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Case Report

An Atypical Presentation of Giant Cell Arteritis with Bilateral Choroidal Ischaemia

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ABSTRACT

Background: Giant cell arteritis is an immune-mediated, medium to large vessel vasculopathy affecting individuals over 50 years of age. It can cause sudden, severe and irreversible loss of vision, most commonly from an arteritic posterior ciliary artery occlusion causing anterior ischaemic optic neuropathy. The optic nerve appearance would typically be swollen and chalky white. Visual reduction secondary to choroidal ischaemia is a much less frequent presentation, the signs of which can be more subtle in appearance, making its early recognition potentially more challenging.

Case Presentation: A 51-year-old male presented to eye casualty complaining of a one-week history of neck pain, intermittent headaches and jaw claudication, associated with reduced vision in his right eye. Presenting visual acuity was hand movements and 6/5 in the right and left eyes respectively. On examination, he was noted to have a right relative afferent pupillary defect, a pale macular area in the right eye with a possible cherry red spot. There was no optic disc swelling. ESR was 34 and CRP was 46 and he was wrongly diagnosed with a non-arteritic central retinal artery occlusion. He subsequently re-presented 5 weeks later with vision loss in his left eye. Best corrected visual acuity was now 6/60 and 6/12 in the right and left eyes respectively. Dilated fundoscopy showed multiple yellow-white lesions in the posterior pole of the left eye and a retinal cotton wool spot. The right optic nerve was pale, and left was normal. A fundus fluorescein angiogram showed delayed choroidal filling and the temporal artery biopsy was suggestive of GCA. He was started on 110mgs of oral prednisolone. After 4 weeks of steroids his BCVA was 6/36 and 6/6 in the right and left eyes respectively. His neck pain, headaches and jaw claudication had all resolved and his ESR and CRP had returned to normal levels.

Conclusion: Our case highlights the need for increased awareness of this uncommon presentation of this potentially blinding disease, to allow prompt and appropriate treatment. Our case is unusual in that despite a delayed diagnosis of 5 weeks, visual acuity initially improved with treatment.

Background

Giant cell arteritis is an immune-mediated, medium to large vessel vasculopathy affecting individuals over 50 years of age. It can cause sudden, severe and irreversible loss of vision, most commonly from an arteritic posterior ciliary artery occlusion causing anterior ischaemic optic neuropathy. The optic nerve appearance would typically be swollen and chalky white. Visual reduction secondary to choroidal ischaemia is a much less frequent presentation, the signs of which can be more subtle in appearance, making its early recognition potentially more challenging. Our case highlights the need for increased awareness of this uncommon presentation of this potentially blinding disease, to

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allow prompt and appropriate treatment. Our case is unusual in that despite a delayed diagnosis of 5 weeks, visual acuity initially improved with treatment.

Case Presentation

A 51-year-old obese (BMI 50) male ex-smoker, presented to eye casualty complaining of a one-week history of neck pain, intermittent headaches and jaw claudication, associated with a 3-day history of reduced vision in his right eye. Presenting best corrected visual acuities (BCVA) were hand movements and 6/5 in the right and left eyes respectively. On examination by the attending doctor he was noted to have a right relative afferent pupillary defect (RAPD), a pale macular area in the right eye with a possible cherry red spot. The absence of optic disc swelling was noted. An initial diagnosis of a right central retinal artery occlusion (CRAO) was made. Urgent ESR and CRP were performed, measuring 34 and 46 respectively. Giant cell arteritis (GCA) was wrongly dismissed and a diagnosis of right non-arteritic CRAO was made. The patient was referred to a TIA clinic for investigation. He had a CT head and carotid Doppler imaging which were normal and was commenced on aspirin and a statin. He was subsequently discharged. Five weeks later he re-presented to eye casualty complaining of a one-day history of reduced vision in his left eye. Best corrected visual acuity was now 6/60 and 6/12 in the right and left eyes respectively. Dilated fundoscopy (illustrated in Figures 1 & 2 showing the right and left eyes respectively) showed multiple yellow-white lesions in the posterior pole of the left eye that were felt to be located at or below the retinal pigment epithelium (RPE) associated with a retinal cotton wool spot. No features of anterior ischaemic optic neuropathy (AION) were seen in either eye. The right optic nerve was documented to be pale but not swollen. The left optic disc was documented to be normal. The right macular area appeared greyish-white. Interestingly the patient's father lost vision in both eyes at the age of 72 from GCA.

