Neurological Case Reports

Case Report
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Case of Idiopathic Central Retinal Artery and Vein Occlusion in a Pediatric Patient

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Abstract

Objective: We are presenting a case of painless progressive left eye visual defect in a healthy fourteen-year-old female.

Methods: This case is about an acute onset of left eye blurriness progressing into no light perception in the same eye, consistent with central retinal artery and vein occlusion in the further dedicated eye exam. Extensive workup to reveal the etiology of the clinical presentation was unremarkable.

Discussion: Based on the presentation, physical exam, and thorough workup findings, the patient was diagnosed with idiopathic central retinal artery and vein occlusion with no identifiable risk factors.

Conclusion: In a young population presenting with acute onset painless vision loss, a comprehensive workup to identify risk factors and possible causes is necessary, and close follow-up must be always present.

Keywords: CRAO; CRVO; Pediatric population; Concurrent CRVO; CRAO

Introduction

Graefe A et al. [1] described the first case of Central Retinal Artery Occlusion (CRAO) in his book in 1859, which was due to embolism. Thought to be a rare condition, with the mean age group for CRAO presentation ranging between 60 to 70 years [2]. Usually is caused by systemic diseases such as coagulation disorders, autoimmune conditions, etc. [3]; a few cases of CRAO without identifiable etiological factors also have been reported.

Case Presentation

We are presenting a case of a fourteen-year-old previously healthy young girl who presented with acute onset painless left eye cloudiness and blurriness for the last four days. Due to unresolved blurred vision, the patient was taken to the optometrist. At the optometrist office, a visual acuity test on the left eye was 20/400 and the right eye 20/30. The fundoscopic exam was remarkable for optic disc edema in the left eye. Based on the eye exam findings, neurology was consulted. Upon further interview with the neurology, the patient reported that eye symptoms started while awake and resting - initially, eye symptoms presented with flashing bright lights, which later developed into blurriness and cloudiness. Eye symptoms at onset were associated with nausea and itchiness, which resolved within a few hours, while visual defect persisted. She mentioned being in a camp two weeks prior presentation, where she stayed in the cabin dormitory. She denied any traumas, insect, tick, or animal bites, exposure to freshwater lakes or rivers. She denied other symptoms such as headache, fever, double vision, pain with eye movements, or any other notable changes in her health. Past medical history was remarkable only for mild allergic reactions, and she was using optic glasses for myopia. Family history was significant for a history of macular degeneration in maternal grandmother. She was living with her family in non-rural settings; no history of alcohol use, smoking, or pet exposures was reported. On further exam, her pupils were equally 5 mm, with prominent left eye Relative Afferent Pupillary Defect (RAPD). The rest of the neurological exam was unremarkable. The ophthalmologist's fundoscopic exam (Table 1, 2) was notable for grade II optic disc edema with diminished foveal reflex and with prominent macular swelling.

Based on the clinical presentation, neurological and ophthalmologic exam findings, the differential diagnosis included infectious or inflammatory papillitis, neuroretinitis, or demyelinating

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Table 1: Eye exam.

Base Eye Exam			
		Right	**Left**
Visual acuity	Near sc	J1	J3
Tonometry	Pressure	20	15
Pupils	Dark	5	5
	Light	2	4
	React	2	2
	APD	None	RAPD
Visual fields	Result	Full	Full
Extraocular movement	Result	Full	Full
Dilation	Both Eyes	1.0% Mydriacyl, 2.5% phenylephrine	
Additional Tests			
		Right	**Left**
Color	Ishihara	15/15	15/15

process. Thorough workup to identify potential causes included complete blood count with differential, erythrocyte sedimentation rate, C reactive protein, IgG/IgM *Bartonella henselae*, and Quinton species antibodies, IgG/IgM toxoplasma antibodies, treponema titers, serum angiotensin-converting enzyme levels, QuantiFERON gold test, Magnet Resonance Imaging (MRI) brain and orbits. The patient requested to complete MRI brain and orbit out-patiently due to the holiday season and was discharged from the Emergency Department (ED) on doxycycline 100 mg BID for possible neuroretinitis given optic disc edema and pigment epithelial detachment on an eye exam.

