Idiopathic Chiasmal Neuritis

Clinical Features and Prognosis

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Objectives: To describe the clinical features of idiopathic chiasmal neuritis in a large cohort of patients and to report their visual and neurologic outcomes.

Design: A retrospective medical record review of consecutive patients with chiasmal neuritis at a single institution. Patients with clinical or radiographic evidence of inflammation involving the intraorbital optic nerve and patients with a systemic inflammatory or neoplastic disorder were excluded.

Results: Twenty patients were identified (14 female, 6 male; mean age, 37 years). Visual acuity at initial examination ranged from 20/15 to light perception. Progressive visual loss beyond 1 month was documented in 1 patient. Twelve of 15 patients who underwent magnetic resonance imaging demonstrated chiasmal enlargement and/or enhancement; 6 patients had 1 or more white matter lesions. Follow-up time ranged from 2 weeks to 22 years, with a mean of 5.7 years. The final median visual acuity was 20/20 (range, 20/15-20/50) and visual fields were normal or improved. Of 15 patients with a minimum follow-up interval of 1 year, 6 developed multiple sclerosis.

Conclusions: The demographic and clinical features of idiopathic chiasmal neuritis resemble those of idiopathic optic neuritis. Visual prognosis is excellent. In this series, 40% of patients subsequently developed multiple sclerosis.

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CHIASMAL NEURITIS IS A clinical syndrome consisting of acute visual loss with a chiasmal visual field pattern and/or radiographic demonstration of chiasmal inflammation. Occasional cases of chiasmal neuritis are due to a systemic inflammatory disease, such as tuberculosi, sarcoidosis, systemic lupus erythematosus, Epstein-Barr disease, or Lyme disease. More commonly, chiasmal neuritis occurs as an idopathic event, either in isolation or in the setting of multiple sclerosis (MS). The clinical course of visual loss in such patients is consistent with demyelinating disease.

The clinical features of idiopathic optic neuritis have been well described, including the risk of development of MS in affected individuals. In contrast, idiopathic chiasmal neuritis is a rare condition whose clinical profile is not as well characterized. Individual case reports and small case series suggest it is a benign entity with a favorable visual outcome, but there are no longitudinal studies that address the issue of subsequent MS. The purpose of this study was to examine the clinical features and outcomes (visual and neurologic) of idiopathic chiasmal neuritis in a large cohort of patients observed at a single institution over a long period.

Methods

This is a retrospective medical record review of consecutive patients with a diagnosis of chiasmal neuritis who were examined and followed up by the authors from 1983 to 2001 at Indiana University and the Midwest Eye Institute. We defined chiasmal neuritis as a syndrome of acute visual loss associated with bitemporal or junctional pattern of visual field loss in which a compressive or hemorrhagic lesion of the chiasm was excluded radiographically.

We determined patients to have the idiopathic variety of chiasmal neuritis if they had no history or other evidence of an infectious, inflammatory, systemic, or neoplastic disorder known to affect the optic chiasm. We excluded patients with optic neuritis in whom enhancement of the intraorbital optic nerve extended to the chiasm. Patients with optic disc edema in either eye at the time of visual loss were also excluded.

We recorded the following information at the first visit for all patients: age; sex; symptoms; ocular, general medical, neurologic, and family histories; and past and current medications. All patients underwent a complete neuro-ophthalmologic examination. At subsequent follow-up visits, new or evolving symptoms, visual acuity, color vision, visual field, and pupill examination and fundus examination results were recorded. Results of all laboratory tests, radiologic studies, and other investigations were noted when available.
### RESULTS

#### DEMOGRAPHIC DATA

Twenty patients were included in the study. There were 6 males and 14 females. All patients were white except for 2 African American women. Age at the time of diagnosis ranged from 13 to 73 years (mean, 37 years; median, 34.5 years). The demographic and background medical details of all patients are given in Table 1.

#### OCULAR HISTORY

Four patients had subnormal baseline acuity in 1 eye owing to preexisting ocular conditions (macular scar in 2 patients, amblyopia in 1 patient, and cataract in 1 patient...)

