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Case Report

Giant Cell Arteritis: A Case Report

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Abstract: Giant cell arteritis (GCA) with ocular involvement is an emergency condition, and treatment with glucocorticoids should be provided immediately upon clinical suspicion of GCA to prevent potential blindness. In this case report, we discuss a case of a 70-years-old Caucasian male presented to the emergency department with one day sudden onset of double vision. This is a teaching case report aimed to discuss the management options, potential complications and visual prognosis of this particular case.

Keywords: Giant Cell Arteritis, Arteritic anterior ischemic optic neuropathy (AAION), Non-arteritic ischemic optic neuropathy (NAION)

1. Introduction

Giant cell arteritis (GCA), previously known as temporal arteritis (TA) [1], is a granulomatous vasculitis mainly affecting the medium and large vessels.[2] Ophthalmic manifestations of GCA vary from retinal infarction, ischemia to optic nerve, cortical blindness, pupillary autonomic dysfunction and cranial nerves (CNs) palsies.[3] GCA is a medical emergency and, once suspected, it warrants immediate diagnosis and management to prevent irreversible damage.[1] We report a suspected case of GCA in a patient presenting with headache, visual loss, diplopia and unilateral sixth nerve palsy.

2. Case Report

A 70-year-old Caucasian male presented with one day sudden onset of double vision. Three days prior to his double vision, he noticed his vision blacked out for 5 seconds, then returned in his left eye. He complained of malaise, new-onset headache predominantly on the left side of his head, and neck muscle pain. There was no history of headache, myocardial infarction or cerebrovascular accidents. His past ocular history included bilateral lens extraction ten years earlier, with good vision. His past medical history included well controlled type II diabetes and well controlled hypertension. He had a slight limp due to a severe fracture in his leg whilst heli-skiing at age of 30 years. His social history was unremarkable. Comprehensive ocular examination was completed: visual acuity (VA) was 6/6 in the right eye and 6/60 in the left eye. The right eye was normal, with normal eye movements, colour vision and field testing. IOP was normal bilaterally. The left eye showed left relative afferent pupillary defect (RAPD) and abducens (sixth) nerve palsy. Following mydriasis, fundoscopy of the left eye revealed pale optic disc swelling with indistinct binderies and retinal whitening at the distribution of cilioretinal arteries (Figure 1). Fundoscopy of the right eye was otherwise normal. Baseline blood lab requested previously by his GP showed normal Hb (Hb:13g/dl, normal ranges for males 13 to 18 g/dl), high ESR (ESR:105, normal range for male 1-10 mm/hr), normal urea (urea: 7 mmol/L, normal range 2.5 to 7.1 mmol/L), high creatinine (creatinine: 170 μmol/L, normal range for male adult 60 - 110 μmol/L), normal HbA1c (5%, normal

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range of type 2 diabetics ≤ 6.5%), normal TFTs and normal LFTs. A clinical suspicion of GCA was made on the basis of our findings.

