Bilateral Simultaneous Optic Neuritis in Childhood Systemic Lupus Erythematosus

A Case Report

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The authors describe an 11-year-old girl with systemic lupus erythematosus (SLE) who developed simultaneous bilateral acute optic neuritis. Severe initial visual loss followed by permanent visual deficit occurred in both eyes despite therapeutic intervention. Recurrence of optic neuritis in one eye caused the vision to deteriorate further. The most probable pathogenesis is occlusive vasculitis involving the small arterioles of the optic nerves.

Key Words: Systemic lupus erythematosus—optic neuritis.

Systemic lupus erythematosus (SLE) is a relatively rare illness during childhood. However, in children it is more acute and severe than in adults. SLE is a systemic disease that affects many organ systems. A variety of ophthalmic manifestations may be seen in SLE, but involvement of the optic nerve in the course of this autoimmune disease is rare. We report the occurrence of simultaneous bilateral acute optic neuritis in a girl with systemic lupus erythematosus that led to permanent severe visual loss in both eyes despite therapeutic intervention.

REPORT OF A CASE

An 11-year-old girl was admitted to the pediatric department in February 1989, with malaise, fever, respiratory distress, and substernal chest pain. While reviewing her past medical history, Raynaud’s phenomenon and periorbital edema were noted. A maculopapular rash was observed on her face upon physical examination. Chest pain was of the pleuritic type. Pericardial friction rub was heard. Splenomegaly was present as well. In the interim, polyarthritis, involving the right knee, the left ankle, and the small joints of the hands and feet, occurred.

Laboratory data showed normal complete blood count; erythrocyte sedimentation rate, 65 mm/h (Westergren); a positive fluorescent antinuclear antibody (FANA) test of a titer 1/1600 with a speckled pattern; anti-double-stranded DNA antibodies, 34.7 U/ml with radioimmunoassay technique (normal: <6 U/ml). The C3 level was normal, but the C4 level was low, at less than 7 mg/dl (normal: 16–45 mg/dl). Lupus erythematosus cells were not
In a recent attack of decreased vision in her right eye, the patient was lost to follow-up for about 1 year, when she returned with complaint of a recent attack of decreased vision in her right eye. Visual acuity of the right eye had deteriorated to counting fingers at 50 cm, but vision of the left eye remained the same as before. There was 2+ relative afferent pupillary defect on the right side and a sluggish pupillary reaction of the left. Both optic discs were flat, while very pale. The visual field revealed persistence of dense central scotomata OU. Malaise, fever, and polyarthritus occurred concomitant with a recurrent attack of optic neuritis in the right eye. Again, a treatment of high-dose oral prednisolone was started, but no improvement occurred in vision of the right eye. Visual-evoked response testing revealed severe abnormality as marked decrease of amplitude in both eyes.

DISCUSSION

Central nervous system involvement occurs in more than one-third of patients with systemic lupus erythematosus (1–3). Optic neuritis in SLE is uncommon (4–16), but can be the initial manifestation (5,7). Among the previously reported cases of optic neuritis in SLE, only two patients were under 12 years old (4,5). Optic neuritis was bilateral in both of these patients; final visual acuity was 20/60 OD and 20/200 OS in the former (4), a 10-year-old female, and light perception OU in the latter (5), an 11-year-old female. The diagnosis of SLE was made in our case, based on the 1982 revised criteria for the classification of systemic lupus erythematosus (17).

Optic neuritis in SLE may present as either papillitis (5,7) or retrobulbar neuritis (10,12). In our patient, although the optic neuritis was dominantly retrobulbar, mild swelling of the disc showing some involvement of the optic nerve head was observed as well.

The main pathogenesis of optic neuritis in SLE is one of a vaso-occlusive disease in small vessels (1). Visual evoked potential testing data in most patients shows the loss of amplitude and is compatible with ischemia to the optic nerve (18). In cases with pure demyelination, only an increase in latency is observed. Jabs and colleagues (12) believe that, in those cases with only demyelination, the demyelination is caused by a milder form of ischemic disease in small vessels (12). In more severe cases however, axonal damage and necrosis occur as well. A general autopsy was done in a known case of SLE with optic neuritis and associated transverse myelitis, sereosis, and organic brain syndrome; the patient died a few days after the onset of coma (5). Gross pathologic examination showed edema of the brain and randomly distributed cysts and areas of softening in both gray and white matter throughout the entire spinal cord. The involved optic nerve was small and sclerotic. The microscopic pathologic findings included endothelial hyperplasia, subintimal thickening of collagen and focal fibrinoid necrosis of arterioles and small arteries associated with perivascular lymphatic and plasma cell infiltration in the spinal
cord and the involved optic nerve. The optic nerve showed marked gliosis and loss of myelin and axons as well (5). Two other postmortem histologic examinations (6,8) confirmed the presence of severe optic nerve demyelination and necrotic myelopathy with normal axons in one patient (8), and moderate to severe focal axonal loss in the other (6).

Visual outcome in SLE optic neuritis is often poor (4-14). In our patient also, a severe drop in visual acuity of both eyes occurred during the first attack, which poorly responded to high-dose oral steroid therapy. Moreover, deterioration of vision in the right eye occurred a few months later, due to a recurrent attack of retrobulbar optic neuritis. The abnormal visual evoked potential in our patient was compatible with the degree of visual loss.

There have been some reports regarding the beneficial effect of intravenously administered "pulse" methylprednisolone in autoimmune optic neuropathy (19,20). The patients, all women except two, with an age range from 26 to 56, suffered from progressive or recurrent optic neuropathy leading to severe visual loss, and none responded to conventional doses of corticosteroid. Only laboratory evidence of collagen vascular disease was present, and none of the patients had overt clinical signs of collagen vascular disease nor fitted neatly into a specific diagnostic category, such as systemic lupus erythematosus. Megadose steroid therapy improved the vision in 11 of 12 patients in one series (20) and 3 of 4 patients in the other series (19). Nevertheless, continued oral administration of prednisolone and cytotoxic drugs were necessary to maintain the vision in most patients.

Considering the results of treatment of optic neuritis with megadose corticosteroids, Spoor (21) suggested a trial of intravenous megadose steroids in patients between the ages of 21 and 45 with optic neuropathies of suspected autoimmune etiology.

Recently, the results of an optic neuritis treatment trial have been published (22). It was concluded that intravenous methylprednisolone followed by oral prednisone speeds the recovery of visual loss due to optic neuritis and results in slightly better vision at 6 months, compared with the placebo group. However, the results of this study could not be applied directly to the cases of autoimmune neuropathy because the patients who had clinical evidence of a systemic disease other than multiple sclerosis were excluded.

Further study is needed to identify the optimal therapy for optic neuritis in systemic lupus erythematosus.

REFERENCES