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Evaluating the incidence of arteritic ischemic optic neuropathy and other causes of vision loss from giant cell arteritis

John J. Chen, MD, PhD[1], **Jacqueline A. Leavitt, MD**[1], **Chengbo Fang, MD**[1,2], **Cynthia S. Crowson, MS**[3,4], **Eric L. Matteson, MD, MPH**[3,4], and **Kenneth J. Warrington, MD**[3]

[1]Department of Ophthalmology, Mayo Clinic

[2]Department of Ophthalmology, The first affiliated hospital of Anhui Medical University

[3]Division of Rheumatology, Department of Medicine, Mayo Clinic

[4]Department of Health Sciences Research, Mayo Clinic

Abstract

Objective—To determine the incidence of permanent visual loss from giant cell arteritis (GCA).

Design—Retrospective, population-based cohort

Participants—All residents of Olmsted Country, Minnesota, USA, diagnosed with GCA between January 1, 1950 and December 31, 2009

Methods—All cases of GCA were identified using the Rochester Epidemiology Project, which is a record-linkage system of medical records for all patient-physician encounters among Olmsted County, Minnesota residents. The medical records were reviewed to identify and determine the cause of permanent vision loss among patients with GCA. Systemic symptoms of GCA and visual outcomes were also determined.

Main Outcome Measures—Incidence and outcomes of permanent vision loss from GCA.

Results—Among the 245 new cases of GCA over the 60-year period, 20 (8.2%) patients suffered permanent vision loss due to GCA. The frequency of arteritic ischemic optic neuropathy (A ION) was 6.9% (95%CI: 4.0%–11.1%) accounting for 85% of cases of permanent vision loss. The frequency of central retinal artery occlusion (CRAO) was 1.6% (95%CI: 0.4%–4.2%) and cilioretinal artery occlusion was 0.4% (95%CI: 0.01%–2.3%). The population-based age- and sex-adjusted annual incidence of A-ION from GCA among persons age ≥ 50 years was 1.3 (95%CI: 0.7–2.0) per 100,000 population. 20% of patients with permanent vision loss from GCA had vision loss without constitutional symptoms of GCA. Overall, there was no significant difference between presenting and final visual acuities.

Corresponding Author: John J. Chen, MD, PhD, Department of Ophthalmology, Mayo Clinic, 200 First Street SW, Rochester, MN 55905, chen.john@mayo.edu, Tel: 507-284-4946, Fax: 507-284-4612.

No conflicting relationship exists for any author.

Conclusion—This population-based data provides the most accurate incidence of permanent vision loss from GCA. This study confirms that visual outcomes from GCA-related vision loss are poor and that 20% of patients with permanent visual loss from GCA can present without systemic symptoms of GCA.

Introduction

Giant cell arteritis (GCA) is a vasculitis of medium and large-sized vessels and is the most common vasculitis in adults in the Western world.[1] Patients often present with systemic symptoms of headaches, scalp tenderness, jaw claudication, malaise, anorexia, fever, and weight loss.[2–5] GCA is a medical emergency because of its potential to cause rapidly progressive irreversible blindness. The most common cause of permanent vision loss from GCA is arteritic ischemic optic neuropathy (A-ION) from inflammatory posterior ciliary artery occlusion,[6,7] which accounts for 81.2% of vision loss according to one large series.[6] Vision loss from A-ION is typically severe with the majority of patients presenting with a visual acuity of less than 20/200 and up to 21% of patients with no light perception.[6,8] Patients can have ocular involvement without systemic manifestations of GCA, which some have termed “occult GCA”, in 5–38% of cases.[9–12]

Other causes of vision loss from GCA include central retinal artery occlusion, posterior ischemic optic neuropathy, and rarely ophthalmic artery occlusion.[6,7] The frequency of ophthalmic manifestations from GCA is poorly defined and ranges from 14 – 70%,[6,7,13–18] which is likely a reflection of selection and/or referral bias in prior studies.