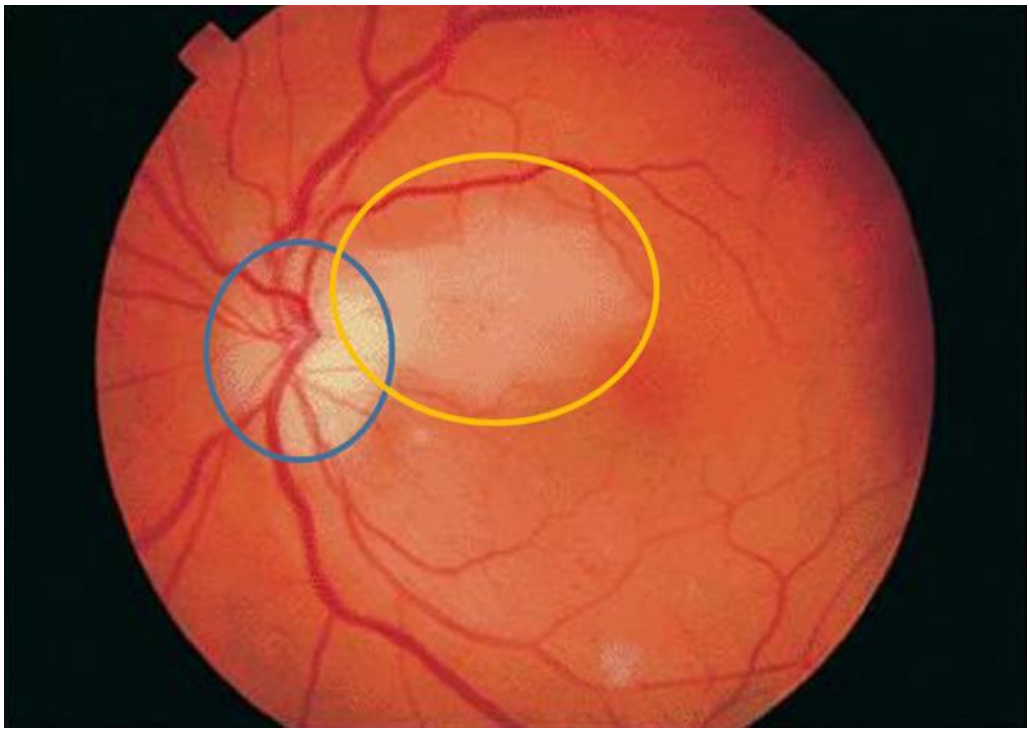

Figure 1. Fundus photography of the left eye revealed pale optic disc swelling with indistinct boundaries (blue circle) and retinal whitening at the distribution of cilioretinal artery (yellow circle); Image taken from: Wills Eye Hospital Atlas of Clinical Ophthalmology.

3. Clinical Features

GCA is the most common cause of vasculitis in patients above 50 years, with a mean age of onset of 70.[1] Women are 2.5 times more likely to have GCA than men, with a lifetime risk of 0.5% for men and 1% for women.[1, 4] GCA has an incidence of 20 per 100,000.[1] GCA significantly affects Caucasians and is more prevalent in Northern European countries, especially Scandinavian countries.[1, 5] It is rare in Black African/Caribbean and Asian populations.[1, 6] There is a strong association between human leucocyte antigen (HLA) and GCA, especially HLA DR4-01.[6] A correlation between the onset of GCA and infection, for example Epstein–Barr virus and herpes simplex virus, has been suggested.[6]

4. Pathology

Granulomatous inflammation with giant cells, lymphocytes and epithelioid cells. There is also disturbance of internal elastic lamina and proliferation of the intima with aneurysm formation (Figure 2).[6]

