Clinical Reasoning: A case of bilateral orbital mass lesions presenting with acute monocular vision loss

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Section 1

An 80-year-old man with medical history of hypertension developed a sudden loss of vision in the left eye without any associated pain, flashes, or floaters. The review of systems was negative for any headaches, jaw claudication, scalp or temporal tenderness, or other symptoms suggestive of polymyalgia rheumatica (PMR). The patient's medications were benazepril and amlodipine for the treatment of hypertension. Clinical examination showed normal vital signs. Visual acuity testing revealed 20/20 vision on the right and hand movement perception only in the temporal field of the left eye, with no light perception on the nasal field of the same eye. Pupils were 3 mm bilaterally, round, and reactive with a relative afferent pupillary defect on the left. Dilated funduscopic examination was unremarkable on the right side and revealed macular whitening and retinal blanching with cherry-red spot on the left. Temporal artery pulses were present. The remainder of the neurologic examination was unremarkable. Laboratory investigations including erythrocyte sedimentation rate, C-reactive protein, lipid profile, and glycosylated hemoglobin were normal.

Question for consideration:

1. What is your differential diagnosis?

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Section 2

A careful ophthalmologic history is an essential initial step in the evaluation of vision loss. Monocular vision loss localizes to an intrinsic ocular pathology (cornea, anterior chamber, or lens) or retinal or prechiasmatic lesions. Patients with primarily mechanical retinal lesions such as retinal detachment typically complain of photopsia before the vision loss whereas vascular lesions of the optic nerve or retina present with sudden, painless, altitudinal, or complete vision loss. Acute lateralized monocular vision loss with relative sparing of the temporal field was suggestive of an intraorbital lesion compressing the optic nerve. An MRI of the orbits with and without contrast was performed, which revealed bilateral enhancing intraconal mass lesions (figure, A), left greater than right. However, the funduscopic findings in this case were consistent with acute central retinal artery occlusion (CRAO). Therefore, the following differential diagnosis was considered in this patient:

1) CRAO is caused by occlusion of the central retinal artery, most commonly from an embolic source. Common etiologies include atherosclerotic plaques along the course of the carotid arteries, aorta, or a cardioembolic phenomenon. CRAO usually presents with an acute monocular vision loss involving the entire visual field. Infrequently, CRAO can be secondary to an inflammatory arteritis, giant cell arteritis (GCA) being the most commonly diagnosed underlying condition. The patient underwent a CT angiogram of the head and neck, which was unremarkable, and a cardiac workup including a 2D echocardiogram and 48-hour cardiac monitoring did not reveal a potential cardioembolic source of ischemia. As mentioned earlier, ESR and CRP were normal, and the patient did not have any systemic symptoms suggestive of GCA.

2) Compressive optic neuropathy from a space-occupying lesion in the orbit usually presents with gradual vision loss and multiple cranial neuropathies along with exophthalmos.

Orbital cavernous venous malformations (cavernous hemangioma) are the most common vascular lesion of the orbit in adults, accounting for 5%–7% of all orbital mass lesions. Although they are largely asymptomatic, sometimes they present with painless, progressive proptosis rather than acute isolated loss of vision.

Orbital pseudotumor is a nonmalignant idiopathic inflammation of the orbital tissues and the most common cause of painful orbital mass in adults. Patients commonly present with pain, proptosis, ptosis, periorbital edema, and restriction of ocular motility. It seldom presents with isolated acute vision loss, especially in the absence of the features mentioned above. Based on its location, it includes conditions such as dacryoadenitis and autoimmune myositis. Tolosa-Hunt syndrome is the term used to indicate its involvement of the cavernous sinus.