Because GCA is a potentially blinding condition, determining the true incidence of permanent vision loss is important to help guide the screening and treatment of this disease. Prior studies were mostly performed at large referral centers, which can suffer from a referral bias towards the most severe patients. The true population-based incidence of the ocular causes of vision loss from GCA was calculated using the resources of the Rochester Epidemiology Project along with a description of previous reports of all visual manifestations of GCA.[19] The goal of this study is to evaluate the population-based incidence of A-ION and other causes of permanent vision loss from GCA using the Rochester Epidemiology Project. This study will examine in more detail the patients presenting with permanent vision loss from GCA. In addition, this study will evaluate the incidence of permanent vision loss in patients with no systemic symptoms of GCA.

Methods

A retrospective review was conducted of all the medical records of patients with newly diagnosed GCA identified through the resources of the Rochester Epidemiology Project (REP), which is a record-linkage system of medical records for all patient healthcare provider encounters among residents of Olmsted County, Minnesota.[20,21] These records capture all medical care at Mayo Clinic and Olmsted Medical Center and their affiliated hospitals, as well as the few private practitioners. This study was approved by the Mayo Clinic and Olmsted Medical Center institutional review boards. All patient records were reviewed to identify newly diagnosed GCA between January 1, 1950 and December 31,

2009. The diagnosis of GCA was based on the 1990 American College of Rheumatology criteria.[3]

The medical records were reviewed to identify patients with permanent vision loss from GCA. The cause of vision loss, visual acuity at presentation and last follow-up examination, and fundus findings were documented. The presenting erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and presence/absence of systemic symptoms of GCA were documented, including headaches, scalp tenderness, jaw claudication, fever, weight loss, and polymyalgia rheumatica (PMR).

Statistical Analysis

Snellen visual acuities were converted to logarithm of minimum angle of resolution (logMAR) values for statistical analysis, and the following LogMAR values were used for non-numeric visual acuities: no light perception (NLP) = 3.0, light perception (LP) =2.3, hand motion (HM) = 2.0, and count fingers (CF) = 1.7, which is a previously used scale to quantify visual loss from GCA.[22]

Descriptive statistics (means, percentage, etc.) were used to summarize the data. Comparisons of characteristics of GCA patients with and without A-ION were performed using Chi-square and rank sum tests. The frequency of A-ION and CRAO were calculated as the number of patients with A-ION or CRAO over the total number of patients with GCA, and 95% Poisson confidence intervals (CI) were calculated.

Age- and sex-specific incidence rates were calculated using the number of incident cases of A-ION as the numerator and population estimates for Olmsted County residents age ≥ 50 years based on decennial census counts as the denominator; linear interpolation was used to estimate population size for intercensal years. Overall rates were age- and sex-adjusted to the 2010 United States white population. Ninety-five percent confidence intervals (95% CIs) were computed for incidence rates assuming the incident cases follow a Poisson distribution. Analyses were performed using SAS version 9.4 (SAS Institute, Cary, NC, USA).

Results

A total of 245 patients were newly diagnosed with GCA from 1950 to 2009. Among those patients, a total of 20 patients (8.2%) (26 eyes) suffered an ischemic event leading to permanent visual acuity or visual field loss. Eighty percent of the patients with permanent vision loss from GCA were women compared to 79% for the remainder of the GCA cohort (p=0.93) (Table 1). Among patients with permanent vision loss, the average (± standard deviation) age was 82.1 ± 6.5 (range 67–92), compared to 75.7 ± 8.2 (range 56–97) in the remainder of the GCA cohort (p<0.001) (Table 1).

Among patients with permanent vision loss, 17 patients (22 eyes) had A-ION (12 unilateral, 5 bilateral), one of which had a concordant cilioretinal artery occlusion in the ipsilateral eye (Table 2 and 3). The frequency of A-ION among the GCA cohort was 6.9% (95% CI: 4.0% – 11.1%) accounting for 85% of cases of permanent vision loss, while cilioretinal artery occlusion (CLAO) was seen in 0.4% (95% CI: 0.01% – 2.3%) of the GCA cohort. Four

patients (4 eyes) had unilateral central retinal artery occlusion (CRAO) (Table 2 and 3), one of which had A-ION in the contralateral eye. The frequency of CRAO among the GCA cohort was 1.6% (95% CI: 0.4% – 4.2%).