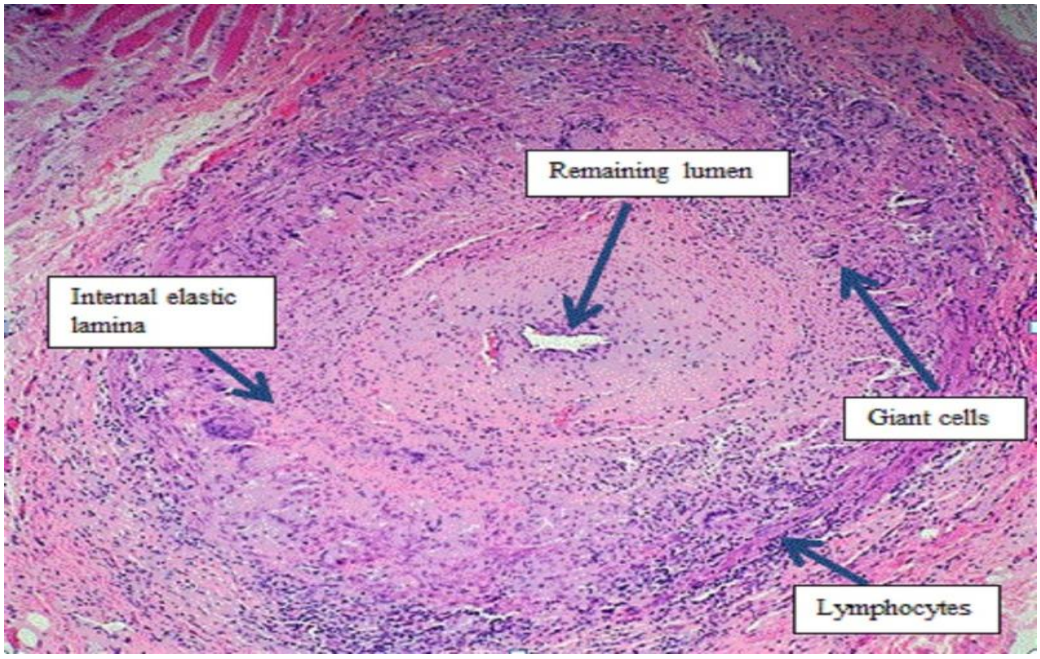

Figure 2. Transmural granulomatous inflammation of all layers of temporal artery, with lymphocytes, giant cells and fragmentated the elastic laminae.[3]

5. Clinical Presentation (Systemic and Ophthalmic Manifestation)

5.1. Systemic Manifestation

The onset of GCA can be insidious or acute.[1] Typical symptoms of GCA include new-onset headache, jaw claudication, and scalp tenderness. [1, 6, 7] Other constitutional symptoms such as fatigue, fever, malaise, anorexia, weight loss and polymyalgia are common.[1, 6, 7] Headaches, which typically occur in 70-80% [1] of patients, may be described as an ache, and are usually located in the temporal region.[1, 6] Although approximately 33% of diagnosed GCA patients have normal temporal arteries, [6] findings such as temporal artery prominence, irregular contour, and pain with no pulse on clinical examination, increase the ratio for a positive temporal artery biopsy (TAB) (Figure 3).[8]

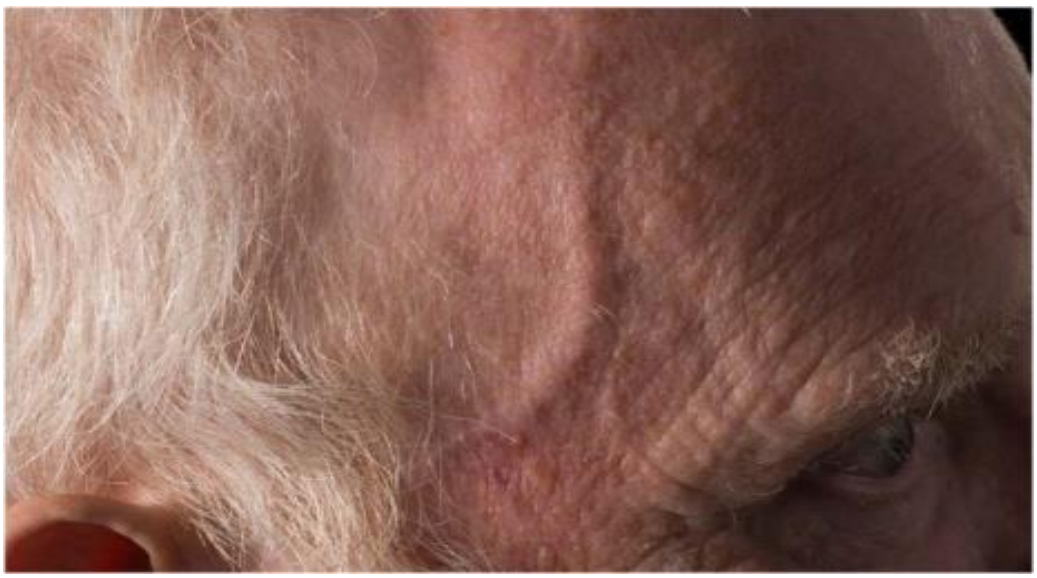

Figure 3. Beaded, prominent, pulseless temporal artery in a patient with positive TAB proven GCA.[6]