An urgent fundus fluorescein angiogram (FFA) and temporal artery biopsy were arranged. A diagnosis of GCA with bilateral choroidal ischaemia was made clinically. The patient weighed 180Kg. He was started on 110mgs of oral prednisolone and 40mg of omeprazole. Eight days following the initiation of oral prednisolone the patients VA had improved in the right eye to 6/24. After 4 weeks of steroids his BCVA was 6/36 and 6/6 in the right and left eyes respectively. His neck pain, headaches and jaw claudication had all resolved and his ESR and CRP had returned to normal levels (8 and 6 respectively). Temporal artery biopsy demonstrated focal lymphocytic infiltrate together with multi-nucleated giant cells and epithelioid histiocytes which are features suggestive of giant cell arteritis. FFA showed delayed choroidal filling in both eyes with evidence of choroidal ischaemia. Background choroidal filling was delayed until 24.9s with localised areas of ischaemia not filling until after 41.1s. There was normal filling of the retinal vessels and no features in-keeping with a previous diagnosis of right CRAO. There was also marked delay in the perfusion of the right optic nerve. After one year of treatment the BCVA in the right eye had decreased to 6/60 secondary to atrophic changes of the macular RPE and optic atrophy, but the LE remained 6/6.

Discussion

GCA is an immune-mediated vasculopathy that typically affects medium to large arteries [1]. The vascular structures most commonly affected include the branches of the external carotid artery (such as the temporal and occipital arteries), the thoracic aorta and axillary arteries, along with the distal subclavian, vertebral and ophthalmic arteries. The main risk factor for the disease is age. It rarely affects people under the age of 50 years and has a peak incidence of 5.9 per 10 000 persons per year in the 70-79-year age range [2]. It affects predominantly woman and has a sex ratio of 2-3:1 [3]. GCA is a medical emergency, requiring prompt treatment as the inflammation can lead to luminal occlusion and ischaemic complications that if left untreated can result in visual loss in 10 to 15% of patients [4].

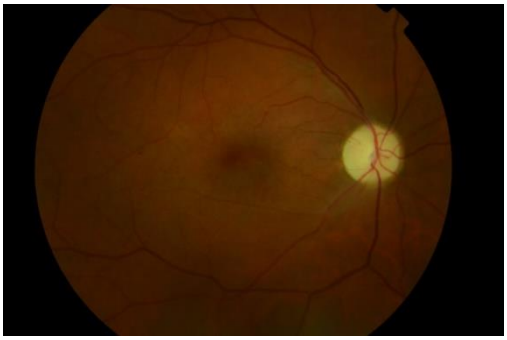

Figure 1: Colour fundus photograph of right eye 5 weeks following initial presentation. There is optic disc pallor and a subtle retinal pigment epithelial hypopigmentation in the macular area.

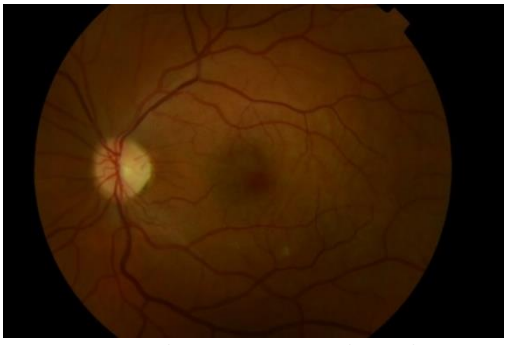

Figure 2: Colour fundus photograph of the left eye showing a cotton wool spot.

Ocular ischaemia secondary to GCA can cause wide ranging symptoms including diplopia, transient loss of all or part of the visual field, transient or irreversible reduced vision, orbital ischaemia and ocular pain secondary to ocular ischaemic syndrome. The most common ophthalmological presentation of GCA is with posterior ciliary artery (PCA) occlusion (81%) causing an arteritic anterior ischaemic optic neuropathy (AAION) [5]. This is associated with a sudden and severe irreversible loss of vision. The optic nerve head is typically swollen with a chalky white appearance. Non-embolic central retinal artery occlusions (CRAO) can also be responsible for loss of vision (14%) along with cilioretinal artery occlusion (22%) [5]. The involvement of more than one arterial territory, as demonstrated by cotton wool spots or a retinal artery occlusion coinciding with an AAION is highly suggestive of GCA. Amaurosis fugax is a worrying symptom, suggestive of a high risk of vision loss [6].

Vision loss in GCA will most often occur in association with an AAION. Isolated acute choroidal ischaemia is a much less frequent presentation, often being more subtle in its clinical picture. As such it may pose a diagnostic challenge for some inexperienced or unwary ophthalmologists and must be considered in elderly patients who have unexplained visual loss with essentially normal fundoscopy. Lack of recognition at this stage would likely result in disease progression and an eventual AAION. Choroidal ischaemia is known to cause white lesions secondary to infarction and subsequent pallor of the RPE and adjacent outer retina [7-9]. The diagnosis of GCA in this case was unfortunately delayed as the patient was misdiagnosed as a CRAO upon initial presentation. In reality the macular ischaemia was secondary to choroidal infarct rather than a CRAO. Figure 3 is an FFA taken 5 weeks after the patient initially presented with vision loss in their right eye which demonstrates delayed choroidal filling, especially prolonged in the macular area and several areas of choroidal ischaemia.