Five days after discharge, the patient presented to the hospital again with worsening visual acuity in the left eye, decreased color vision, interval worsening of optic disc edema on the fundoscopic exam with flame hemorrhages extending into the macula. The patient was not able to complete an outpatient MRI brain and orbits. Eye exam during the second presentation was remarkable for grade III optic disc edema with obstruction of vessels, diffuse flame hemorrhages, tortuous engorged veins in the left eye. In the face of worsening eye symptoms and eye exam findings, stat MRI brain/orbits with and without contrast, MRI of Cervical, Thoracic, and Lumbar (C/T/L) spines with and without contrast, further lab work including (i) infectious such as Herpes Simplex Virus (HSV), Varicella-Zoster Virus (VZV), *West Nile*, *Bartonella quintana*, and *henselae* Polymerase Chain Reaction (PCR); (ii) inflammatory and autoimmune workup such as IgG blood, Neuromyelitis Optica Aquaporin 4 (NMO AQP4), Myelin Oligodendrocyte Glycoprotein (MOG) antibodies), Antinuclear Antibodies (ANA), antiphospholipid panel, Antineutrophil Cytoplasmic Antibodies (ANCA) panel, rheumatoid factor were obtained. Lumbar puncture with thorough Cerebrospinal Fluid (CSF) analysis was completed. All infectious, inflammatory, and vasculitis workups, as well as MRI C/T/L spine, were unremarkable.

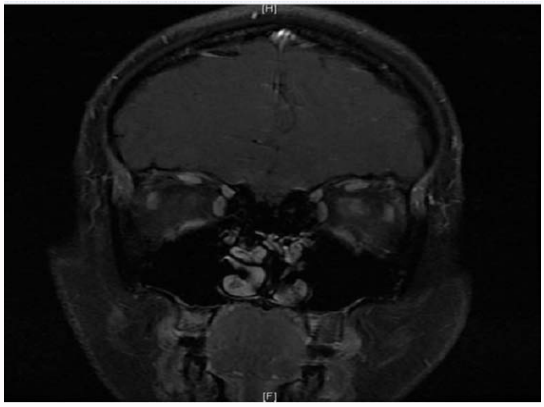

Figure 1: Coronal fat-suppression MRI of the orbits. T1 weighted sequence with subtle enhancement in the perineural soft tissues surrounding the left optic nerve represent left optic peri-neuritis.

MRI brain and orbits with and without contrast (Figure 1) showed a short segment of perineural enhancement of the retrobulbar optic nerve without enhancement of the central optic nerve.

Lumbar puncture results (Table 3) did not reveal any findings

Table 2: Slit lamp and Fundoscopic Exam.

Slit Lamp and Fundus Exam			
		Right	**Left**
External exam	External	Normal	Normal
Slit Lamp Exam	Lids/Lashes	Normal	Normal
	Conjunctiva/ Sclera	White and quiet	White and quiet
	Cornea	Mild decreased tear break up time	Clear
	Anterior Chamber	Deep and quiet	Deep and quiet
	Iris	Pharm dilated	Pharm dilated
	Lens	Clear	Clear
	Vitreous	Normal, no cell	Normal, no cell
Fundus Exam	Disc	Normal, distinct rims, pink, flat, no obscuration	Grade II disc edema, with no obscuration of vessels, and no hemorrhages
	C/D Ratio	0.25	0
	Macula	Normal, good foveal reflex	Diminished foveal reflex, foveal exudate, and associated swelling of the macula. In the region between the fovea and disc, approximately 1/2-disc diameters from disc, there is a fluffy, white, bilobled infiltrate.
	Vessels	Normal	Tortuous veins
	Periphery	Normal	Normal

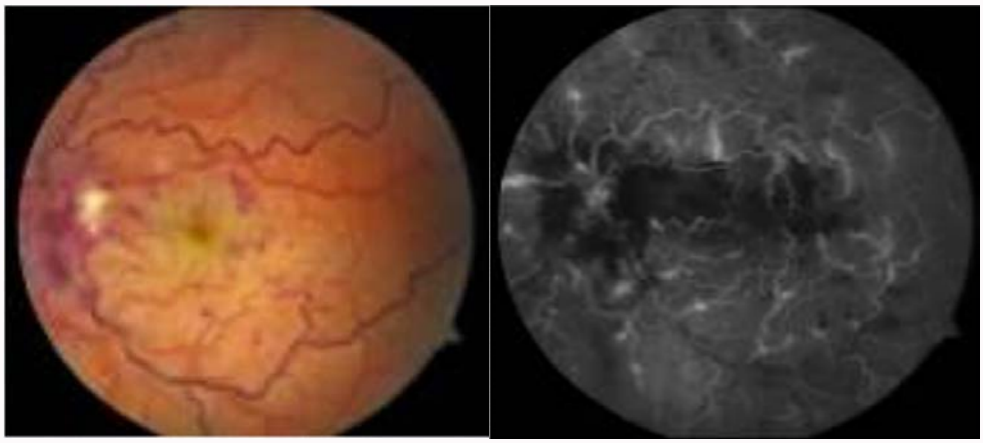

Figure 2: (a) Fundoscopic exam of the left eye: Significant disc edema-grade 2-3, 360 diffuse flame hemorrhages, tortuous veins, and scattered Dot and Blot Hemorrhage (DBH). (b) Fluoresce in Angiography (FA) of the left eye: Delayed Arterial Time (AT).