The overall population-based age- and sex-adjusted annual incidence of A-ION from GCA among persons age ≥ 50 years was 1.3 (95 % CI: 0.7 – 2.0) per 100,000 population. The age-adjusted incidence rate of A-ION from GCA was higher among women (1.6; 95% CI: 0.7 – 2.4 per 100,000 population) than men (1.0; 95% CI: 0.0 – 1.9 per 100,000 population).

Among the 17 patients with A-ION, there were 14 patients (18 eyes) presenting with disc edema from acute A-ION. Among patients with acute A-ION, 10 of 14 patients (14 of 18 eyes) had pallid disc edema at presentation (Table 3). Three patients (4 eyes) had chronic vision loss and presented with optic nerve pallor without optic disc edema at initial examination. These patients had prior arteritic ischemic optic neuropathy, most likely arteritic anterior ischemic optic neuropathy, but could also have had prior posterior ischemic optic neuropathy because optic disc edema was never documented.

The presenting visual acuity for patients with A-ION from GCA ranged from 20/20 to NLP with 15 of the 23 eyes worse than 20/200 (Table 2 and 3). Final visual acuities ranged from 20/20 to NLP with an average follow-up of 4.25 years. While three eyes improved by > 2 lines, overall there was not a significant improvement in visual acuity ($p = 0.78$, Table 2 and 3). Eyes presenting with NLP vision did not show any recovery despite steroid treatment. Eyes presenting with hand motion or LP mostly did not show significant improvement, but a few improved to CF. Three eyes from two patients presenting with LP or CF vision progressed to NLP despite immediate steroid treatment.

For the 4 patients with CRAO, the presenting visual acuity ranged from 20/250 to NLP (Table 2 and 3). While no optic disc edema was present, it is possible that patients with CRAO could have had concordant ophthalmic artery occlusion, especially the patient that was NLP. One patient improved from 20/250 to 20/100 and another patient improved from LP to CF, otherwise no recovery was noted.

There was a trend toward better visual outcomes among patients presenting in the more recent half of the cohort (1980–2009) compared to patients presenting in the first half (1950–1979), with an average visual acuity of 20/300 vs HM, respectively ($p=0.062$).

Sixteen of 20 patients with permanent vision loss had at least one systemic symptom of GCA, which included headaches, scalp tenderness, jaw claudication, fever, weight loss, and/or PMR (Table 1 and 4). Therefore 4 (20%) patients with biopsy proven GCA and permanent vision loss had no systemic symptoms of GCA. These patients all had an elevated ESR with an average of 74.3 ± 10.9. In comparison, 9 (4%) of GCA patients without vision loss had no GCA systemic symptoms ($p=0.002$).

The average ESR was 72.1 ± 21.2 among patients with permanent vision loss compared to 80.6 ± 57.5 for remainder of the GCA cohort ($p=0.51$) (Table 1 and 3). With a cut off of age/2 for men and (age+10)/2 for women,[23] ESR was considered normal in 2 of 20 patients. If the lab reference defined range of abnormal was used (≥29 for women and ≥22 for men),

all 20 patients had an elevated ESR. CRP was only obtained in 4 patients and was elevated in all of them with an average of 70.5mg/L ± 22.6. The CRP was not obtained in the two women that had an ESR less than (age+10)/2.

Discussion

Among this large geographically defined cohort of 245 patients with GCA, 8.2% suffered permanent vision loss from an ischemic insult from GCA. Others have previously reported a higher incidence of permanent vison loss from GCA, ranging from 10.2% to 48.8%.[6,13,16–18,24–27] A-ION accounted for the vision loss in 85% of this GCA cohort, which is similar to prior studies.[6,16,22,25]

