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Short Communication

When Sight Fails Suddenly: Acute Vision Loss in a Tricenarian

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Abstract

We describe the case of a 36-year-old right-handed businessman who presented with acute, painless bilateral vision loss. Initial clinical and ophthalmologic examinations did not reveal ocular pathology, and neurophysiological testing showed only borderline abnormalities. Neuroimaging identified bilateral parieto-occipital lesions, and subsequent vascular imaging demonstrated severe intracranial internal carotid artery occlusion with collateral vessel formation consistent with Moyamoya vasculopathy. Further review of medical history revealed underlying rheumatoid arthritis with positive RA factor and anti-CCP antibodies, suggesting autoimmune-mediated chronic vasculopathy as the etiology. This case highlights the diagnostic complexity of sudden bilateral vision loss in young adults and emphasizes the rare but significant association between rheumatoid vasculitis and secondary Moyamoya syndrome.

Keywords: *Moyamoya Syndrome, Rheumatoid Vasculitis, Bilateral Vision Loss*

Section 1: Patient Presentation

A 36-year-old right-handed businessman presented with a history of sudden, painless bilateral loss of vision, which began abruptly on 19 March 2021. The visual loss was profound, with complete inability to perceive visual stimuli, and occurred without associated neurological features such as diplopia, headache, cranial nerve deficits, limb weakness, sensory disturbances, seizures, or transient visual obscurations. There was no preceding febrile illness, vaccination, or trauma to suggest an inflammatory or infectious prodrome. Visited us on 28th june 2021, the patient reported approximately 90% spontaneous improvement in vision. The patient initially consulted an ophthalmologist for sudden bilateral vision loss, where he received multivitamin injections, and oral tablets. During this evaluation, he was also found to have newly detected hypertension, for which he was started on Amlodipine 2.5 mg once daily. Past medical history was notable for cataract surgery in 2012 at the age of 27 years, without further documented evaluation. There was no prior history of similar visual complaints. The patient also had a 4 year history of small and large joint pains, previously diagnosed as Rheumatoid Arthritis (RA), for which he was treated with methotrexate for one year before discontinuing therapy. He has remained asymptomatic for the past 3 years. The patient reports chronic tobacco chewing for the past 12 years. Examination at our centre showed blood pressure of 140/90 mmHg. Neurologic examination revealed normal pupils, preserved color vision, intact visual fields, normal discs, hypertensive retinopathy changes grade 2, and normal ocular movements without any motor deficits. Higher mental function tests revealed error in flower pot drawing and error in calculation with normal Albert's line test [Figure 1].

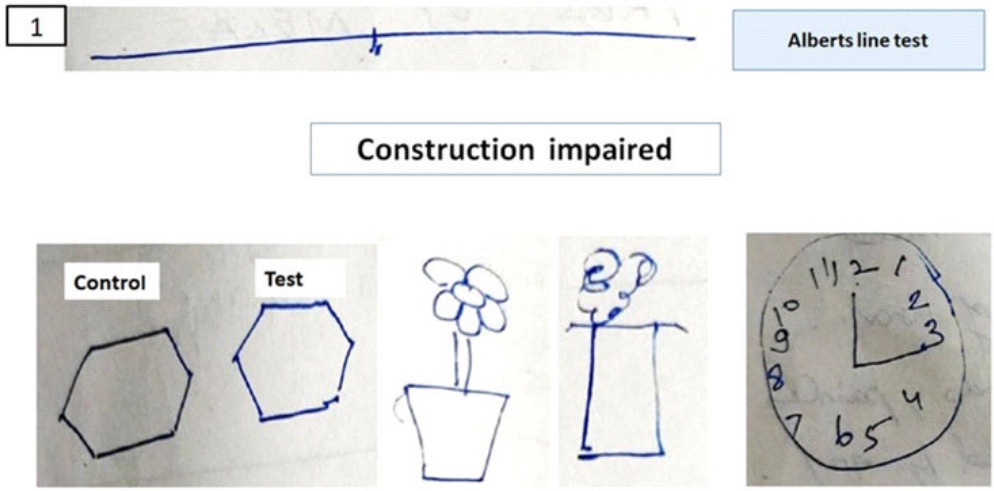

Figure 1. Shows error in flower pot drawing, clock drawing with normal Albert's line test.