Table 3: CSF workup results.

CSF workup	Results
Color CSF	Colorless
Character CSF	Clear
Nucleated cells CSF	2
Total nucleated cell CSF	2
RBC CSF	2
Xanthochromia CSF	No
Neutrophils CSF	1
Lymphocytes % CSF	82
Monocytes % CSF	17
Cells counted CSF	72
Glucose, CSF-STAT	52
Oligoclonal bands	none
Protein CSF	28
Neuromyelitis Optica antibody titers	<1:1
Varicella zoster	0.00

consistent with infectious or demyelinating pathologies.

Left optic nerve sheath biopsy results did not show any acute or chronic inflammation or granulomas signs. On the further discussion of the case, the dermatology team suggested the possibility of Sneddon syndrome (thrombo-occlusive syndrome with livedoid cutaneous features), livedoid vasculopathy, and popular sarcoid. Skin biopsy was obtained with unremarkable results. With the exam and workup findings, the patient started on a three-day course of high-dose intravenous steroids and was discharged home on oral steroid taper for suspected perineuritis/papillitis leading to venous engorgement.

One day after discharge, the patient returned to the hospital with no light perception in the left eye. Fundoscopic exam (Figure 2a) and fluorescence angiography (Figure 2b) findings were notable for serous retinal discharge and very prolonged arterial filling time, arteriovenous transit time, and impaired perfusion of an entire retina consistent with central retinal vein and artery occlusion.

She was admitted to Neurology service for further stroke workup. Vascular imaging of the head and neck did not show any atherosclerotic plaque, stenosis, occlusion in the head/neck, or any signs of vasculitis. Hypercoagulable and cardiac workup was utterly unremarkable. Due to a completely unremarkable diagnostic workup, she was diagnosed with idiopathic central retinal vein and artery occlusion and started antiplatelet therapy with daily aspirin. On further follow up visual impairment was not resolved yet.

Discussion

The approach to acute vision loss is very well researched. While monocular vision loss localizes to the visual tract before the optic chiasm, binocular visual defects are consistent with pathology of the visual tract from the optic chiasm to calcarine fissure. Painful monocular vision loss is mostly characteristic of optic nerve inflammation/infection, while painless monocular visual defect raises suspicion for ischemic changes of the optic nerve [4]. CRAO is one of the rare conditions causing monocular painless visual defects with a mean age group of 60 to 70 years [2]. Causes of CRAO usually include systemic conditions as coagulation disorders, autoimmune conditions, etc. [3]. In recent years a few cases of CRAO without identifiable etiology factors were reported. The cases of an eleven-year-old female and thirteen-year-old male with idiopathic CRAO were reported by Ratra et al. [3] in 2012. Another case of idiopathic CRAO was reported by Heckler LV et al. [5] in 2008 in a healthy six-year-old male who presented with acute onset right eye vision loss, and all workup was unremarkable. In 2002 Lee WB et al. [6] described another case of an eight-year-old boy with left eye CRAO without any etiological cause that could be revealed on extensive workup. One of the recent cases was published by Liu et al. [7] describing acute onset painless visual loss in the right eye with ophthalmologic exam findings being consistent with CRAO with completely unremarkable workup.

All the above cases were notable for idiopathic CRAO in an otherwise healthy young population. In our case, a fourteen-year-old healthy girl was found to have central retinal vein and artery occlusion. Hayreh SS analyzed the cause of combined CRAO and CRVO. The author described that CRVO was related to occlusion in the vein in the optic nerve leading to blockage of retinal circulation and resulting in complete retinal circulation impairment and CRAO. In addition, it was described that in combined CRAO and CRVO, CRAO was not related to central retinal artery embolism or thrombosis but was a result of CRVO [8].

Idiopathic concurrent CRVO and CRAO are extremely rare,

especially in the pediatric population. A case report on this rare neurological condition was published by Azman MSB et al. [9] in 2017. The publishers described the case of a nine-year-old girl with sudden painless vision loss in her right eye. Fluorescence angiogram was notable for CRAO and CRVO. Further extensive workup did not reveal any identifiable etiology leading to a diagnosis of idiopathic CRAO and CRVO. She was treated with intravenous steroids with improvement in right eye vision to 1/60.

We are presenting another case of idiopathic concurrent CRAO and CRVO in an otherwise healthy fourteen-year-old female patient without any risk factors and completely unremarkable diagnostic workup findings.

Conclusion

Even if idiopathic CRAO or combined CRAO and CRVO are very rare in an otherwise healthy pediatric population, as published by the abovementioned authors and us, it is still quite possible to come across. In a young population presenting with acute onset painless vision loss, an extensive workup to identify risk factors and possible causes is necessary. Close follow-up must be present at all times.

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