The population-based annual incidence of A-ION from GCA was 1.3 per 100,000. Johnson and Arnold published the only prior incidence study on A-ION and found an annual incidence of 0.36 per 100,000 in the state of Missouri and Los Angeles County, California.[28] However, their study was dependent on a survey questionnaire to practicing ophthalmologists and therefore inherently suffers from recall and selection bias. Prior studies have indicated an annual incidence of GCA of 19.8 per 100,000.[29,30] Assuming 8.2–48.8% of these patients suffer permanent vision loss, 85% from A-ION as discussed above, the annual incidence of A-ION from GCA of 1.3 per 100,000 in the current study is likely closer to the actual incidence of this complication. The higher incidence in the current study may also reflect the population of Olmsted County, which is 90.3% Caucasian according to the 2000 US census data. GCA predominantly affects individuals of Northern European descent and therefore this study is best generalizable to other Caucasians with GCA in the United States.[31]

The current study confirms that vision loss from A-ION and CRAO from GCA is quite severe with 19 of 26 (73%) eyes presenting with visual acuity worse than 20/200. 15% of eyes had an improvement of 2 lines or better in final visual acuity. Based on prior studies, this likely reflects improved eccentric fixation rather than true improvement in the majority of cases because others have found similar poor rates of recovery, where 10–15% patients treated with immediate steroids had an improvement 2 lines or better in visual acuity, but only 4–5% showed a corresponding improvement in visual field.[22,32] In our series, patients presenting with poor vision had little chance of recovery despite immediate steroid treatment. No patients presenting with NLP had any improvement and patients presenting with HM or LP vision at most improved to CF. The main goal of steroids is to prevent vision loss in the fellow eye, which is thought to occur within days in up to 50% of cases of GCA if left untreated.[6]

There was a trend toward better visual outcomes in more recent years (1980–2009) compared to patients seen in the more distant past (1950–1979). This was driven primarily by 2 patients that ended up NLP bilaterally that presented before 1980. Interestingly, both patients sought medical attention with mild vision loss and the diagnosis of GCA was missed until they had complete loss of vision in both eyes. There have been no cases of bilateral blindness since 1980, which is likely because medical professionals are now more aware of GCA and typically started patients on prednisone if there are any concerns of GCA.

Eighty percent of patients with permanent vision loss from GCA were women, which is in concordance with the overall female predominance of GCA in this as well as other cohorts.[22,29,30,33,34] Interesting, Johnson and Arnold found no sex predilection for A-ION in their incidence study.[28]

20% of patients with permanent vision loss from GCA had no premonitory systemic symptoms of the disease, which is similar to what was reported by Hayreh and colleagues.[10] This was significantly higher than the 4% occult presentation in the GCA cohort without permanent vision loss. With prior studies indicating an occult presentation in 5% to 38% of cases of GCA, systemic symptoms of GCA cannot be relied upon to rule out an ischemic complication from GCA,[9–12] especially because patients with permanent vision loss are more likely to present without systemic symptoms of GCA.

The primary limitation of this study is its retrospective nature. Not all patients had formal visual fields performed and therefore visual fields were not included in the study. As with any retrospective study, the lack of a symptom documented in the history does not necessarily mean the patient did not have them. For this reason, some visual symptoms, such as amaurosis preceding the vision loss, were not included in this study.

Overall, this study provides the most accurate population-based incidence of A-ION from GCA. At an annual incidence of 1.3 per 100,000, it is higher than previously reported at 0.36 per 100,000. Despite the higher incidence of A-ION, the rate of permanent vision loss from A-ION and other GCA-related causes of vision loss among GCA patients was 8.2%, which is lower than tertiary referral-based series on visual manifestations of GCA. This is a major strength of a population based study, which avoids referral bias to tertiary referral-based centers that can increase the rate of visual manifestations of GCA and recall bias from a survey-based study that can artificially reduce, or possibly increase the estimate of A-ION.

