CASE REPORT

Acute central retinal vein occlusion following episode of myopericarditis in an adolescent patient

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SUMMARY

A 15-year-old African-American boy presented with unilateral decrease in vision following an episode of myopericarditis. His ocular examination was significant for diffuse retinal oedema and intraretinal haemorrhages. He was diagnosed with a central retinal vein occlusion and was referred to rheumatology for further evaluation following negative hypercoagulable studies. He was later found to have systemic lupus erythematosus. Approximately 8 months after presentation, the patient's vision improved and remained stable at 20/40.

BACKGROUND

Central retinal vein occlusion (CRVO) is a major cause of significant visual impairment and blindness. Its 'blood and thunder' fundus appearance results from thrombosis causing disc swelling, diffuse retinal and preretinal haemorrhages, and cotton wool spots.¹ Presentation is usually subacute vision loss in contrast with sudden vision loss associated with central retinal artery occlusion. An afferent pupillary defect may be present if venous stasis is severe enough to cause slowed retinal perfusion. It affects men and women equally and is a disease predominantly of the elderly with major risk factors being age, hypertension, hyperlipidaemia, glaucoma and diabetes mellitus.²

CRVO is uncommon in younger adults and even rarer in the paediatric and adolescent population.¹ In these instances, one might consider unusual causes such as blood dyscrasias causing a hyperviscosity syndrome predisposing an individual. The exact cause of hyperviscosity can vary from rheumatological disease to lymphoma, or even oral contraceptive use in young women.³ A central retinal venous occlusion may be either ischaemic or non-ischaemic in nature, ischaemic having a worse prognosis.⁴

We present a rare case of unilateral non-ischaemic CRVO following an episode of myopericarditis in a 15-year-old African-American boy. To our knowledge he is the youngest patient reported with CRVO and also the only reported case associated with myopericarditis.

CASE PRESENTATION

A 15-year-old African-American boy presented to the emergency room with progressive right nasal hemianopia. The patient reported that the scotoma was noticed earlier that evening and slowly progressed leaving only a temporal sliver of clear vision in the right eye. He denied any recent trauma, pain, pain with eye movements, headache,

flashes, floaters or associated neurological symptoms.

The patient was discharged from the hospital 1 day prior after being treated for myopericarditis of unknown aetiology. At that admission, the patient presented with chest pain that increased when lying down. He was found to have diffuse ST segment elevation on ECG and progressively elevated troponins. Echocardiography revealed decreased left ventricular function and pericardial effusion. The patient was treated with intravenous immunoglobulin and discharged after clinical improvement. There were no reported ocular symptoms during that admission.

Visual symptoms began early in the evening on the day of presentation and initial ophthalmological examination showed best-corrected visual acuity of 20/70 with eccentric fixation in the right eye and 20/20 in the left eye. Colour plates were found to be 11/11 with eccentric fixation in the right eye and 11/11 in the left eye. Pupil examination showed a sluggish right pupil with a relative afferent pupillary defect, and a normal reactive pupil on the left. Intraocular pressures were 15 mm Hg in the right and 12 mm Hg in the left. On slit-lamp examination, the anterior segment was found to be normal bilaterally. Fundus examination of the right eye revealed disc oedema, disc haemorrhage, vessel tortuosity and macular whitening involving the fovea with normal periphery. Left eye fundus examination was within normal limits. Given the severity of ocular symptoms and clinical findings along with recent history of pericarditis and elevated sedimentation rate, the patient was admitted for a hypercoagulable and rheumatological workup (table 1) with presumptive diagnosis of optic neuropathy of unknown aetiology.

The following afternoon, the patient was found to have worsening retinal findings in the right eye with vascular sheathing and intraretinal haemorrhages in all four quadrants without evidence of vitritis; left eye remained normal. The patient was started on oral acyclovir and prednisone for presumptive diagnosis of acute retinal necrosis. The patient was discharged after 3 days with stable ocular findings and instructions for close outpatient follow-up.