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<table>
<thead>
<tr>
<th>Patient No./Sex/Age, y</th>
<th>Time From Symptom Onset to Initial Visit, wk</th>
<th>Symptoms</th>
<th>Ocular</th>
<th>Medical and Neurologic</th>
<th>Family</th>
</tr>
</thead>
<tbody>
<tr>
<td>1/F32</td>
<td>10 d</td>
<td>Progressive distortion in the right eye</td>
<td>Macular scar in the left eye</td>
<td>Migraine</td>
<td></td>
</tr>
<tr>
<td>2/F34</td>
<td>4</td>
<td>Changes on Amsler test in the left eye without metamorphopsia</td>
<td>Macular scar in the right eye followed by Amsler grid testing</td>
<td></td>
<td></td>
</tr>
<tr>
<td>3/M13</td>
<td>10</td>
<td>Blur and pain with movement in the right eye, Uhthoff symptom, associated flu-like illness</td>
<td>Sinus drainage</td>
<td>Maternal cousin with MS</td>
<td></td>
</tr>
<tr>
<td>4/M45</td>
<td>3</td>
<td>Blur in the right eye, cannot read out of left eye</td>
<td>Tobacco, DM</td>
<td></td>
<td></td>
</tr>
<tr>
<td>5/F24</td>
<td>4</td>
<td>Pain with movement in the left eye, HA then fuzzy vision</td>
<td>DM, heart disease, epilepsy</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6/F31</td>
<td>3</td>
<td>Bright, light-blocking central vision in the left eye</td>
<td>Had feeling of being pushed forward that lasted a few wk 5 y ago</td>
<td>Heart disease</td>
<td></td>
</tr>
<tr>
<td>7/M41</td>
<td>8</td>
<td>Visual loss for 2 mo in the right eye, photopsias in the right eye, 3-wk pain in the left eye</td>
<td>Had feeling of being pushed forward that lasted a few wk 5 y ago</td>
<td></td>
<td></td>
</tr>
<tr>
<td>8/M31</td>
<td>6</td>
<td>Blur in the left then right eye</td>
<td>Childhood esotropia and amblyopia in the right eye</td>
<td>DM</td>
<td></td>
</tr>
<tr>
<td>9/F45</td>
<td>6</td>
<td>Photophobia and glare in the right eye, then blur</td>
<td>DM</td>
<td>HTN, DM Breast CA</td>
<td></td>
</tr>
</tbody>
</table>
| 10/F60                 | 2-3                                         | Bright light flashing and acute visual loss in the left eye then blur in the right eye | Optic neuritis in the left eye (with disc edema | HTN diagnosed at time of visual loss 
| 11/F18                 | 2                                           | Painless visual loss in the right eye | Cleft palate repair | HTN, DM, CAD |
| 12/F73                 | 2-3                                         | Difficulty seeing television and reading, HA | Cataract in the right eye | Migraine, HTN, CAD, renal artery stenosis, post renal graft |
| 13/F25                 | 5                                           | Blur in both eyes with blind spots | Rumbliness of right tarsus 4 wk before visual loss | DM, stroke, breast CA |
| 14/M49                 | 2                                           | Dark spots in both eyes, progressive | | Aunt with MS |
| 15/F33                 | 3                                           | Grayish fog in the right eye that progressed to both eyes across 3 wk | Hyperlipidemia | Stroke |
| 16/F42                 | 5                                           | Progressive blur in the right eye for 4 wk, which moved to both eyes with further progression for 2 wk, pain | 2 Episodes of painless blur in the right eye 13 y and 2 y before chiasmal neuritis, treated each time with prednisone | Stroke |
| 17/F35                 | 10 d                                        | Progressive fogginess in the right eye with bright sparkles | | |
| 18/F41                 | 3                                           | Progressive dark haze in the center of the left eye | | |
| 19/M45                 | 3                                           | Fuzzy then dark vision in the right eye | 2 Prior episodes of visual loss in the left eye that resolved without treatment | Chronic HA |
| 20/F29                 | 2                                           | Dark in the right then left eyes | | |

Abbreviations: CA, cancer; CAD, coronary artery disease; DM, diabetes mellitus, HA, headache; HTN, hypertension; MS, multiple sclerosis.
tient). Three other patients had previously experienced monocular visual loss that later recovered. In one of these patients, visual loss was accompanied by disc edema and was diagnosed as optic neuritis. In the second patient, 2 episodes of painless visual loss occurred 13 years and 2 years before the chiasmal neuritis, each episode having been treated with prednisone. The third patient also reported 2 prior episodes of monocular visual loss that spontaneously recovered, though no diagnosis or treatment had been offered.

NEUROLOGIC AND FAMILY HISTORY

One patient reported a transient episode of numbness around the torso that preceded visual loss. No patient had a diagnosis of MS. Two patients had a close family member with the disease.

ACUTE EPISODES

Visual loss was monocular in 8 patients and bilateral simultaneous or rapidly sequential in 10 patients. Two patients had preexisting visual loss in 1 eye that was severe enough to preclude this distinction (left eye in patient 1; right eye in patient 2). Only 4 of 20 patients (20%) reported eye pain. Three patients had accompanying photopsia and 2 patients experienced glare and/or photophobia. The duration of symptoms before our first examination ranged from 10 days to 2.5 months, with an average of 26 days. Only 1 patient had documented progression of visual loss 1 month from symptom onset (patient 16). However, the history or follow-up was insufficient to make this determination in 3 patients (patients 2, 15, and 18).