Scalp tenderness is often a warning sign and precedes a headache by a few weeks.[1] Jaw claudication presents in under 50% of GCA patients while tongue claudication can be present but is less common.[6, 9] Patients may present with polymyalgia rheumatic (PMR) in 50% of cases, where they suffer from prominent muscle and morning stiffness mainly in the proximal griddle locations.[1] Other GCA complications include large-vessel aneurysms and stenosis such as thoracic and abdominal aortic aneurysm.[6] Clinically, they may present as chest, back pain and intermittent limb claudication.[6] Rarer complications include scalp necrosis and stroke.[6]

5.2. Ophtalmic Manifestation

Arteritic anterior ischemic optic neuropathy (AAION), headed by amaurosis fugax, is the foremost cause of irreversible visual loss in patients with GCA.[10] Amaurosis fugax may lead to permanent loss of vision if disregarded, in which there is a total vision black out for few seconds[1], most commonly because of temporary ischemia of the optic nerve head.[10] Visual loss occurs because of occlusion mainly in the PCAs, infrequently the CRA and rarely the ophthalmic artery.[10] Visual loss is mainly unilateral, however, bilateral loss of vision has been reported.[10] Approximately 50% of affected eyes have hand movement vision or worse, with RAPD.[10] Ocular motor imbalance and diplopia occurs in 2-15% of patients with GCA because of ischemia affecting brainstem, ocular motor muscles and extraocular muscles.[10] GCA can affect various cranial nerves (CNs) including bilateral abducens nerve[12] and unilateral abducens nerve[11]. Figure 4 illustrates the blood supply to the optic nerve and the posterior segment of the eye.[14]

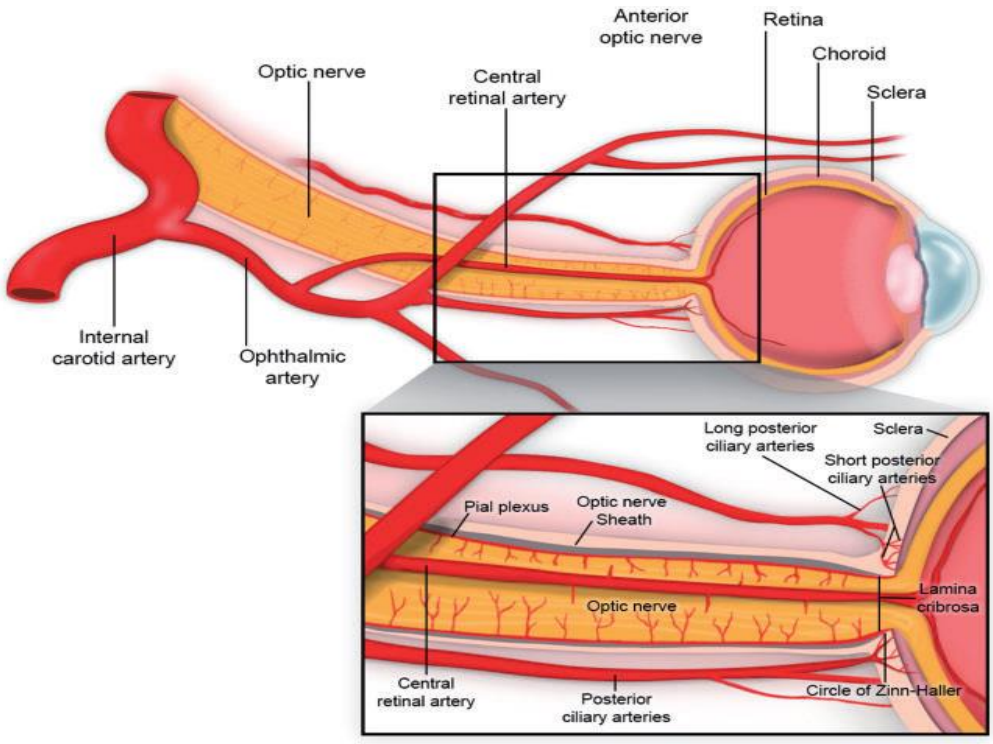

Figure 4. GCA in the orbit disrupts the ophthalmic artery (OA) and its branches, especially the central retinal artery (CRA) and posterior ciliary arteries (PCAs). The PCAs supply the choroid, which feeds the photoreceptors in the optic nerve head and outer third of the retina. The CRA perfuse blood to the inner two thirds of the retina. The ophthalmic artery also supplies blood to the extraocular muscles. Ischaemia of these structures causes diplopia and ophthalmoparesis.

