High dose intravenous steroid therapy for severe posterior segment uveitis in Behçet’s disease

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**Aim:** To evaluate the safety and effectiveness of high dose intravenous steroid therapy (HDIST) in Behçet’s disease patients with severe posterior segment uveitis attacks.

**Methods:** Five patients with ocular Behçet’s disease were treated with HDIST for severe posterior segment attacks. Two patients had vasculitis, one patient had papillitis, and the other two had retinitis; four patients also had accompanying severe vitritis. The visual acuities and improvement in ocular signs after HDIST were evaluated.

**Results:** During HDIST, patients had no systemic complications caused by treatment. All patients responded to HDIST with evidence of a decrease in intraocular inflammatory activity and improved visual acuities within a mean time of 7 (1–15) days of commencing treatment. During follow up three out of five patients had new posterior segment attacks.

**Conclusion:** HDIST is effective in controlling severe, vision threatening acute posterior segment Behçet’s uveitis attacks and in improving visual function in a short period of time.

Posterior segment involvement, which is usually in the form of retinitis and retinal vasculitis in Behçet’s disease, is a significant cause of severe visual loss unless diagnosed early and treated properly. High dose intravenous steroid treatment (HDIST) is a form of steroid therapy recommended as an initial treatment in optic neuritis, for the treatment of uveitis in patients with multiple sclerosis, Vogt-Koyanagi-Harada disease, and other serious inflammatory diseases. Its efficacy in the prevention of severe visual loss in acute Behçet’s disease has also been reported.

We treated five patients with Behçet’s disease with acute posterior segment involvement with high dose intravenous methylprednisolone between May 1994 and March 1999. Patients with serious posterior segment attack (retinitis, vasculitis, papillitis, vitritis) that threatened the vision profoundly were selected for the treatment. All patients met the international diagnostic criteria for Behçet’s disease.

Patients were admitted to hospital and underwent a detailed medical examination. Intravenous methylprednisolone, 1 g, was administered over 1 hour for 3 days followed by initiation of oral methylprednisolone, 1 mg/kg/day, which was gradually tapered and then either stopped or continued at a maintenance dose of 10 mg/day. Adjunctive immunosuppressive treatment was initiated in patients who were not already on immunosuppressants.

**REPORT OF CASES**

**Case 1**
A 31 year old woman presented with acute visual loss in the left eye. She had a right ocular prosthesis (traffic accident). She had had the diagnosis of non-ocular Behçet’s disease for 3 years. Examination of the left eye revealed a visual acuity of 0.2 and an area of vasculitis at the temporal vascular arcade with accompanying vitritis and macular oedema. HDIST was administered followed by oral methylprednisolone. Azathioprine 2 mg/kg/day was initiated. After 15 days, vision in the left eye improved to 0.6 with regression of vasculitis and vitritis. The visual acuity improved to 1.0 by the first month. Oral methylprednisolone was gradually tapered and discontinued during the ensuing 4 months. During 6 years of follow up no new ocular attacks occurred. Immunosuppressive treatment was continued for 4 years to control her extraocular signs of Behçet’s disease. Since she has had no ocular or extraocular signs for the past 2 years immunosuppressive treatment has been discontinued.

**Case 2**
A 46 year old woman, diagnosed with Behçet’s disease 2.5 years ago, had a complaint of acute visual loss in the right eye. She had a history of several episodes of anterior uveitis in both eyes (seven times in the right eye, once in the left eye), all of which responded to topical steroid therapy. On examination her visual acuity was counting fingers at 1 foot in the right eye and 1.0 left eye. She had papillitis in the right eye. HDIST was administered followed by oral methylprednisolone. The visual acuity improved to 0.8 on the second day and to 1.0 after 3 days. At the end of the first week azathioprine 2 mg/kg/day was initiated. Oral methylprednisolone was tapered gradually and discontinued after 4 months. During 2.5 years of follow up she had papillitis once and anterior uveitis four times in the right eye while taking immunosuppressive treatment.