At the initial visit, visual acuity ranged from 20/15 to light perception OD (median, 20/80 OD) and 20/15 to hand motion OS (median, 20/40 OS). In 11 of 38 eyes, visual acuity was 200/200 or worse; 2 eyes were excluded owing to poor baseline vision (20/200 and counting fingers) from preexisting ocular disease. In 1 patient, visual acuity was reduced to 20/200 OU at the initial visit.

The pattern of visual field loss was a bitemporal hemianopia in 8 patients and junctional scotoma in 12 patients. Fundus examination did not reveal any retinal or disc pathology that could explain the visual loss.

LABORATORY AND RADIOGRAPHIC FINDINGS

Laboratory investigations included measuring complete blood count, angiotensin-converting enzyme, lysozyme, syphilis serologies, Westergren erythrocyte sedimentation rate, antineutrophil antibodies, and antineutrophilic cytoplasmic antibodies. One patient had an elevated antineutrophil antibody titer of 1:640 with a speckled pattern, but tests were negative for antibodies to double-stranded DNA, ribonucleic protein, and Smith antigen; she had no clinical evidence of vasculitis.

Of the 6 patients who had a head computed tomographic scan, 1 patient demonstrated chiasmal enhancement and 5 patients had normal results. Fifteen patients underwent cranial magnetic resonance imaging (MRI) with and without contrast, one of whom had already undergone a computed tomographic scan. Twelve of these 15 patients (80%) demonstrated chiasmal enlargement and/or enhancement. The chiasm was well visualized and appeared normal in the remaining 3 patients. One or more deep white matter lesions were found in 6 of 15 patients who underwent MRI scans.

Six patients also underwent lumbar puncture, with normal cerebrospinal fluid study results in 4 of them. The other 2 patients had elevated IgG concentrations, 1 of whom also had oligoclonal bands. The results of radiographic testing and cerebrospinal fluid analysis are summarized in Table 2.

VISUAL OUTCOMES

The length of follow-up ranged from 2 weeks to 22 years, with an average follow-up time of 69 months (5.7 years). No follow-up was available in 1 patient. Seventeen patients had received interventional treatment: oral prednisone in 9 patients and intravenous methylprednisolone in 8. Four eyes had preexisting subnormal acuity and there was insufficient information for 2 patients (4 eyes); thus, the determination of final acuity was based on 32 affected eyes. Final acuity ranged from 20/15 to 20/50 (median, 20/20) and was 20/20 or better in 20 of 32 eyes. Results of formal visual field testing were available in these 32 eyes, of which 24 showed improvement and 8 had normal visual fields. The visual and neurologic outcomes of all patients are given in Table 3.

NEUROLOGIC OUTCOMES

None of the patients had a neurologic deficit other than visual loss at the time of their initial neuroophthalmologic consultation. Fifteen patients were followed up for at least 1 year; 6 of them later developed clinically definite MS when they experienced another acute neurologic (nonvisual) deficit with radiologic and laboratory findings consistent with demyelination. The interval between diagnosis of idiopathic chiasmal neuritis and MS were 3 months (2 patients), 5 months (1 patient), 1 year (1 patient), 3 years (1 patient), and indeterminate (1 patient). Of the 6 patients who had white matter lesions on initial MRI, 2 developed MS, 3 did not, and no information was available for 1 patient. Of the 2 patients who had abnormal cerebrospinal fluid at their initial visits, 1 developed MS.

COMMENT

The demographic profile of our patients with idiopathic chiasmal neuritis resembles that of idiopathic optic neuritis. This is not unexpected, given that these 2 entities represent different manifestations of the same pathologic process, ie, demyelinating disease. Intuitively, the clinical profile of the acute attack in the 2 disorders would be expected to be similar as well. The clinical features and long-term outcomes of idiopathic optic neuritis are well defined based on data from the large, prospective multi-center Optic Neuritis Treatment Trial. However, comparable information regarding chiasmal neuritis is scanty.