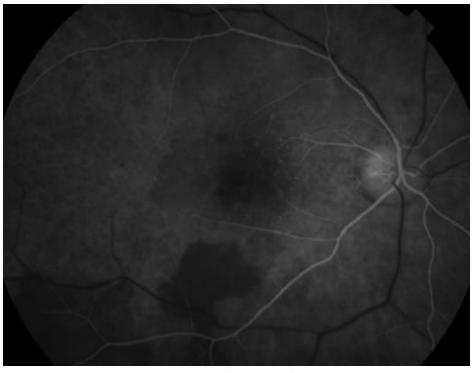

Figure 3: Arterial phase fundus fluorescein angiogram of the right eye showing generalised delayed choroidal filling with a well-defined inferior infarct secondary to a small choroidal artery occlusion. A larger inferotemporal infarct is evident as a wedge-shaped choroidal filling defect, with its apex pointing towards the posterior pole. There is delayed perfusion of the optic nerve, especially nasally.

The anatomy and haemodynamics of the choroidal circulation in health and disease has been expertly described in the past by SS Hayreh and colleagues [9]. The exact pattern of choroidal ischaemia and whether there is an associated AAION is determined by which branches of the PCA are involved and where the watershed areas are located, which can vary between individuals. In our case it is clearly evident that there is a well-defined inferior infarct that is circular in shape secondary to a small choroidal artery occlusion. A larger inferotemporal infarct is evident as a wedge-shaped choroidal filling defect, with its apex pointing towards the posterior pole. This is likely secondary to the occlusion of a larger choroidal artery or a long PCA as described by SS Hayreh [9]. The macular area is also involved with a small, round infarct, likely representing the occlusion of a small terminal choroidal arteriole. There is a subtle temporal macular choroidal filling defect that explains the reduction in vision secondary to macular dysfunction. The severity of choroidal ischaemia can vary significantly ranging from ischaemic dysfunction to frank RPE infarction [9]. The submacular choroid is known to be especially vulnerable to ischaemia, more than any other area of choroid given the multiple watershed zones that meet in this area [9, 10].

Interestingly in our case the patient's visual acuity demonstrated a significant improvement with treatment with high dose oral prednisolone despite a significant delay (5 weeks) in the initiation of treatment. Also, of note is the positive family history for GCA in this case and the relatively young age at presentation. The aetiology of GCA is complex with both environmental and genetic factors seeming to influence both disease development and progression [11]. An increased incidence of GCA has been correlated with epidemics of Chlamydia pneumoniae, parvovirus B19 and Mycoplasma pneumonia in Denmark [12]. An increased knowledge of genetics has recently led to the recognition of genetic susceptibility for GCA. Several familial aggregations of the disease have been shown to share certain HLA alleles that have been implicated in giving genetic influence on the disease susceptibility and severity [11]. Our case here may be yet another example of the importance family history and genetic influence plays for the onset of this disease.

In conclusion, this is an atypical case of visual loss secondary to macular choroidal ischaemia. It highlights that not all cases of GCA with visual loss present with a typical chalky white swollen optic disc secondary to AAION.

Conclusion

GCA can present with ocular features other than AAION. Choroidal ischaemia can be an uncommon manifestation of GCA, along with CRAO and new onset diplopia secondary to cranial nerve palsies. Although visual loss secondary to an AAION is usually catastrophic and irreversible, VA may show improvement if loss of vision occurs secondary to isolated choroidal ischaemia even following a significant delay in the initiation of treatment.

GCA should not be dismissed in patients who present with visual loss and no evidence of papillitis and it should be remembered that mildly raised inflammatory markers are still abnormal and need adequate clinical explanation.

Abbreviations

AAION: Arteritic Anterior Ischaemic Optic Neuropathy
AION: Anterior Ischaemic Optic Neuropathy
BCVA: Best Corrected Visual Acuity
CRAO: Central Retinal Artery Occlusion
CRP: C-Reactive Protein
CT: Computerised Tomography
ESR: Erythrocyte Sedimentation Rate
FFA: Fundus Fluorescein Angiogram
GCA: Giant Cell Arteritis
HLA: Human Leukocyte Antigen
PCA: Posterior Ciliary Artery
RAPD: Relative Afferent Pupillary Defect
RPE: Retinal Pigment Epithelium
TIA: Transient Ischaemic Attack

Conflicts of Interest

None.

Ethics Approval and Consent to Participate

Not applicable.

Consent

Informed consent for publication of the patient clinical details and clinical images was obtained from the patient.

Availability of Data and Materials

The authors declare that all the data in this article are available within the article.

Competing Interests

None.

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Author Contributions

CW conceived and wrote the manuscript. MA edited the manuscript and created images. WMA supervised and edited the manuscript.

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