Abstract

Objective—To evaluate the prevalence and clinical profile of patients with biopsy-proven arteritic anterior ischemic optic neuropathy presenting with preserved visual acuity of 20/40 or better and those with an initial poor visual acuity of 20/50 or worse through a retrospective chart review

Results—Nine of 37 patients with arteritic anterior ischemic optic neuropathy presented with a preserved visual acuity of 20/40 or better in the affected eye. All patients with preserved visual acuity had initial visual field defects that spared the central field. All 37 patients immediately received high-dose corticosteroid therapy. Visual acuity worsened by > 2 lines in one of nine patients (11%) with preserved visual acuity, with a corresponding progression of visual field constriction.

Conclusion—Although preserved visual acuity of 20/40 or better has traditionally been associated with the nonarteritic form of anterior ischemic optic neuropathy, giant cell arteritis should still be strongly considered, especially if they have giant cell arteritis systemic symptoms.

Introduction

The visual sequelae of giant-cell (temporal) arteritis are often dramatic and irreversible; nonetheless, a minority may have minimal visual dysfunction, potentially deflecting attention from the correct diagnosis. Since early diagnosis and treatment of giant-cell arteritis is crucial in preventing blindness, we report our experience in giant-cell arteritis patients who presented with preserved visual acuity of 20/40 or better.

Patients and Methods

Medical records of all patients with anterior ischemic optic neuropathy secondary to biopsy-proven giant-cell arteritis seen in the neuro-ophthalmology clinics at the Michigan State University and at the Baylor College of Medicine were reviewed. Temporal artery biopsies were considered positive if there was disruption of the internal elastic lamina with infiltration of mononuclear cells in the arterial wall, with or without giant cells [1]. Subjects were excluded if they had concomitant non-giant cell arteritis-related retinal or other optic nerve pathology, or incomplete follow-up data. All patients were seen by a neuro-ophthalmologist, had a detailed ocular and medical history, as well as a comprehensive neuro-ophthalmologic evaluation. The primary outcome measure was the best corrected Snellen visual acuity on their initial and final clinic visits. Improvement or worsening of visual acuity on final follow-up was defined as a change of more than two Snellen lines, except in the following circumstances: (1) if the initial visual acuity was counting fingers, a follow-up visual acuity of 20/200 or better was defined as an improvement, while a follow-up visual acuity of hand motion or worse was defined as worsening; and (2) any follow-up visual acuity change to light perception or no light perception was considered as worsening visual acuity. Color vision testing (Ishihara or AO Hardy-Rand-Rittler pseudoisochromatic plates) and visual fields testing using either kinetic (Goldmann, Haag-Streit) or static (Humphrey visual field analyzer, Humphrey Instruments, San Leandro, CA) perimetry were also performed. Color vision was documented as a quotient of the number of correct color plates divided by the total number of color plates shown (with 1 as perfect). Visual field defects were classified into either of the following predominant patterns: arcuate, altitudinal, central scotoma, or generalized constriction. Final Goldmann or Humphrey visual fields were defined as stable if they were within 15° of the initial visual fields and without the development of a new scotoma within the central 15° of vision. The following data were also recorded from the medical charts: giant-cell arteritis systemic signs and symptoms (e.g., headache, scalp tenderness, jaw or tongue claudication, diplopia, myalgia, arthralgia, anorexia, weight loss, numbness), history of...
systemic steroid or aspirin intake prior to the onset of giant-cell arteritis symptoms, erythrocyte sedimentation rate on their initial and final clinic visit, and the type and duration of giant-cell arteritis therapy (e.g., intravenous methylprednisolone, oral prednisone, steroid-sparing anti-inflammatory medications, aspirin).

Patients were divided into two subgroups for analysis: the preserved visual acuity group (Snellen visual acuity of 20/40 or better) and the poor visual acuity group (Snellen visual acuity of 20/50 or worse). Continuous data were expressed as mean ± SD, while categorical variables were expressed as percentages. Student’s two-tailed t-test for continuous variables was performed using the SPSS software, version 17.0 (SPSS Inc, Chicago, IL). Approval for the study was obtained from the respective institutional review boards and was compliant with the Health Information Portability and Accountability Act.

Results

Demographics

A total of 37 patients (43 eyes) with arteritic anterior ischemic optic neuropathy was included in our study, 76% from Michigan State University and 24% from Baylor College of Medicine; nine patients (9 eyes) had a visual acuity of 20/40 or better (preserved visual acuity group) and 28 patients (34 eyes) had a visual acuity of 20/50 or worse (poor visual acuity group). There was no significant difference in the age at diagnosis (preserved visual acuity mean age of 75 years, SD = 6 years versus poor visual acuity group mean age of 76 years, SD = 6 years, \( p = 0.68 \)). None of the patients had polymyalgia rheumatica nor were on systemic steroids prior to the onset of giant cell arteritis symptoms. Three of the patients in the preserved visual acuity group were on aspirin prior to the onset of giant cell arteritis symptoms, while there were nine in the poor visual acuity group.

Giant-Cell Arteritis Signs and Symptoms

The patients with preserved visual acuity were referred to our clinic because of blurred vision (67%), peripheral vision loss (33%), amaurosis fugax (11%), or diplopia (11%). Aside from visual loss, eight of nine patients (89%) in the preserved visual acuity group and 26 of 28 patients (71%) in the poor visual acuity group presented with systemic giant-cell arteritis symptoms (Table). Although the duration of giant-cell arteritis symptoms prior to visual loss was shorter in the preserved visual acuity group (mean = 18.9 ± 15.85 days) compared to the poor visual acuity group (mean = 33.5 ± 35.72 days), this was not statistically significant (\( p = 0.98 \)).

