TEMERTY FACULTY OF MEDICINE
UNIVERSITY OF TORONTO

Ischemic Ocular Manifestations of Giant Cell arteritis: A Canadian Case Series

Mariam Issa MD(C),[1] Laura Donaldson MD PhD,[2] Edward Margolin MD[2,3]

[1]Faculty of Medicine, University of Toronto, Toronto, Ontario, Canada, [2]Department of Ophthalmology and Vision Sciences, Faculty of Medicine, University of Toronto, Toronto, Ontario, Canada
[3]Department of Medicine, Division of Neurology, Faculty of Medicine, University of Toronto, Toronto, Ontario, Canada

Introduction

Giant cell arteritis (GCA) affects the ocular circulation in more than half of cases and can cause irreversible bilateral blindness if not rapidly recognized and treated.

Diagnosis can be challenging due to the variable clinical presentation and lack of systemic symptoms in approximately 20% of cases, which are termed occult GCA.

We compared clinical and laboratory findings of patients with occult vs. systemic temporal-artery biopsy (TAB) positive GCA with ocular manifestations.

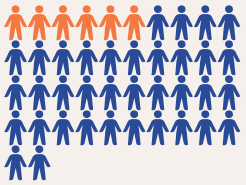
Objective

- Determine the frequency of occult GCA
- Compare the levels of inflammatory markers in patients with occult versus systemic disease
- Determine the vascular circulations involved
- Study the incidence of other neuro-ophthalmologic manifestations of GCA, such as cranial nerve palsies

Methodology

Retrospective chart review of patients with TAB-positive GCA with ocular manifestations seen at a tertiary neuro-ophthalmology practice between 2015 and 2020 and focused on comparing the characteristics of patients with occult vs. systemic GCA.

Demographics



Occult GCA was seen in 6/42 (14.3%) of patients.

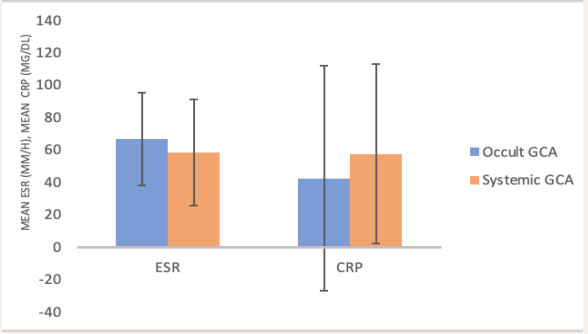
No significant difference in sex distribution between groups.

Occult GCA affected older patients (occult GCA; 84.6 ± 5.3 years, systemic GCA; 75.9 ± 8.2,

Occult GCA patients had worse presenting visual acuity (occult GCA: 2.66 ± 0.05 LogMAR, systemic GCA: 1.09 ± 1.18 LogMAR)

Results

Figure 1: Erythrocyte Sedimentation Rate (ESR) and C-Reactive Protein (CRP) Levels in GCA



- 92.9% of patients had elevation of at least one inflammatory marker and 42.5% of patients had elevation in both. One patient with occult GCA and one patient with systemic GCA had normal inflammatory markers.

Key message

Absence of systemic symptoms and normal markers are not sufficient to rule out GCA.

- There was no significant difference in ESR/CRP levels or ocular signs/symptoms between occult vs systemic groups.
- AION was the most common ischemic ocular manifestation of both occult and systemic GCA. One patient with occult GCA had isolated cranial nerve (CN) six palsy.

Key message

GCA should be ruled out in all patients who are older than 50 years of age who present with isolated sixth nerve palsy

- Involvement of multiple ocular circulations was seen in six patients, all in the systemic group.
- AION was diagnosed in 20% (2/10) of patients with symptoms of amaurosis fugax.

Table 1: Clinical Signs/Symptoms and Ischemic lesions in Patients with Ocular Manifestation of GCA

Symptom/Lesion		Occult Ocular GCA N = 6	Ocular and Systemic GCA N = 36	P value
Ocular Signs/Symptoms	Blurred Vision	4 (66.7)	20 (55.6)	0.69
	Diplopia	1 (16.7)	4 (11.1)	0.57
	Ptosis	0 (0)	1 (2.8)	1.0
	Halos	0 (0)	1 (2.8)	1.0
	Visual field defect	0 (0)	2 (5.4)	1.0
	Optic disc edema	3 (50)	13 (36.1)	0.14
	Pallid edema	2 (33.3)	7 (19.4)	0.59
	Optic disc pallor only	2 (33.3)	5 (13.9)	0.26
	RAPD	5 (83.3)	22 (61.1)	0.40
Cranial Symptoms	Jaw claudication	-	18 (50.0)	-
	Scalp tenderness	-	8 (22.2)	-
	Headache	-	20 (55.6)	-
	Dizziness/light-headedness	-	1 (2.8)	-
	Neck pain	-	1 (2.8)	-
Systemic manifestations	Fever	-	4 (11.1)	-
	Weight loss	-	6 (16.7)	-
	Fatigue	-	8 (22.2)	-
	Myalgia	-	2 (5.6)	-
	PMR	-	7 (19.4)	-
	Loss of appetite	-	1 (2.8)	-
Ocular ischemic symptoms	AION	5 (83.3)	18 (50.0)*	0.2
	CRAO	0 (0)	3 (8.3)*	1.0
	PION	0 (0)	1 (2.8)	1.0
	Amaurosis fugax	0	10 (27.8)	0.31
CN Palsies	CWS	0 (0)	6 (16.7)	0.57
	CN4P	0 (0)	4 (11.1)	1.0
	CN6P	1 (16.7)	1 (2.8)	0.27
	Normal neuro-ophthalmic exam	0 (0)	3(8.3)	1.0

RAPD: Relative Afferent Pupillary Defect, AION: Anterior ischemic optic neuropathy, CRAO: Central retinal artery occlusion, PION: Posterior ischemic optic neuropathy, CWS: Cotton wool spots m CN4P: Cranial nerve 4 palsy, CN6P: Cranial nerve 6 palsy . *One patient in the systemic group had bilateral ocular symptoms

Conclusion

Patients with occult GCA are typically older but ocular ischemic manifestations are similar in occult and systemic GCA. Levels of inflammatory markers are similar in both cohorts.

GCA should always be considered in older patients who present with signs of retinal and/or optic nerve head ischemia and new onset of CN palsies.