Moreover, GCA can affect intracranial and extracranial blood vessels, producing homonymous loss of the visual field because of stroke in the occipital cortex.[14]

6. Differential Diagnosis (DDX)

Non-arteritic ischemic optic neuropathy (NAION) should be differentiated from occult GCA.[10] Unlike GCA, NAION has no systemic symptoms, amaurosis fugax or haematological abnormalities.[15] In contrast to NAION, AAION usually has chalky white optic disc oedema.[10] PMR and GCA are closely related disorders with PMR characterised by morning stiffness and pain in the muscles of shoulder, cervical region and pelvic joints, with no vision loss.[10] Connective tissue disease and vasculitis that overlap or mimic GCA, including systemic lupus erythematosus and rheumatoid Arthritis can be considered as other DDX for GCA.[10] These conditions, unlike GCA, involve multiple organs. Other DDXs to consider if the patient is presenting only with systemic symptoms include myasthenia gravis, thyroid dysfunction, malignancy, myeloma, amyloidosis and tuberculosis.[10]

7. Diagnosis

Diagnosing GCA can be challenging as the symptoms are non-specific and because there is a wide variety of phenotypes,[6, 16] and requires a thorough examination combined with proper investigations.[17] GCA must be suspected in patients presenting with new-onset headaches and other symptoms (mentioned previously).[1]

8. Ophthalmic Examination

If GCA is suspected, a thorough assessment of VA, colour vision, RAPD, ocular motility, IOP, anterior segment and fundoscopy examination for the posterior segment following dilatation should be conducted.[14] Automated visual fields testing should be completed promptly if a patient presents with visual loss.[14] Fundoscopic findings (Figure 5) include signs of ischemia, such as cotton wool spots and pallid nerve head in acute presentation, and optic disc pallor with cupping as a late GCA sequalae.[14]