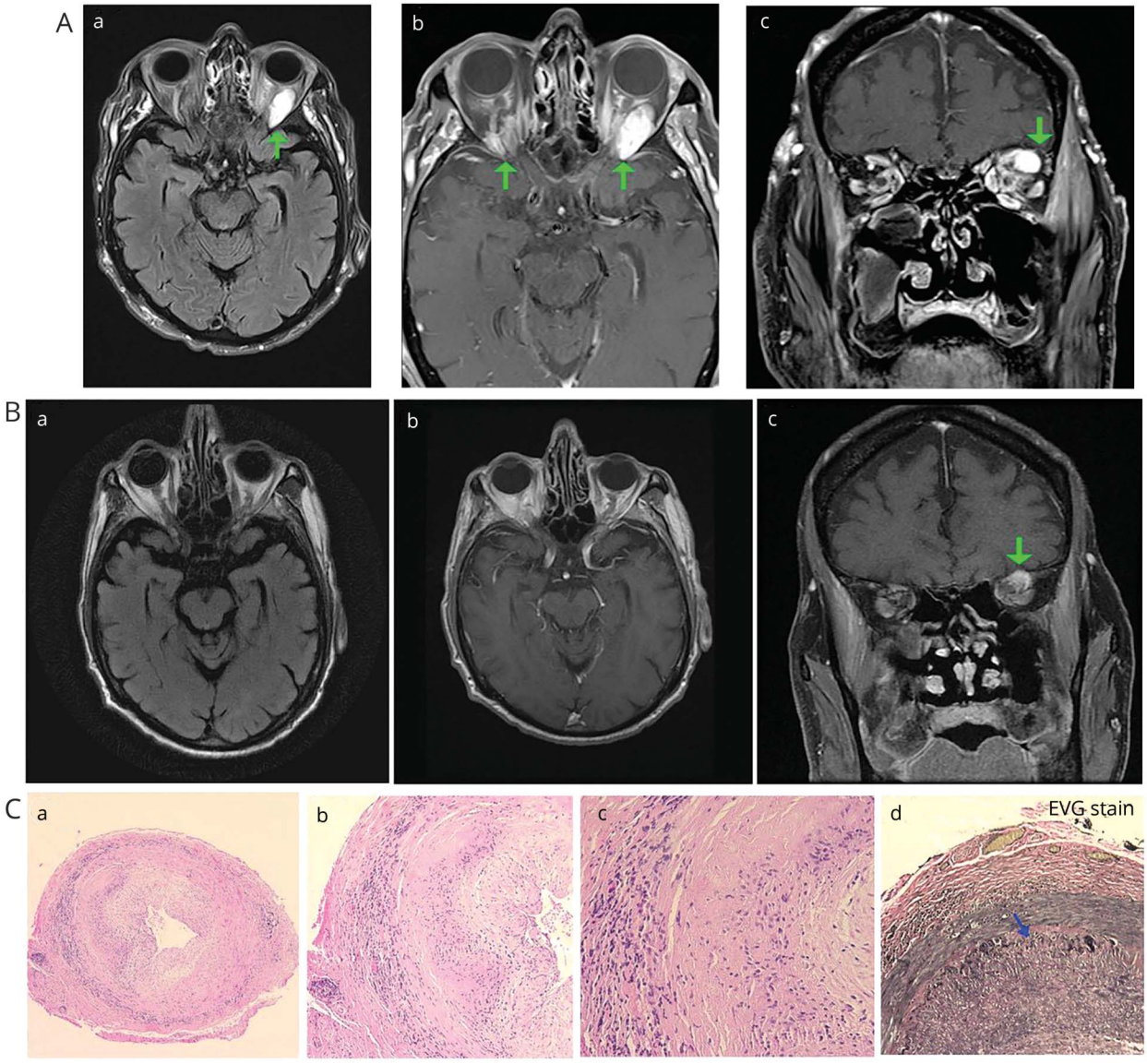
Cavernous-carotid fistula is an abnormal communication between the internal carotid artery and the cavernous sinus and can be traumatic or spontaneous. It presents with chemosis, proptosis, pulsatile exophthalmos, and other cranial nerve deficits. It can lead to ophthalmic vein varices that over time may also lead to central retinal vein occlusion.

3) Although ischemic optic neuropathy (ION) commonly presents with altitudinal visual deficit, it is worth considering in a patient with acute vision loss and vascular risk factors.

Nonarteritic ION is the most common form of ION. It is analogous to small vessel disease stroke of the optic nerve and has a similar risk factor profile including hypertension, diabetes mellitus, hypotension, and rarely use of vasopressor medications. The clinical features are swollen optic disc, decreased cup-to-disc ratio (normally 0.3), and uncommonly, simultaneous involvement of the fellow eye.

Arteritic ION usually occurs in patients over 70 years of age. GCA is the most common etiology. It is usually accompanied by local and systemic features suggestive of an underlying inflammatory condition such as new-onset headache, jaw pain, fever, myalgias and weight loss. Simultaneous involvement of the fellow eye is uncommon.

Figure MRI brain



(A) (A.a: axial FLAIR; A.b: axial T1 + contrast; A.c: coronal T1 + contrast). MRI brain shows bilateral orbital enhancement, left greater than right (green arrows). (B) (B.a: axial FLAIR; B.b: axial T1 + contrast; B.c: coronal T1 + contrast). MRI brain shows resolution of bilateral orbital enhancement (green arrow). (C) (C.a, C.b, C.c ×4, ×10, ×40): panarteritis. (C.d [×10]): elastic Van Gieson stain shows fragmentation of the internal elastic lamina (blue arrow).