Initial Clinical and Laboratory Findings

In the poor visual acuity group, 32 of 34 eyes (94.1%) had a visual acuity of 20/200 or worse. Initial color vision was significantly better in the preserved visual acuity group (mean = 0.77 ± 0.24) compared to the poor visual acuity group (mean = 0.05 ± 0.14), \( p < 0.0001 \) (Table). In the preserved visual acuity group, initial color vision was not significantly different between the affected eye (mean = 0.77 ± 0.24) and the fellow eye (mean = 0.85 ± 0.18), \( p = 0.44 \); in the poor visual acuity group, initial color vision was significantly worse in the affected eye (mean = 0.05 ± 0.14) compared to the fellow eye (mean = 0.72 ± 0.32), \( p < 0.0001 \). All patients in this series had visual field defects at presentation (Table), although central vision was spared in patients with preserved visual acuity (Figure).

A trend toward initial high erythrocyte sedimentation rate was seen in the poor visual acuity group (mean 82.15 ± 36.97 mm/hr) compared to the preserved visual acuity group (mean = 61.44 ± 33.82 mm/hr), but this was not significant, \( p = 0.14 \).

Final Clinical and Laboratory Findings

Final parameters of visual function were measured after a mean follow-up of 3 years (range = 0.17–14.9). None of the patients with preserved visual acuity improved, while the final VA in five of 34 eyes (14.7%) in the poor visual acuity group improved by at least two Snellen lines. Final visual acuity worsened by more than two Snellen lines in one of nine eyes (11.1%), from 20/20 to 20/40, in the preserved visual acuity group and in nine of 34 eyes (14.7%) in the poor visual acuity group. Despite high-dose corticosteroid treatment, 27 of 34 eyes (79.4%) with an initial poor visual acuity had a final visual acuity of 20/200 or worse. Final color vision was significantly better in the preserved visual acuity subcohort (mean = 0.80 ± 0.22) compared to the poor visual acuity subcohort (mean = 0.14 ± 0.25), \( p < 0.0001 \). Color vision did not significantly change from initial to final visit in either group (preserved visual acuity group \( p = 0.76 \), poor visual acuity group \( p = 0.13 \)). Visual fields improved by at least 15 degrees compared to the initial visual fields in three of nine eyes (33.3%) in the
preserved visual acuity group, and none improved in the poor visual acuity group (Figure). Of the five eyes from the poor visual acuity group whose visual acuity improved on follow-up, none had a corresponding improvement in their visual fields or in their color vision. All patients had residual visual field defects. Progression of peripheral visual field constriction by more than 15 degrees was seen in two of nine preserved visual acuity eyes (22.2%) and in two of 34 poor visual acuity eyes (5.9%). In the whole series, two patients from the poor visual acuity group had subsequent involvement of the fellow eye; in one patient, the fellow eye involvement occurred within days of corticosteroid administration.

A trend toward lower erythrocyte sedimentation rate on the last clinic visit was seen in the preserved visual acuity group (mean = 14.78 ± 12.87), although this was not significantly different from the poor visual acuity group (mean = 24.85 ± 23.06), *p* = 0.12.

### Treatment

Treatment was administered to all patients in this series. High-dose oral prednisone was given to all patients, intravenous methylprednisolone to 17 of 37 (46%) patients, oral methylprednisolone to one patient (3%), steroid-sparing agent (i.e., methotrexate, plaquenil) to six patients (16%), aspirin (81 to 325 mg daily) to 19 patients (51%), and aspirin/dipyridamole to one patient (3%).

### Discussion

In our series of 37 patients with anterior ischemic optic neuropathy secondary to biopsy-proven giant-cell (temporal) arteritis, 21% had an initial preserved visual acuity of 20/40 or better. This is similar to other studies investigating giant cell arteritis-related visual loss (21–23%), although these studies also included patients with retinal, choroidal, or cortical ischemia [2,3]. All of our patients with preserved visual acuity had visual field defects that spared the central vision, thus preserving...
visual acuity and color vision. Jaw claudication was the most common nonvisual giant-cell arteritis symptom.

Patients with preserved visual acuity showed a trend toward a shorter delay to diagnosis compared to those with poor visual acuity suggesting that the diagnosis may have been made earlier in the preserved visual acuity subcohort, before the ischemia could have progressed and worsened.

All patients were immediately treated with parenteral or oral high-dose steroids. One patient developed a subsequent arteritic anterior ischemic optic neuropathy in the fellow eye within days of parenteral high-dose corticosteroid initiation, while the visual acuity deteriorated by
more than two Snellen lines in 23% of eyes. High-dose corticosteroid therapy takes approximately one week to abort the arteritic process in the posterior ciliary artery walls; studies have shown that any visual deterioration often occurred within the first week of corticosteroid initiation [4–8]. Hayreh and Zimmerman [4] postulated that steroid effect latency and optic nerve head hypoperfusion could be responsible for worsening vision despite high-dose steroids. This study emphasizes that treatment is not aimed at improving vision, but rather at preventing further visual deterioration in both eyes and in controlling the systemic vasculitic process. The clinician should make patients aware of the possibility of worsening vision at the time of initial diagnosis, despite high-dose steroids.

Although the clinical suspicion for giant-cell arteritis may be lower in patients with anterior ischemic optic neuropathy and visual acuity of 20/40 or better, evaluation for giant-cell arteritis should still be performed (i.e., temporal artery biopsy). Other indices should be taken into consideration (i.e., age > 50 years old, jaw or tongue claudication, new headache, abnormal visual fields, elevated erythrocyte sedimentation rate and C-reactive protein (CRP) [9–10]). These patients should be treated promptly to prevent irreversible visual loss and other vasculitis-related comorbidities.

References