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In the early 1900s, a few articles described acute chiasmal visual field defects in patients with MS, and pathologic studies confirmed that demyelinating foci could involve the chiasm. The first article that demonstrated a clinicopathologic correlation was written by Bell et al in 1975; they documented the findings of an exploratory craniotomy. Sacks and Melen described clinical features in 3 patients with chiasmal neuritis due to MS. With the advent of computed tomographic imaging and then MRI, the diagnosis could be established with more confidence. The clinical features of chiasmal neuritis were further expanded by Spector et al, who reported on a series of 6 patients with chiasmal visual loss due to MS. These authors noted that the condition has a predilection for young to middle-aged women, improves spontaneously, and carries a generally favorable visual outcome. A second series of 6 patients with chiasmal neuritis was described by Newman et al in 1991, further characterizing this syndrome, including its radiographic features. Their patients, like those in previous reports, were predominantly female and generally enjoyed good visual recovery. These authors pointed out that pain was an accompanying symptom in 3 patients (50%) and that visual loss was protracted during 1 to several months in 4 patients. Four of the 6 patients had an abnormal chiasmal appearance on MRI, demonstrating high-signal ab-
normalities on T2-weighted images with enlargement and/or enhancement. One patient developed an acute transverse myelopathy 3 months after onset of visual loss.

In our series of 20 patients, 14 were female (70%). The mean age was 37 years. The oldest patient in our series (age 73 years) is also the oldest reported patient in the literature. The chief symptom in this series was visual loss that was monocular in approximately half of cases. Unlike idiopathic optic neuritis in which more than 90% of patients report pain, only 20% of our patients with chiasmal neuritis reported accompanying eye pain. Interestingly, as in optic neuritis, visual phenomena were common in our series, described by 25% of patients. These photopsias were transient and typically described as sparkles, glare, or too much light. Visual loss worsened across a period of days to weeks, then stabilized and improved. Progressive visual loss beyond 1 month occurred in only 1 patient and was not associated with a worse visual outcome. Visual loss was moderately severe with visual acuity of 20/200 or worse in one-third of eyes.

Magnetic resonance imaging is a very helpful diagnostic tool in this setting, as in idiopathic optic neuritis. In addition to ruling out extrinsic lesions of the chiasm, in most patients (80%), MRI provided support for the diagnosis, showing enlargement and/or enhancement of the chiasm. Cerebrospinal fluid analysis was performed in only 6 patients and its results were largely normal.

Treatment for chiasmal neuritis has no formal guidelines and must be individually determined. Most patients were offered treatment because their bilateral visual loss was functionally disabling and it was hoped that corticosteroids would facilitate recovery. Almost half of these patients were treated with oral prednisone, because information from the Optic Neuritis Treatment Trial suggesting that intravenous methylprednisolone was preferable was not yet available. Regardless of treatment, all affected eyes improved substantially, 97% reaching a visual acuity of 20/40 or better, 63% recovering to 20/20. Visual fields also improved or normalized in all patients. Thus, the course of visual loss and extent of recovery in chiasmal neuritis resembles that of optic neuritis.

This is the first series to examine the neurologic prognosis of chiasmal neuritis across an extended period. Only 1 patient had a prior diagnosis of demyelinating optic neuritis (patient 11). Two others (patients 16 and 20) had experienced transient visual loss that may have been due to optic neuritis. Two patients had 1 preceding symptom that may have been demyelinating (illusory motion and truncal numbness in patients 7 and 13, respectively). In the remaining 15 patients, chiasmal neuritis was the first recognized demyelinating event. Six patients (40%) in our cohort group developed clinically definite MS at 3 months to 3 years from onset of chiasmal neuritis. In 4 of these 6 patients, the second demyelinating event occurred within 1 year. In previous studies regarding optic neuritis, the presence of white matter lesions on MRI and cerebrospinal fluid abnormalities was strongly predictive of the development of MS. In our study, the number of patients studied is insufficient to draw conclusions regarding the predictive power of the MRI findings. Of the 4 patients who had white matter lesions on MRI at the time of their visual loss, 2 subsequently developed more extensive demyelinating disease. One of the 2 patients who had cerebrospinal fluid abnormalities developed MS.

In conclusion, our series supports the assumption that idiopathic chiasmal neuritis is clinically similar to idiopathic optic neuritis in terms of patient demographics, clinical course, visual outcomes, and risk for the subsequent development of MS. The only notable difference was absence of eye pain accompanying visual loss in pa-
patients with idiopathic chiasmal neuritis. The clinical similarity between chiasmal neuritis and optic neuritis suggests a shared pathophysiology, and we would recommend that management of chiasmal neuritis follow the guidelines put forth by the Optic Neuritis Treatment Trial for optic neuritis. Whether ancillary testing such as MRI or lumbar puncture has the same predictive value for the development of MS cannot be determined at this time.

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REFERENCES


In 1795, Dr Isaac Thompson concocted an eye water of zinc sulfate, saffron, camphor, and rose water. It was sold as late as 1939. This is 1 of a series of 32 medical trade cards advertising the product from 1875 through 1895.

Ophthalmological Ephemera

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