Interferon alfa-2a in the Treatment of Ocular Adamantiades-Behçet’s Disease

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1. INTRODUCTION

Adamantiades-Behçet’s disease (ABD) is a chronic relapsing multisystemic vasculitis involving small and large vessels. The etiology of the disease still remains unknown. Relapsing ocular involvement is one of the major manifestations of Adamantiades-Behçet’s disease, characterized by iritis (Fig. 1), uveitis, retinal occlusive vasculitis (Fig. 2), and optic nerve neuropathy that often lead to blindness if untreated. Retinal detachment and secondary glaucoma are severe complications of occlusive retinal vasculitis. Surgical intervention of these complications often leads to a recurrence of the inflammation limiting the surgical results. In the case of Adamantiades-Behçet’s disease with eye involvement, a combination of systemic corticosteroids and cyclosporine A is currently the treatment of choice, although severe side effects, such as Cushing syndrome, osteoporosis or renal failure and hypertension frequently occur. Interferon alfa (IFNα) has been shown to be an effective treatment in mucocutaneous ABD. It has also been suggested to improve ocular lesions. We examined the effects of interferon alfa-2a (6–9 Mio IU 3x/week) in a case series of 24 patients with ocular involvement.
2. PATIENTS AND METHODS

Since 1999 we included 24 consecutive ABD patients with ocular involvement who provided informed consent in an IFNα treatment protocol. The mean follow-up since initiation of the treatment was 18 months (6-36 months). The mean age at the onset of the disease was 28 years (18-38 years), whereas the beginning of ocular symptoms 30 years (23-40 years). At the time of initiation of the treatment all 24 patients showed oral ulcerations, whereas some patients presented genital ulcerations (n=7), arthritis (n=9), skin lesions (n=16), or cerebral involvement (n=1). All patients underwent a complete ophthalmologic examination including visual acuity measurement, slitlamp examination of the anterior segment, and indirect ophthalmoscopy of the vitreous body and fundus. We detected iritis in 20 of the 24 patients, 21 had ocular vasculitis, and 12 patients had a neuropathy of the optic nerve.

Patients received IFNα-2a (6-9 Mio IU 3x/week) subcutaneously as long-term therapy. Treatment was initiated at relapse of the eye disease. Corticosteroids (prednisolone 100 mg/day/p.o.) were administered additionally, and were tapered within 2 weeks to a maintenance dose of 10 mg/day. IFNα-2a was lowered to 6 and then to 3 Mio IU 3x/week, provided a 4-month period without ocular inflammation. After a 6-month period without ocular inflammation corticosteroids were discontinued.

Figure 1. Hypopyon iritis