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Section 3

Questions for consideration:

1. What are the next steps in the management of this patient?
2. What additional tests would you order to narrow the differential diagnosis?

A complete workup for an embolic source remained negative. Keeping the possibility of an uncommon presentation of a common and treatable condition in mind, the diagnosis of GCA was considered in this elderly man with vision loss, despite the lack of usual systemic and inflammatory markers associated with GCA. Given the time sensitivity of GCA treatment, empiric therapy with steroids was initiated (1,000 mg/d for 3 days, followed by 60 mg orally daily with a tapering schedule afterwards). A cerebral angiogram did not reveal any arterial or venous pathology. The patient's symptoms improved marginally, with perception of hand movements in both temporal and nasal field of the left eye. A repeat MRI of the brain and orbit was performed 3 days after the treatment that showed a significant reduction in the size of the orbital mass (figure, B). Eventually a temporal artery biopsy (TAB) showed transmural inflammation with histiocytes and lymphocytes along with moderate amount of intimal hyperplasia and fragmentation of the internal elastic lamina, thus confirming the diagnosis of GCA (figure, C)

Discussion

Monocular loss of vision is an ocular emergency. Urgent assessment, rapid diagnosis, and early treatment initiation has been shown to improve clinical outcomes and reduce disability. We present a case of GCA presenting as lateralized monocular vision loss in a context of bilateral intraorbital mass lesions that responded to steroids.

While the differential diagnosis of the intraorbital masses includes intraorbital lymphoma, sarcoidosis, and meningioma, we propose that these lesions were perhaps a manifestation of a local inflammatory response along the GCA spectrum, as has been previously reported in literature.[1–3] Furthermore, the lack of a ring-enhancing appearance of these masses argues against a lymphoma. Similarly, response to steroids and the imaging features, both on MRI and head CT, do not support the likelihood of a meningioma. Sarcoidosis is a systemic granulomatous inflammatory disorder and can present similarly but remains low on differential in a man with no history of any respiratory conditions. The lack of the intraorbital mass biopsy is a limitation of this report. The biopsy was deferred due to the rapid reduction of the size of this mass associated with improvement in symptoms.

GCA is an inflammatory vasculopathy affecting medium and large vessels with well-developed vasa vasorum.[4] Given how rare coexisting intraorbital inflammation is, any accurate estimates of their incidence along with GCA are not currently available.[5,6] However, unlike the current case, orbital inflammation is also accompanied by other typical GCA clinical features including new-onset headache, scalp tenderness, jaw and tongue claudication, systemic symptoms such as fever, weight loss, malaise, and joint pain, and increased inflammatory markers. PMR is found coexistent with GCA in about 30%–40% of cases.[4] The diagnosis heavily relies on the presence of abnormal inflammatory markers. In fact, a large cohort study showed that only 4% of biopsy-proven GCA cases have normal inflammatory markers.[7] TAB remains the gold standard in diagnosing this condition and characteristically shows panarteritis (figure, C.a, C.b, C.c). The presence of giant cells is not required to make the diagnosis. The fragmentation of internal elastic lamina is a highly pathognomonic feature of GCA (figure, C.d).

Rapid institution of glucocorticoids (GC) remains the mainstay of treatment of GCA, with variable duration of treatment depending on individual treatment response as GCA is associated with a high risk of relapse.[8] Several steroid-sparing agents like TNF-α inhibitors have also been studied with varying degree of success.[9] More recently, tocilizumab, an interleukin-6 α receptor blocker, has been shown to reduce relapse rates when used with GC in a phase III study.[10] Although these results are promising, more research is needed to evaluate the safety and efficacy of this agent.

This case represents an uncommon presentation of a common condition and emphasizes the importance of considering GCA in the differential diagnosis of any form of acute monocular vision loss. The atypical features of this case, including the lack of headache, myalgia, and other GCA systemic symptoms with normal inflammatory markers and the presence of bilateral orbital mass lesions made for a challenging diagnosis. The treatment of GCA is time-sensitive and delayed treatment is associated with poor visual outcomes. Despite several atypical features, a high vigilance for the possibility of this condition led to the correct diagnosis and prevented a potentially poor long-term visual outcome.

Author contributions

Dr. Bhatt: study concept and design, analysis and interpretation of data, critical revision of manuscript for intellectual content. Dr. Landman: study concept and design, analysis and interpretation of data. Dr. Sidani: analysis and interpretation of data. Dr. Asdaghi: study supervision, study concept and design, analysis and interpretation of data, critical revision of manuscript for intellectual content.

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Disclosure

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