of uveitis, the ideal immunosuppressant has yet to be found. The three main classes of immunosuppressives that are widely used today in addition to glucocorticosteroids are antimetabolites, T cell inhibitors and alkylating agents. Antimetabolites include azathioprine, methotrexate and mycophenolate mofetil (MMF). T cell inhibitors include cyclosporine and tacrolimus. Alkylating agents include cyclophosphamide and chlorambucil. Cyclosporine and azathioprine have been found to be effective in the treatment of Behçet’s disease in randomized controlled trials, whereas the efficacy of other agents is shown by uncontrolled case series.\(^5\)

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References