Atypical Giant Cell Arteritis Causes Bilateral Vision Loss Despite Normal Inflammatory Markers and Negative Ultrasound

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01. Introduction

In suspected giant cell arteritis (GCA), treatment decisions must prioritise clinical suspicion over reassuring investigations [1].

We present a case of bilateral arteritic anterior ischaemic optic neuropathy occurring in the setting of low-grade inflammatory markers and a negative temporal artery ultrasound, reinforcing that diagnosis in GCA remains fundamentally clinical.

02. Timeline

Week 0: Initial Assessment

79-year-old Female
- Progressive bilateral painless vision loss (4 weeks)
- VA 6/5 (right), 6/6 (left) to 6/60 bilaterally
- **Headache + proximal girdle pain initially misattributed**
- **Jaw pain not initially recognised as claudication**
- CRP 5.9 mg/L, ESR 37 mm/hr, and platelets 422 ×10⁹/L
- MRI unremarkable

Week 3: Ongoing Concern

- **Temporal artery ultrasound negative**
- Jaw claudication confirmed
- No alternative diagnosis identified.

Week 11: Ischaemia Identified

- Thickened, non-pulsatile temporal artery
- **FFA: Marked delayed choroidal filling, supporting arteritic AION**
- IV methylprednisolone initiated

Week 16+: Diagnosis

- Temporal artery biopsy: Positive
- Subsequent systemic vascular instability, supporting wider vasculitic burden

03 Investigations

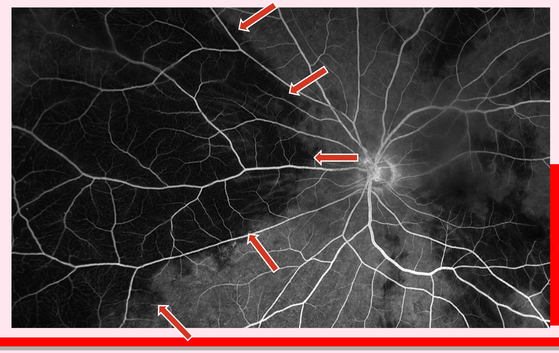

Figure 1. 31-second FFA of right eye demonstrating patchy delayed choroidal filling

04. Discussion

Why is this case important?
- High clinical probability of GCA (GCAPS >10) at presentation
- Bilateral vision loss and red flag features present
- Corticosteroid initiation was significantly delayed

Diagnostic tension
- Headache and polymyalgia initially attributed to chronic disease/MSK pathology
- Jaw pain not recognized as claudication early
- Inflammatory markers were only mildly elevated
- Negative temporal artery ultrasound provided false reassurance

What the literature says
- ESR, CRP and platelets may be normal or mildly elevated in biopsy-proven GCA cases [2]
- Ultrasound sensitivity is variable and operator dependent [1]
- Temporal artery biopsy remains the diagnostic gold standard [1]
- Delayed choroidal filling on FFA reflects posterior ciliary artery hypoperfusion and supports arteritic disease
- Visual loss is often sudden and irreversible

Key reflection
- GCA remains a clinical diagnosis
- Negative investigations do not exclude disease
- Treatment should not be delayed when clinical suspicion is high

GCA Probability Score

	Score	-3	0	1	2	3
Demographics	Age (Years)		≤49	50-60	61-65	≥66 ★
	Sex			M	F ★	
Duration	Onset of symptoms		>24 weeks	12-24 weeks	6-12 weeks	<6 weeks ★
Laboratory	CRP		0-5 mg/L	6-10 mg/L	11-25 mg/L	≥25 mg/L
Symptoms	Headache		N	Y ★		
	Polymyalgic		N		Y ★	
	Constitutional		N	Single		Combination
	Ischaemic		N			Y ★
Signs	Visual (AION, CRAO, Field loss, RAPD)		N			Y
	Temporal artery abnormality		N	Tenderness	Thickening	Pulseless ★
	Extra-cranial artery abnormality		N	Thickening	Bruit	Pulse loss
	Cranial nerve palsy		N			Y
Alternative	Alternative diagnosis	Y	N			

Table 1: GCAPS at presentation indicated high probability of GCA despite low-grade inflammatory markers and negative ultrasound. Stars indicate features identified during diagnostic pathway. [3] GCAPS: 11-17 = High probability. Atypical headache, polymyalgia and temporal artery abnormality were recognised at a later presentation.

05. Conclusion

Negative ultrasound and low-grade inflammatory markers do not exclude GCA in high-risk patients (GCAPS ≥10).
Diagnosis remains clinical.
Key actions:
- ❖ Start corticosteroids immediately when GCA is suspected with visual symptoms
- ❖ Do not delay treatment for confirmatory testing
- ❖ Urgent fast-track rheumatology referral is required

This case highlights how over-reliance on investigations can delay treatment and contribute to irreversible visual loss.

06. References

1. Bilton EJ, et al. Eye. 2023;37:2365–73.
2. van der Geest KSM, et al. JAMA Intern Med. 2020;180:1295–304.
3. Laskou F, et al. Clin Exp Rheumatol. 2019;37(Suppl 117):104–8.