


Case report

Central retinal artery occlusion from *Streptococcus gallolyticus* endocarditisRita Serras-Pereira,¹ Diogo Hipolito-Fernandes ,¹ Luísa Azevedo,² Luísa Vieira¹

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SUMMARY

Central retinal artery occlusion (CRAO) is a rare but blinding disorder. We present a case of a 81-year-old woman with multiple cardiovascular comorbidities admitted to the emergency department due to sudden, painless vision loss on left eye (oculus sinister (OS)) on awakening. The patient also reported long standing fatigue associated with effort that started 4 months before admission. She presented best corrected visual acuity of counting fingers OS. Funduscopy OS revealed macular oedema with cherry red spot pattern. Blood cultures came positive for *Streptococcus gallolyticus* in the context of a bacteremia and native mitral valve vegetation identified on transoesophageal echocardiography. CRAO of embolic origin was admitted in the context of an infective endocarditis. CRAO can be the first manifestation of a potentially fatal systemic condition and thus multidisciplinary approach is warranted with close collaboration between ophthalmologists and internists in order to provide proper management and the best possible treatment.

BACKGROUND

Central retinal artery occlusion (CRAO) is a rare but blinding disorder characterised by an obstruction of retinal blood flow that may be due to an embolus, thrombus, vasculitis, traumatic vessel wall damage or spasm. The major risk factors can be divided in arteritic and non-arteritic in nature, the latter representing more than 90% of cases, and CRAO can be further classified as permanent, transient and with cilioretinal artery sparing, an artery present in 15%–30% of the population.^{1 2} Incidence of non-arteritic CRAO is approximately 1–2 in 100 000.³ Embolism from an atherosclerotic plaque is the most frequent cause and risk factors are similar to those for cardiovascular and cerebrovascular events: hypertension, diabetes mellitus, tobacco use, carotid and coronary artery disease.¹ In the European Assessment Group for Lysis in the Eye trial, 73% of patients with diagnosis of CRAO had hypertension, 40% had at least 70% of carotid stenosis, 22% had coronary artery disease, 20% atrial fibrillation and 17% valvular heart disease and in patients with no prior history of cardiovascular risk factors, at least one was identified in 78% of patients.³ CRAO usually presents with acute and painless loss of monocular vision with 80% of patients having a final visual acuity (VA) of counting fingers or worse.^{1 2} Classical fundoscopic findings include retinal oedema, cherry red

spot, retinal arteriolar attenuation and a normal optic disc.³ Occasionally, emboli may be seen in the central retinal artery (CRA) or its branches.³ Visual outcomes in CRAO are typically poor, although with some variability, depending on factors such as duration of CRAO, the presence of cilioretinal artery and aetiology of the emboli.³ Management of CRAO is independent from its cause and can be divided in acute, with attempts to restore ocular perfusion; subacute, with prevention of secondary ocular complications such as neovascularisation and glaucoma; and long term, with prevention of other end-organ ischaemic events.¹

Infective endocarditis (IE) is likewise a rare condition, potentially life-threatening, with mortality rates as high as 40% annually.⁴ Systemic embolisation can occur in up to 80% of patients with IE and the risk seems to be greater with mitral valve involvement.^{1 4 5} Valvular vegetations can release emboli and lead to amaurosis fugax or retinal artery occlusion.⁴ CRAO is a very rare complication of IE. *Streptococcus gallolyticus*, a gram-positive, non-enterococcal, group D *Streptococcus* from the *S. bovis* group, is an opportunistic pathogen asymptotically found in the gastrointestinal tract of humans and an increasing cause of bacteremia and IE in the elderly.^{5 6} It is estimated to be the aetiological agent of IE in 11%–14% of cases.⁵

To the best of our knowledge, this is the first reported case of CRAO due to *S. gallolyticus* IE and highlights the importance of multidisciplinary approach.

CASE PRESENTATION

A 81-year-old woman with prior history of valvular cardiomyopathy (moderate mitral and aortic valve stenosis), paroxysmal atrial fibrillation under oral anticoagulation, implanted pacemaker, type II diabetes mellitus, hypertension, dyslipidaemia, obesity and peripheral venous insufficiency was admitted to the emergency department of a tertiary care hospital due to sudden, painless loss of VA on the left eye (OS) on awakening. On ophthalmological examination, she presented best corrected VA (BCVA) of 20/25 right eye (oculus dextrus (OD)) and counting fingers OS. Pupils were isochoric and isoreactive, no restrictions on extraocular movements. On biomicroscopy, bilateral pseudophakia was identified. Funduscopy OS revealed macular oedema with cherry red spot pattern. The clinical diagnosis of CRAO OS was made and the patient was immediately treated with systemic



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Reminder of important clinical lesson