On the first outpatient visit, optical coherence tomography (OCT) revealed massive subretinal fluid. In addition, fluorescein angiography showed disc oedema without vascular leakage (figure 1). Repeat testing of the right eye 1 week later showed improvement in vision with corresponding improvement of fundus appearance on dilated examination and

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Table 1 Results of diagnostic testing during admission and outpatient follow-up

	Results	Reference range
Infectious		
Adenovirus DNA, QN PCR	Not detected	<500 copies/mL
Parvovirus B19 PCR	Not detected	
Echovirus type 6 Ab	<1:10	<1:10
Echovirus type 7 Ab	<1:10	<1:10
<i>Echovirus type 9 Ab</i>	<1:160 (H)	<1:10
Echovirus type 11 Ab	<1:10	<1:10
Echovirus type 30 Ab	<1:10	<1:10
Rubella IgM	0.31	≤0.89 IU (negative)
<i>Coxsackie A-9 Ab</i>	1:16 (H)	<1:8
Enterovirus PCR	Not detected	
CMV PCR	Not detected	
HIV 1/2 Ab + P24Ag	Non-reactive	
RPR	Non-reactive	
Chlamydia/gonococcal	Negative	
Blood cultures	No growth	
Rheumatological		
Rheumatoid Factor	Negative	
<i>ANA titre</i>	1:640 (H)	<i>speckled pattern</i>
Cardiolipin IgG Ab	14.78	<23 (GPL)
Cardiolipin IgM Ab	2.5	<11 (MPL)
Serine protease 3	5	0–19 AU/mL (negative)
Myeloperoxidase Ab	3	0–19 AU/mL (negative)
Anti-Hu	Negative	
Anti-Ri	Negative	
Ribosomal P protein Ab	3	<29 AU/mL (negative)
<i>Anti-RNP</i>	441.11 (H)	<20 EU (negative); >25 EU (positive)
Scl-70 Ab	2.39	<20 EU (negative)
SS-B Ab	3.18	<20 EU (negative)
<i>SS-A Ab</i>	31.57 (H)	<20 EU (negative); >25 EU (positive)
Centromere Ab	26	<29 AU/mL (negative)
dsDNA Ab	11.2	<25 IU (negative)
Lyme Ab	negative	
ACE	29	18–101 U/L
Coagulation panel		
PT	13.8	12.2–14.5 s
INR	1.1	0.8–1.2
<i>PTT</i>	20.0 (L)	23.0–36.0 s
<i>Fibrinogen</i>	614(H)	200–480 mg/dL
<i>D-dimer</i>	>1600 ng/mL	<200ng/mL
CBC		
<i>Platelet count</i>	402(H)	100–400 × 10 ⁹ /L
<i>ESR</i>	124(H)	0–13 mm/h
<i>CRP</i>	3.7(H)	≤0.5 mg/dL
WBC	9.9	4.5–14.5 × 10 ⁹ /L
<i>RBC</i>	2.84 (L)	4.5–5.3 × 10 ¹² /L
<i>Hb</i>	7.9 (L)	13.0–16.0 g/dL
<i>Hct</i>	24.1 (L)	37.0–49.0%
MCV	84.9	78.0–98.0 fL
MCH	27.8	25.0–35.0 pg
MCHC	32.8	31.0–37.0 g/dL

ACE, angiotensin-converting enzyme; ANA, antinuclear antibody; CBC, complete blood count; CMV, cytomegalovirus; CRP, C reactive protein; dsDNA, double-stranded DNA; ESR, erythrocyte sedimentation rate; Hb, haemoglobin; Hct, haematocrit; INR, international normalised ratio; MCH, mean corpuscular hemoglobin; MCHC, mean corpuscular hemoglobin concentration; MCV, mean corpuscular volume; PT, prothrombin time; PTT, partial thromboplastin time; RBC, red blood cell; RNP, ribonucleoprotein; WBC, white blood cell.
Italics denotes abnormal value

Figure 1 Fluorescein angiography of the right eye, on initial outpatient follow-up visit, showing disc oedema without vascular leakage.

subretinal fluid on OCT. With continued improvement steroids were tapered over 1 month (figures 2 and 3). Five months after initial presentation, the patient showed complete resolution of fundoscopic findings with best-corrected visual acuity measuring 20/40 in the right eye. Last follow-up examination nearly 1 year later still showed best-corrected visual acuity of 20/40 and normal fundoscopic appearance in the right eye.