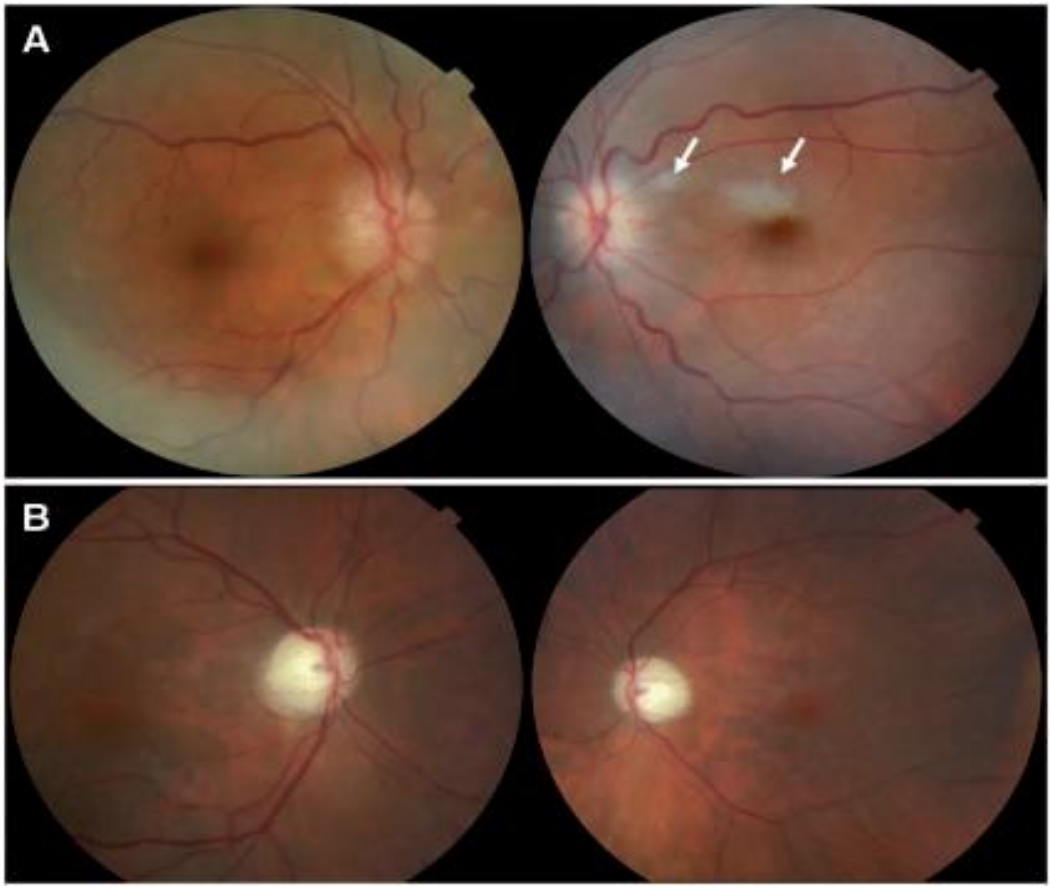

Figure 5. Fundoscopic findings in GCA patients. (A) Fundi of patient presenting with acute bilateral loss of vision due to GCA. There is bilateral pallid oedema of the optic nerve head and cotton wool spots (white arrows) in the left eye.[14] (B) Late sequalae of AION showing bilateral optic disc pallor with cupping.[14]

Fluorescein angiography demonstrates hypoperfusion and delayed perfusion of choroid, retina or both in GCA.[14] A highly suggestive feature of GCA is large swathe of choroidal hypoperfusion (Figure 6).[14] Laboratory blood test includes erythrocyte sedimentation rate (ESR) and C- reactive protein (CRP) are characteristically elevated[1, 6] with ESR>50.[1] However, in some cases, inflammatory markers can be normal.[18] Other serological markers that may predict the diagnosis of GCA include anaemia and mildly elevated liver enzymes.[1, 6]