Table 1 Laboratory tests on admission and on discharge

	Laboratory tests on admission	Laboratory tests on discharge
Erythrocytes	3.03×10 ¹² /L↓	3.62 10 ¹² /L↓
Haemoglobin	8.5×10 g/L↓	10.4×10 g/L↓
Hematocrit	26.3%↓	32.5%↓
Mean cell volume	86.8 fl	89.8 fl
Mean cell haemoglobin	28.1 pg	28.7 pg
Red cell distribution width	13.4%	15.1%
Leucocytes	13.24×10 ⁹ /L↑	5.79×10 ⁹ /L
Neutrophils	11.25×10 ⁹ /L↑	3.75×10 ⁹ /L
Eosinophils	0.2×10 ⁹ /L	0.12×10 ⁹ /L
Basophils	0.4×10 ⁹ /L	0.9×10 ⁹ /L
Lymphocytes	0.97×10 ⁹ /L↑	1.57×10 ⁹ /L
Monocytes	0.96×10 ⁹ /L	0.30×10 ⁹ /L
Platelets	249×10 ⁹ /L	228×10 ⁹ /L
C-reactive protein	152.5 mg/L↑	0.9 mg/L
Prothrombin time	25.7 seg↑	14.1 seg
Activated partial prothrombin time	31.5 seg↑	35.1 seg
Urea	90 mg/dL↑	52 mg/dL↑
Creatinine	1.62 mg/dL↑	1.18 mg/dL↑
Estimated glomerular filtration rate	30 mL/min/1.73↓	44 mL/min/1.73↓
Lactic dehydrogenase	339 U/L↑	227 U/L↑
Sodium	134 mEq/L	142 mEq/L
Potassium	4.6 mEq/L	4.7 mEq/L
Chloride	107 mEq/L	106 mEq/L
Calcium	8.1 mg/dL	9.5 mg/dL
Magnesium	1.89 mg/dL	1.82 mg/dL

acetazolamide and isosorbide dinitrate, ocular massage and topical ocular hypotensives.

On further questioning, the patient also reported long standing fatigue associated with effort that started 4 months before admission and worsened the previous week, when she started feeling febrile. On physical examination, the patient was alert, attentive and oriented; febrile (temperature 38°C); haemodynamically stable; oxygen saturation of 97% (at room air) and

on auscultation, cardiac sounds were rhythmic and a systolic murmur was evident.

On complementary examination, blood tests revealed normocytic normochromic anaemia and increased inflammatory markers (table 1). ECG showed ventricular pacing rhythm and chest radiography showed an increased cardiothoracic ratio. Cranial CT did not reveal any acute parenchymatous brain lesions.

The patient was admitted to the internal medicine department for further aetiologic investigation. A few days after admission, blood cultures came positive for *S. gallolyticus spp gallolyticus* sensitive to penicillin and a native mitral valve vegetation was identified on transoesophageal echocardiography. CRAO of embolic origin was admitted in the context of an IE. Optical coherence tomography confirmed an ischaemic and oedematous internal retina compatible with CRAO diagnosis (figure 1)

TREATMENT

The patient was treated with intravenous ceftriaxone 2 g/day for 28 days and was discharged after full clinical stabilisation, 33 days after admission.

OUTCOME AND FOLLOW-UP

The ophthalmology department collaborated closely with the internal medicine department. The patient refused to be submitted to any further ophthalmologic examinations or treatments but maintained periodic follow-up and was stable since discharge. At the time of the last visit, BCVA OS was counting fingers, anterior segment without significant changes, intraocular pressure (IOP) within normal limits and retinal atrophy with optic disc pallor on funduscopy, without signs of iris, optic disc or retinal neovascularisation.

DISCUSSION

CRAO due to IE is a rare event with few cases reported in the literature. The authors report the first case of CRAO due to *S. gallolyticus* endocarditis. CRAO is an ophthalmologic emergency and can be the first manifestation of a potentially fatal systemic condition, such as IE, with overlapping risk factors for major cardiovascular and cerebrovascular events.³