DISCUSSION

CRVO is primarily a disease of the elderly where risk factors of hypertension, diabetes and atherosclerosis are more prevalent. One major review suggested that 7.5–19.8% of CRVOs are found in patients under the age of 50, leading to the assumption

Figure 2 Serial optical coherence tomography (OCT) of the right eye. (A) Massive subretinal fluid on first outpatient visit with best-corrected visual acuity (BCVA) of 5/400. (B) Decrease in amount of subretinal fluid 1 week later with BCVA of 20/400. (C) Small residual fluid remained at 1 month follow-up with BCVA of 20/60. (D) OCT revealed resolution of subretinal fluid at 2-month follow-up with final BCVA of 20/40.

that the prevalence is likely lower in individuals under 20 years of age.⁵ It is believed that in young adults, CRVO is a different disease from retinal vein occlusion in adults.¹ It has been described with various terminology including benign retinal

vasculitis, optic disc vasculitis and papillophelbitis.⁶ One must be careful in adopting these terms to the adolescent population as there is limited literature and only a few anecdotal reports of varying aetiology.

Figure 3 Serial fundus photos of the right eye. (A) Significant retinal oedema with vascular engorgement and disc haemorrhage seen at first outpatient visit. (B) Improvement in retinal and disc oedema at 1-week follow-up. (C) Further improvement at 1-month follow-up with resolution of disc haemorrhage and trace residual retinal oedema. (D) Complete resolution of disc and retinal oedema with residual macular exudates at 2-month follow-up.

Known risk factors associated with development of CRVO in young adults (less than 40 years old) include glaucoma, sarcoidosis, vasculitis, increased orbital or intraocular pressure, hyphema, hyperviscosity syndrome of various aetiology, high homocysteine and HIV.⁶ Given the rarity of presentation, one might consider similar risk factors could potentially exist in the adolescent population as well.

There are several possible aetiologies to consider in our patient. The onset of symptoms shortly after being treated for myopericarditis could raise the possibility of venous stasis, from reduced ejection fraction, leading to venous backlog and the patient's CRVO. The aetiology of the patient's myopericarditis itself must also be considered as a possible cause of CRVO. Viral studies during the patient's episode of myopericarditis revealed elevated echovirus type 9 and mildly elevated cocksackie A-9 titres. Rheumatological workup initiated during admission was found to be negative along with HIV studies. Possible hyperviscosity following administration of intravenous immunoglobulin must also be considered given the proximity of symptoms to administration. However, only one dose, with a slow infusion rate, was given during admission and therefore unlikely to precipitate the event.⁷ Hypercoagulability panel showed elevated D-dimer, fibrinogen, platelets, ESR and CRP; however, these values could be elevated due to generalised inflammation.

Given our patient's age and presenting factors, we believed it to be reasonable to initiate a hypercoagulopathy and rheumatological workup. Coverage for acute retinal necrosis (ARN) with acyclovir was initiated due to the evolution of retinal findings and potential devastating outcome of ARN in such a young patient. Ongoing outpatient rheumatological studies revealed elevated anti-ribonucleoprotein and SS-A antibody titres leading to the presumptive diagnosis of systemic lupus erythematosus (SLE).⁸ Literature search revealed cases of CRVO in individuals with SLE, however, those cases were associated with positive lupus anticoagulant or antiphospholipid antibodies; which were found to be negative in our patient.^{9 10}

To our knowledge, this is the youngest patient reported with CRVO and associated myopericarditis. The diagnostic workup and treatment can be difficult in this population given the lack of known cases and published literature. In our investigation, no published algorithm exists for the diagnostic workup of an adolescent presenting with a CRVO. It has been shown that the specific cause of CRVO in young adults is not known.⁶ It is reasonable to assume that this may hold true in the adolescent population and more obscure causes must be investigated.

However, if associated systemic findings are present, as with myopericarditis in our case, it is reasonable to consider a multidisciplinary approach in evaluation to ensure occult systemic aetiologies are not missed.

Learning points

- ▶ Although extremely rare, central retinal vein occlusions (CRVOs) do occur in the adolescent population.
- ▶ Obscure aetiologies must be considered when evaluating an adolescent patient with CRVO.
- ▶ A multidisciplinary approach should be utilised in such cases to ensure occult systemic diseases are not missed.

Contributors AD and LA were involved in the conception, analysis and interpretation of the data and critical revision of the manuscript. SS contributed by analysing and interpreting the data and critically revising the manuscript.

Competing interests None.

Patient consent Obtained.

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