Questions (Clinical Reasoning)

1. Which differentials should be considered for acute, bilateral, painless visual loss in a young male, given the historical clues?

2. What is the localization of lesion based on clue from neurological examination?

3. What vascular or systemic conditions in young adults can produce such clinical syndrome?

4 What investigation will narrow the differential diagnosis?

Section 2: Answering the Questions

1. The differential diagnosis for acute, bilateral, painless visual loss in a young male includes optic nerve, retinal, and cortical causes based on the historical clues. Optic neuropathies remain primary considerations, including a demyelinating process presenting as painless papillitis, Leber hereditary optic neuropathy (particularly relevant given the history of cataract surgery at a young age and treatment given), and toxic or nutritional optic neuropathy associated with chronic tobacco use. Retinal contributions such as acute-on-chronic hypertensive retinopathy may further account for the visual impairment, with tobacco exposure potentially adding a component of toxic retinopathy or tobacco amblyopia. Additionally, bilateral occipital lobe involvement leading to cortical blindness must be considered, with differentials such as posterior reversible encephalopathy syndrome (PRES), cerebral rheumatoid vasculitis (history of RA) and MELAS, especially in the context of young age and early-onset cataracts.

2. The neurological examination localizes the deficit to the central visual pathways, most consistent with bilateral occipital cortical involvement. The presence of normal pupils, intact visual fields, a normal fundus aside from mild hypertensive changes, and preserved color vision effectively rules out significant optic nerve or retinal pathology. Additional findings impaired calculation suggesting left parieto-occipital dysfunction and constructional deficits indicating right parietal involvement further support a lesion affecting the bilateral parieto-occipital regions rather than the anterior visual pathways.

3. In young adults, posterior circulation ischemia leading to this clinical syndrome can arise from several vascular and systemic conditions, including inflammatory vasculitis (RA), vertebrobasilar arterial dissection, inherited or acquired prothrombotic states, and non-atherosclerotic arteriopathies such as Moyamoya vasculopathy.

4 Magnetic resonance imaging (MRI) of the brain with MR angiography (MRA) is the key investigation to narrow the differential diagnosis. MRI will identify parieto-occipital cortical or sub cortical lesions suggestive of PRES or other structural pathologies, while MRA helps detect underlying cerebral vasculopathy such as vessel narrowing, Moyamoya disease, vasculitis, or arterial dissection. Additionally, normalization of MRI findings on follow-up imaging at around three months would support a diagnosis of PRES.

Discussion and Conclusion

Moyamoya disease is a rare, chronic cerebrovascular disorder characterized by progressive stenosis and occlusion of the terminal intracranial internal carotid arteries and their proximal branches, with development of fragile compensatory collateral vessels that resemble a “puff of smoke” on angiography the hallmark radiographic feature of the condition. This process results in chronic cerebral hypoperfusion and predisposes affected individuals to ischemic or hemorrhagic strokes. (1) When similar angiographic features occur in association with an underlying systemic condition, the term moyamoya syndrome is preferred. Recent reviews emphasize that autoimmune and inflammatory disorders represent important secondary causes of moyamoya vasculopathy in young adults. (2)

In this patient, the presentation of sudden, profound, bilateral, painless visual loss with near-complete spontaneous recovery suggested a transient or evolving cerebral ischemic phenomenon rather than optic neuropathy or retinal pathology. Normal pupillary reactions, preserved color vision, and a normal fundus (aside from mild hypertensive changes) effectively rule out significant anterior visual pathway lesions (optic nerve, retina) pointing instead toward bilateral occipital cortical dysfunction, as later confirmed by imaging and cognitive testing (errors in figure drawing and calculation suggest parieto-occipital involvement). This pattern is typical of cortical blindness seen in posterior circulation or watershed ischemia due to compromised blood flow in the posterior cerebral territories or border zones.