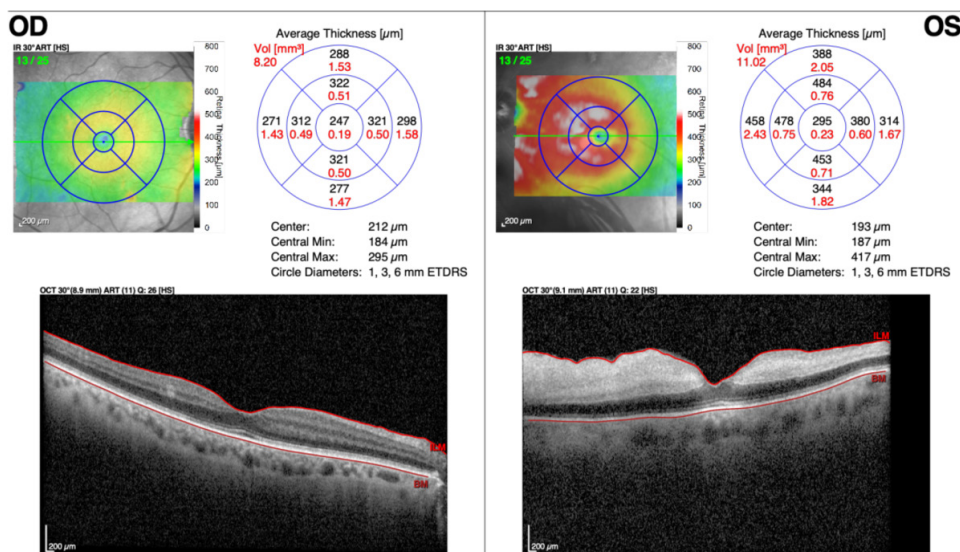


Figure 1 Optical coherence tomography imaging showing hyperreflective and thickened inner retinal layers that correspond to retinal oedema left eye. Oculus dextrus, OD; oculus sinister, OS.

Any process that leads to the obstruction of blood flow to the CRA can be a cause of CRAO.³ Embolism from an atherosclerotic plaque is the leading cause of CRAO, most frequently in the internal carotid artery followed by the aortic arch and heart.^{1,3} Less commonly, CRAO can be caused by a thrombotic event in the setting of vasculitis, especially giant cell arteritis, or hypercoagulable state such as hyperhomocysteinaemia, factor V Leiden, protein C and S deficiencies, sickle cell disease, paraneoplastic syndromes, among others.^{1-3,7} Certain ocular conditions can also lead to CRAO by decreasing ocular perfusion across the optic nerve head: acutely raised IOP, optic nerve head drusen and preretinal arterial loops.³ These less frequent causes should always be considered in patients without classic cardiovascular risk factors. As far as we know, CRAO due to an embolus from a cardiac vegetation is a very rare complication of IE with no estimated prevalence in the current literature.

IE should be suspected in patients with risk factors for the disease and an acute or subacute illness.⁸ Damaged endothelium in the setting of valvular stenosis can lead to the formation of a platelet and fibrin thrombus with risk of bacterial colonisation, which in turn may lead to further endothelial injury and culminate in the formation of an infected vegetation.⁹ Eighty to ninety per cent of cases involve gram-positive cocci, with *S. aureus* identified as the most frequent causative pathogen in developed nations and *S. viridans* group in developing nations.⁹ IE has a wide clinical presentation, often non-specific, and embolic events may prompt patients to seek medical attention in the first place.

The documentation by an ophthalmologist of an embolic event may be central to diagnostic guidance in the emergency room, as this case highlights. An IE should always be suspected in a patient with a heart murmur and documented cardiac pathology, particularly with positive inflammation markers and bacteremia. In the setting of degenerative valvular stenosis, non-specific presentation of long standing fatigue and an embolic event (including CRAO), a diagnostic hypothesis of IE should not be dismissed even if concurrent cardiovascular risk factors are present. In the present case report, a medical history and general examination pointed towards a septic embolus due to IE even in the presence of multiple cardiovascular risk factors.

IE is a potentially life-threatening condition with high mortality rates and treatment should be started as soon as the diagnosis is made. Likewise, there is a high degree of morbidity and mortality associated with CRAO, not only due to severe vision compromise and decreased quality of life, but also due to immediate and long-term ocular and systemic events.² There is an increased risk of stroke, acute myocardial infarction and death in patients with CRAO admitted to hospitals as CRAO represents end-organ ischaemia and potentially underlying atherosclerotic disease that puts patients at risk of future ischaemic events.^{1,2} Lavin *et al* reported an incidence of 36.7% of critical carotid artery disease and 20% of major echocardiographic abnormalities, such as myocardial infarction, heart failure and critical valvular disease, in CRAO patients.¹⁰

A multidisciplinary approach is warranted with close collaboration between ophthalmologists and other specialists in order to provide proper management and the best possible treatment to these patients.

Learning points

- ▶ Central retinal artery occlusion (CRAO) can be the first manifestation of a potentially fatal systemic condition, such as IE, with overlapping risk factors for major cardiovascular and cerebrovascular events.
- ▶ CRAO is a very rare complication in the setting of an infective endocarditis (IE).
- ▶ IE should be suspected in patients with risk factors for the disease and an acute or subacute illness.
- ▶ The documentation by an ophthalmologist of an embolic event may be central to diagnostic guidance in the emergency room.
- ▶ Close collaboration between specialists from different areas is fundamental in order to provide optimal management to our patients.

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