**Case 3**
A 52 year old woman, diagnosed with Behçet’s disease 3.5 years, presented with sudden decrease of vision in the left eye. She had a history of acute posterior uveitis attacks (six times) in the left eye and had been taking oral steroids and immunosuppressive treatment (cyclosporine A 200 mg/kg/day, azathioprine 100 mg/day). Her visual acuity was 0.7 right eye and hand movement left eye. Examination of the left eye disclosed vasculitis and accompanying vitritis. She had posterior subcapsular cataracts in both eyes. HDIST was administered followed by oral methylprednisolone. By the ninth day her vision was counting fingers at 1 foot with an improvement of posterior segment inflammation. At the end of the second week her visual acuity was 0.2, which had been recorded to be the best visual acuity during the remission period. Oral methylprednisolone was tapered gradually over 2 months and continued at the maintenance dose. Meanwhile she had been taking azathioprine and cyclosporine at the same doses. After she discontinued oral methylprednisolone by mistake she had a recurrence of the attack and presented with a visual acuity of counting fingers at 1.5 feet. On examination she had papillitis, retinitis, and severe vitritis. She was administered HDIST for the second time. The visual acuity in her left eye improved minimally by the fourth day (counting
The visual acuity of the left eye was 0.3 right eye and the funduscopy examination he had retinitis and vitritis with a visual acuity of 0.05 right eye and 0.2 left eye (Fig 1). He was diagnosed with Behçet’s disease based on recurrent oral aphthous ulcers, folliculitis, uveitis, and HLA-B51 positivity. HDIST was administered followed by oral methylprednisolone. Azathioprine was continued at the same dose.

In conclusion, we recommend HDIST as an initial therapy in severe acute posterior segment uveitis attacks in Behçet’s disease with substantial visual impairment for prompt control of the inflammation and faster recovery of visual function. In our opinion, HDIST spares the potential side effects of high dose oral steroid treatment. In such cases of Behçet’s disease with severe ocular involvement, the initial dosages of oral steroids which are started for the control of acute inflammation frequently exceed 100 mg prednisone per day. The initiation doses are higher and the tapering period is definitely

**DISCUSSION**

The clinical course of Behçet’s disease is characterised by exacerbations and remissions of unpredictable duration and frequency. The retinal disease is the most disabling manifestation of Behçet’s disease. Attacks of inflammation may occur as frequently as every few weeks. These recurrent retinal vascular occlusive episodes ultimately lead to irreversible alterations and widespread retinal atrophy. Vision in patients with untreated Behçet’s disease is usually lost, on average, 3-4 years after onset of eye symptoms.5-8

Many different treatment schedules have been tried in Behçet’s disease. Corticosteroids (topical, periorcular injection and systemic routes) have a beneficial effect on the acute manifestations of inflammation.9 Though the disease may initially respond to treatment with systemic corticosteroids, it will invariably become resistant to a pale optic disc. The visual acuity improved to counting fingers at 5 feet and by the 12th day it increased to 0.3 with resolution of retinitis. Oral methylprednisolone dose was tapered over 4 months and continued at the maintenance dose of 10 mg/day for 3 months. During 3 years of follow up he had retinitis once in the left eye, but HDIST was not considered necessary as the visual acuity was not significantly affected.

Fingers at 5 feet and after 7 weeks it was 0.05. She has been taking two immunosuppressive agents and oral steroid at the maintenance dose without any new attacks for the last 6 months.

**Case 5**

A 29 year old man who had the diagnosis of ocular Behçet’s disease and had been taking azathioprine 100 mg/day for a year presented with a sudden decrease of vision in the left eye. The visual acuity was 0.3 right eye and the funduscopy revealed a pale optic disc. The visual acuity of the left eye decreased to counting fingers at 2 feet which had been recorded to be 0.3 previously, and the examination disclosed retinitis and vitritis. He also had bilateral posterior subcapsular cataracts. HDIST was administered followed by oral methylprednisolone. Azathioprine was continued at the same dose. By the sixth day the visual acuity improved to counting fingers at 5 feet and by the 12th day it increased to 0.3 with resolution of retinitis. Oral methylprednisolone dose was tapered over 4 months and continued at the maintenance dose of 10 mg/day for 3 months. During 3 years of follow up he had retinitis once in the left eye, but HDIST was not considered necessary as the visual acuity was not significantly affected.
longer than the oral steroid regimen that we start after HDIST, thus making the patient more prone to long term side effects of steroid therapy. Besides, our clinical impression is that the response to high dose oral steroids is not as immediate as in HDIST. We initiate adjunctive immunsuppressive therapy for controlling the disease long term if the patient is not on immunosuppressants. Azathioprine is our drug of choice in the treatment of severe Behçet’s uveitis. The onset of action of immunosuppressants, particularly azathioprine, is delayed and benefit begins 6–12 weeks after starting the medication.\textsuperscript{16} For this reason, oral steroids should be tapered rather slowly during this period. Azathioprine with its steroid sparing effect allows the dosage of oral steroids to be lowered to a more acceptable level.

References
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