The baseline MRI brain (Figure 2A) dated 21, June 2021 and demonstrated bilateral parieto-occipital infarcts. A repeat MRI performed five months later (Figure 2B) showed residual infarcts in the same distribution, and the accompanying MRA revealed bilateral internal carotid artery (ICA) occlusion with narrowing of both middle cerebral arteries (MCA). Subsequent digital subtraction angiography (DSA) confirmed ICA occlusion distal to the ophthalmic artery, with recruitment of collateral vessels supplying the MCA and ACA territories, consistent with Moyamoya disease, Suzuki angiographic Grade 3 (Figure 3). [1]

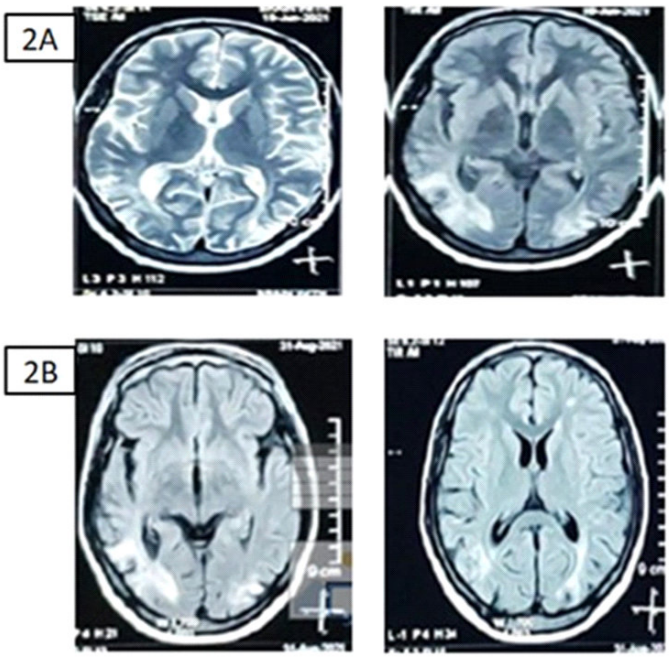

Figure 2A. Magnetic Resonance Imaging (MRI) brain showing bilateral parieto-occipital infarcts in T2 and flair sequences and **2B** shows bilateral parieto-occipital infarcts in flair sequences.

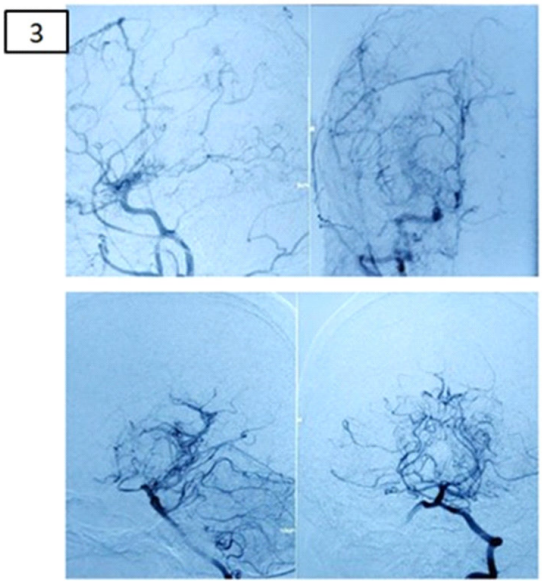

Figure 3. Digital Subtraction Angiography (DSA) ICA occlusion distal to the ophthalmic artery, with recruitment of collateral vessels supplying the MCA and ACA territories, consistent with Moyamoya disease, Suzuki angiographic Grade 3.

Extensive laboratory evaluation showed positive rheumatoid factor and anti-CCP antibodies, with negative CRP, ANA profile, ANCA, and antiphospholipid antibodies. Serum homocysteine and lipid parameters were within normal limits.

The clinical backdrop of rheumatoid arthritis introduces additional complexity: whereas idiopathic MMD is characterized by a lack of inflammation, moyamoya syndrome may be linked to systemic vascular disorders such as rheumatoid cerebral vasculitis, which can lead to ongoing changes in intracranial blood vessels and narrowing that ultimately result in a moyamoya configuration. Reports have described similar associations where autoimmune vascular injury predisposes to moyamoya-like occlusive disease. [3]

Thus, this case illustrates how a systemic immune-mediated vasculopathy can lead to chronic progressive steno-occlusive cerebrovascular disease, manifesting clinically as cortical blindness due to bilateral occipital infarcts in a young adult, a presentation that, without high suspicion and appropriate angiographic evaluation, might be misattributed to optic nerve or retinal pathology. MRI/MRA/DSA are critical in such scenarios, as they not only confirm the diagnosis but also guide further management, including revascularization strategies to prevent future ischemic events.

Conflict of Interest

The authors declare no conflict of interest.

Acknowledgement

None

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