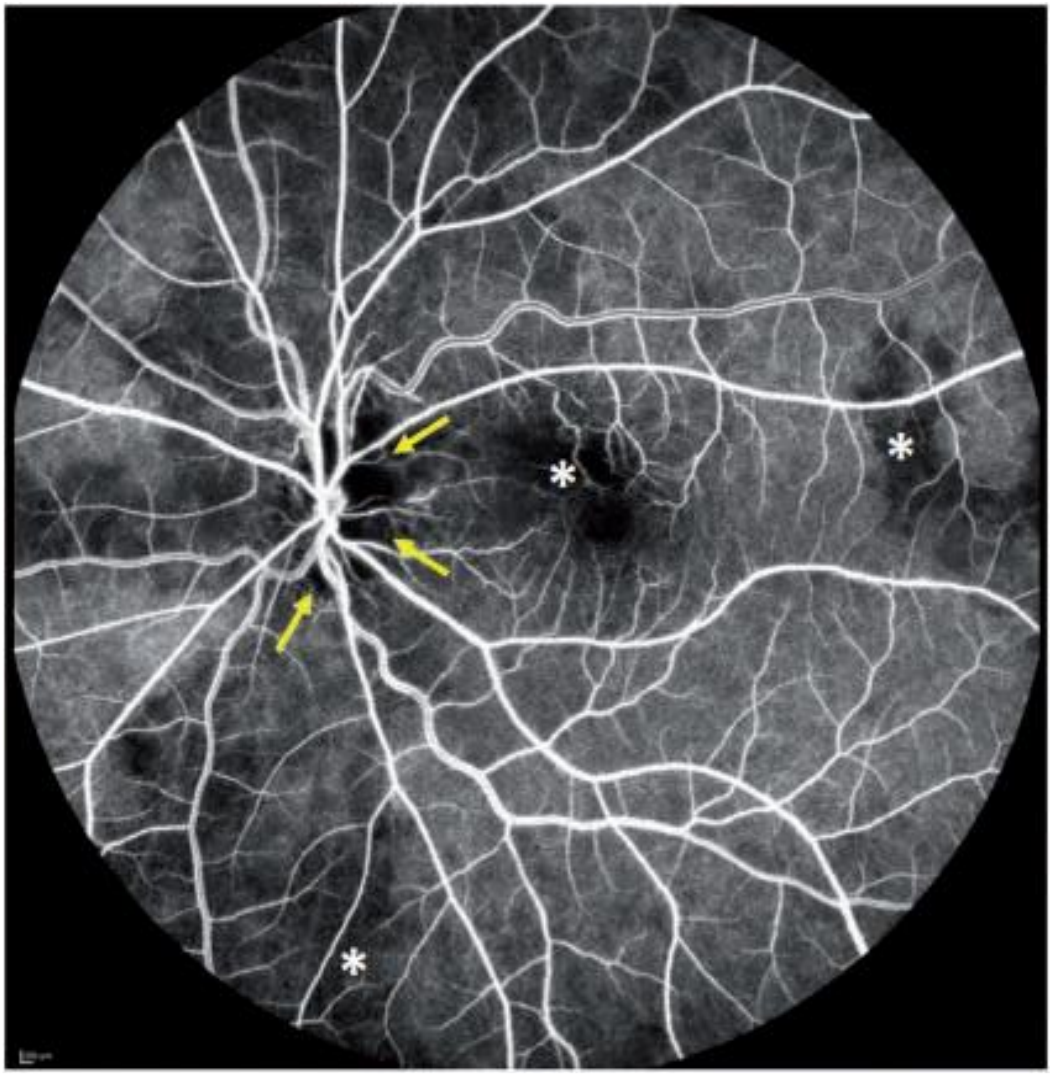

Figure 6. The optic nerve head is demarcated by yellow arrows. Swathe of choroidal hypoperfusion are illustrated (asterisks). [14]

The gold standard investigation to diagnose GCA is TAB, however, it may be negative in 30% of cases.[1, 19] Skip areas are common, thus a larger sample size (>2cm) is recommended when obtaining a TAB.[1] Classical pathological findings are illustrated in Figure 2. TAB should not delay initial treatment and needs to be done within 2 weeks of starting it.[1] The American College of Rheumatology (ACR) established the diagnosis criteria for GCA.[20] These criteria include new onset headache, age of onset ≥50 years, ESR> 50mm/hr, temporal artery tender/reduced pulsation and temporal artery biopsy demonstrating vasculitis with multinucleated giant cells or granulomatous inflammation.[20] If the patients score 3 or more of the 5 criteria, they will have a specificity of 91.2% for GCA and a sensitivity of 93.5% [20, 21] Imaging modalities such as ultrasonography (US) and magnetic resonance imaging (MRI) can be used to determine the presence of GCA.[19] Vascular US can be utilised to detect temporal artery occlusion, oedema and stenosis in GCA. Typically there is a "halo sign", which is a non-compressible hypoechoic ring surrounding an arterial lumen that signifies an edematous thickening of the arterial wall because of inflammation.[6] MRI can reveal non-specific inflammation signs such as enhancement of the optic nerve, perineural sheath or chiasm (Figure 7).[14] If GCA patients present with systemic symptoms, other screening tests can be done,

including anticardiolipin antibodies,[10] TFTs, anti-cytoplasmic neutrophil antibodies, dipstick urinalysis, chest X-ray and echocardiogram.[14]