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Abbreviations

GCA	giant cell arteritis
A-ION	arteritic ischemic optic neuropathy
CRAO	central retinal artery occlusion
ESR	erythrocyte sedimentation rate
CPR	C-reactive protein
PMR	polymyalgia rheumatica
REP	Rochester Epidemiology Project

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Table 1

Demographic characteristics of patients with giant cell arteritis

Characteristics	No vision loss (N=225)	Vision loss (N=20)	*P* value
Age, years, mean (±SD)	75.7 (±8.2)	82.1 (±6.5)	<0.001
Female sex	178 (79%)	16 (80%)	0.93
ESR, mm/hr, mean (±SD)	80.6 (±57.5)	72.1 (±21.2)	0.51
No systemic sx of GCA	9 (4%)	4 (20%)	0.002
Fever > 100° F	37 (17%)	3 (15%)	0.85
Weight loss	54 (24%)	1 (5%)	0.048
Headache	168 (75%)	9 (45%)	0.003
Jaw claudication	98 (44%)	11 (55%)	0.34
Scalp tenderness	96 (45%)	5 (25%)	0.08
Headache, Jaw claudication or scalp tenderness	199 (98%)	14 (70%)	0.012
Polymyalgia rheumatica	61 (27%)	5 (25%)	0.84

Values in table are n (%) unless otherwise specified.

Table 2

Causes of vision loss among giant cell arteritis patients.

Cause of vision loss	# of patients*	# of eyes	Avg Initial Visual Acuity (range)	Avg Final Visual Acuity (range)	P value
A-ION	17	22	20/800 (20/20-NLP)	20/650 (20/20-NLP)	0.78
CRAO	4	4	HM (20/250-NLP)	CF (20/100-NLP)	0.70
CLAO	1	1	CF	CF	

A-ION: arteritic ischemic optic neuropathy, CRAO: central retinal artery occlusion, CLAO: cilioretinal artery occlusion, NLP: no light perception, HM: hand motion, CF: count fingers

Table 3

Patients with permanent vision loss from giant cell arteritis.

Patient	Age	Gender	Ocular sx	Eye(s) affected	Fundus findings	Initial VA	Final VA	ESR	CRP	GCA sx
1	83	F	A-ION	OU	Pallid disc edema	NLP OD, CF OS	NLP OU	102		Yes
2	77	F	A-ION OS + CRAO OD	OU	Pallid disc edema OS. Retinal whitening OD	20/250 OD, NLP OS	20/100 OD, NLP OS	107		Yes
3	67	M	A-ION	OS	Pallid disc edema	HM	CF	80		Yes
4	74	F	A-ION	OD	Pallid disc edema	LP	LP	103		Yes
5	83	F	CRAO	OS	Retinal edema	LP	CF	76		Yes
6	82	M	A-ION + CLAO	OS	Disc edema, retinal whitening temporal to the disc	CF	CF	79		No
7	92	M	A-ION	OU	Pallid disc edema	Bare LP OU	NLP OU	67		Yes
8	73	F	A-ION	OD	Pallid disc edema	20/20	20/20	108		Yes
9	84	F	A-ION	OU	Pallid disc edema	CF OU	CF OD, 20/70 OS	81		No
10	83	F	CRAO	OD	Attenuated arteries	NLP	NLP	79		No
11	81	F	A-ION	OS	Mild disc edema with hemorrhages	CF	CF	52		Yes
12	86	F	A-ION	OU	Pallor	20/40 OU	20/40 OU	38		Yes
13	83	F	A-ION	OD	Pallor	LP	CF	57		Yes
14	81	F	A-ION	OS	Pallid disc edema	LP	HM	52		Yes
15	92	F	A-ION	OD	Superior disc edema	20/50	20/50	44		Yes
16	82	M	A-ION	OS	Disc edema	20/400	20/70	60		Yes
17	80	F	A-ION	OD	Pallid disc edema	20/150	20/70	69	90	Yes
18	78	F	A-ION	OU	Pallid disc edema	HM OD, 20/25 OS	HM OD, 20/20 OS	58	48.1	No
19	87	F	A-ION	OS	2+ pallor	20/200	20/200	50	90	Yes
20	92	F	CRAO	OD	Retinal edema and cotton wool spot	HM OD	HM OD	80	54	Yes

A-ION: arteritic ischemic optic neuropathy, CRAO: central retinal artery occlusion, CLAO: cilioretinal artery occlusion, HM: hand motion, CF: count fingers, LP: light perception, NLP: no light perception

Using population-based data, we provide the most accurate incidence of arteritic ischemic optic neuropathy from giant cell arteritis, which was 1.3 per 100,000 in patients over the age of 50.