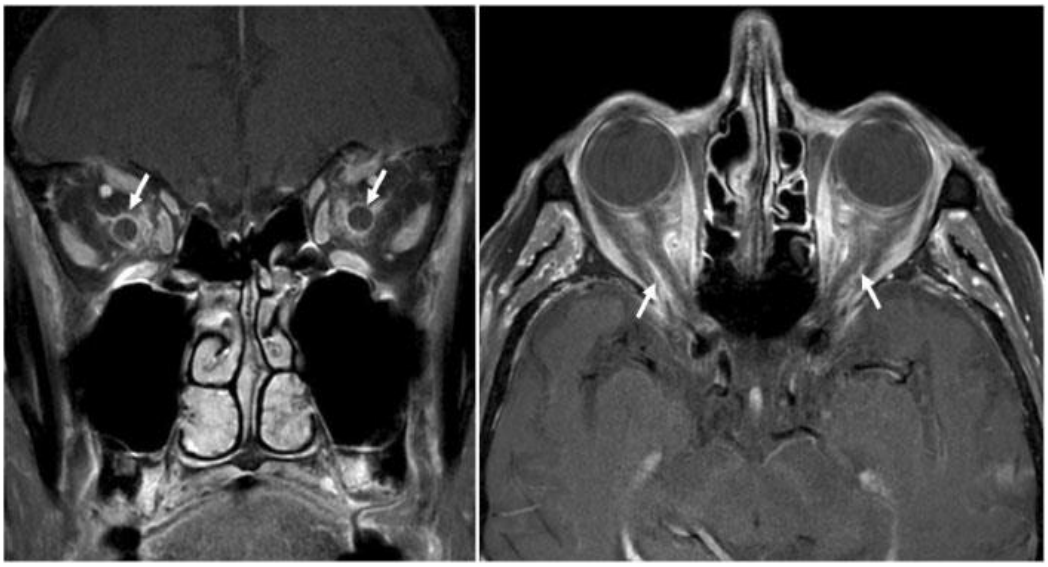

Figure 7. Coronal and axial contrast-enhanced T1-weighted fat-suppressed MRI of the orbits indicating optic nerve sheaths enhancement (white arrows).[14]

9. Management

Upon suspecting GCA, glucocorticoid (GC) should be started immediately to avoid irreversible damage.[6, 14] Patients presenting with cranial ischemic symptoms and/or ophthalmic symptoms should be commenced on high dose pulse treatment of GC with intravenous 500-1000mg methylprednisolone for 3-5 days. The patient can then be switched to high-dose of oral prednisone (100-120mg/day).[6, 14] The EULAR and BSR guidelines recommend instant GCA management with 1 mg/kg GC (up to 60 mg/day) or 40–60 mg/day.[6] This is to lower the incidence of ischemic complications, particularly to avoid vision loss.[6] An initial dose of 40 mg/day is considered sufficient for patients without cranial ischemic symptoms.[6] Tapering the dose and duration depends on the patient's response.[6] The BSR guidance suggests tapering of GC over a period of 1-2years according to the response.[22] Glucocorticoids, although beneficial, may have significant adverse effects, especially for doses above 40mg/day.[6, 14] The determination of previous comorbidities such as cataract, diabetes mellitus hypertension, peptic ulcer disease, osteoporosis, cardiovascular disease and glaucoma is, therefore, crucial as GC may worsen disease process.[6] Low dose aspirin has been utilised to treat GCA as an adjunctive therapy, however, there are no randomised control trials assessing the role off aspirin in preventing ischemic complications.[14] Recently, the function of toclilizumab in GCA has been assessed and it demonstrated that toclilizumab improves the risk-benefit long-term profile for GCA management.[23] However, the study did not evaluate the effect of toclilizumab and its dosing protocols on vision complications.[23] Our patient presented with symptoms, signs and high inflammatory markers suggestive of GCA. Treatment must be provided immediately, as detailed above, to avoid any irreversible outcomes. The patient had multiple pre-existing comorbidities, including diabetes, hypertension and cataract, which warrants careful administration/ prescription of GC and follow-up.

10. Conclusion

GCA can present with a wide range of symptoms and signs including new-onset headache, horizontal diplopia, malaise, unilateral sixth CN palsy and RAPD. GCA is an

emergency condition, and treatment with GC should be provided immediately upon clinical suspicion of GCA to prevent potential blindness.

Consent: Informed consent was obtained from the patient to publish their anonymised data.

Interest of conflict: Author declares no interest of conflict.

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