Case Report

A Cardiac Pseudoaneurysm as a Thromboembolic Source: Acute Visual Loss due to Cardiac Emboli

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Received 10 December 2019; Accepted 2 March 2020; Published 11 March 2020

Acute visual loss is rarely caused by a heart condition. This manuscript transcribes a case report of a 36-year-old patient with a 2-year history of aortic valve replacement due to bicuspid aortic valve endocarditis that presents to the emergency department with an acute right eye visual loss. After ophthalmologic investigation identified a central retinal artery occlusion, a transthoracic echocardiography was performed to search for a possible cardiac embolus, despite the patient presenting INR values of 2-2.5 for the last year. A mitral-aortic intervalvular fibrosa pseudoaneurysm was identified. A transoesophageal echocardiography was then performed, identifying a small clot logged inside the pseudoaneurysm that protruded to the left ventricle outflow tract.

After INR-adjusted warfarin treatment to levels between 3 and 4, the pseudoaneurysm was surgically closed. This is a rare case since the likely source of embolism to the central retinal artery was the thrombus logged inside the pseudoaneurysm despite a standardly accepted therapeutic INR.

1. Introduction

The mitral-aortic fibrosa (MAF) consists of a thin layer of fibrous avascular tissue that separates the anterior mitral leaflet from the posterior portion of the aortic root [1]. It is limited by the left atrium (posterolaterally), the left ventricular outflow tract (LVOT, inferiorly), and the pericardium (superiorly). It is prone to infection, and, as a result, a pseudoaneurysm (PMAF) may develop—an uncommon condition that typically consists in a pulsatile cavity in the mitral-aortic junction communicating with the left ventricular outflow tract (LVOT) [1–3]. It is most frequently caused by aortic valve endocarditis, particularly in prosthetic valves [1, 4]. It may be identified by transthoracic echocardiography (TTE) and transoesophageal echocardiography (TOE) in postcardiac surgery asymptomatic patients or may present itself with many unspecific symptoms: the current literature describes fever and active signs of endocarditis in nearly 40% of PMAF patients and heart failure signs in 16%. Only 12% presents with cerebrovascular or embolic events [2]. It may be a severe condition if complete rupture occurs [5], but treatment is available through surgical or percutaneous closure [2, 4, 6].

2. Case Presentation

2.1. History of Presentation. A 36-year-old male presented to the emergency department with complaints of acute (approximately 1-hour evolution) and painless visual loss of the right eye. He was promptly observed by ophthalmologic investigation identified a central retinal artery occlusion, a transthoracic echocardiography was performed to search for a possible cardiac embolus, despite the patient presenting INR values of 2-2.5 for the last year. A mitral-aortic intervalvular fibrosa pseudoaneurysm was identified. A transoesophageal echocardiography was then performed, identifying a small clot logged inside the pseudoaneurysm that protruded to the left ventricle outflow tract.

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2.2. Past Medical History. Previous medical history included a known bicuspid aortic valve disease. 2 years later, and after a dental procedure, the patient developed an aortic valve endocarditis due to an infection from *Streptococcus sanguinis*. This led to an ischemic stroke, from which the patient recovered completely. He was under antibiogram-directed antibiotic therapy for 1 month when a second stroke happened, and he was then submitted to an urgent surgical replacement of the aortic valve for a mechanical prosthesis that went well. He was discharged with a transthoracic echo referring a normofunctional prosthesis with no other significant alterations and an indication for warfarin therapy for INR target between 2 and 3. At the time of presentation of this case, the patient had medical records available that confirmed INR values between 2 and 2.5 for the last year.

2.3. Differential Diagnosis. Differential diagnosis for single eye acute visual loss includes an acute stroke/transient ischemic accident, compressive lesion, trauma, infection, Purtscher retinopathy and occlusion of the central retinal artery due to either atherosclerotic disease, embolism, arteritis or the presence of a hypercoagulable state (due to neoplastic disease or inherited pro-coagulative diseases, for example) [7].

2.4. Investigations. A fundoscopic exam was immediately performed and identified the foveola assuming a cherry red spot-like image with retinal whitening, which suggested that the visual loss had been caused by a central retinal artery occlusion (Figure 1(a)). Due to the patient’s previous medical history, cardiology observation was required to search for a cardiac source of embolism. The patient’s physical exam was normal, with cardiac auscultation identifying a clear metallic click as the second cardiac sound and a slight nonradiating systolic murmur (I/VI). A TTE was performed, showing a nonobstructive aortic prosthesis and a periprosthetic echolucent pulsatile region with a hyperechogenic mobile structure inside (Figures 1(b) and 2). To better characterize these findings, a TOE was performed (Figures 1(c), 1(d), 3, and 4) revealing a free hyperechogenic highly mobile structure suggestive of a free thrombus inside a pseudoaneurysm in the mitral-aortic intervalvular fibrosa, protruding into the LVOT through a small orifice (supplementary material video 1 and 2).
tract; Ao: ascending aorta.

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3. Discussion

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complication with a very rare clinical presentation. In our

understanding, the PMAF created a low-flow condition that,

despite an INR between 2 and 2.5, still allowed a thrombus

formation. The embolization of this small thrombus was

the highly probable cause for the central retinal artery occlu-

sion that caused the patient’s symptoms. Regarding ophthal-

mological investigation, and after fundoscopy identification

of retinal whitening with a macular cherry red spot, no fur-

ther tests are indicated [7].

A review of all relevant English language articles pub-

lished until 2010 identified 88 PMAF cases in the literature:

only 11 patients presented with a cerebrovascular or an

embolic complication. Of these, a clot was identified inside

the pseudoaneurysm in only 5 cases [2]. Stroke due to a

thrombus inside the PMAF was previously described, but to

our knowledge, this is the first described case of central reti-

nal artery occlusion.

Acute visual loss is rarely due to a cardiac cause, but in

patients with high embolic risk conditions, it should be sys-

tematically thought of, since diagnosis may be difficult [5]

but specific adequate treatment is available [6]. In this case,

the final diagnosis was a cardioembolic central retinal artery

occlusion, and most patients continue to experience severe

vision loss during follow-up despite the current standard

therapy [8].

This case highlights that in patients with a high embolic

risk, the search for a cardiac embolism after any possible

embolic event must be thorough. According to guidelines

and recommendations, TTE is the first tool, and sometimes

a TOE is also required [3]. This report represents a clinical

scenario where a rare extracardiac manifestation (visual loss)

was the event that prompted an exhaustive investigation that

allowed surgical correction for the underlying problem.

Conflicts of Interest

There are no conflicts of interest.

Supplementary Materials

Supplementary 1. Video 1: transoesophageal echocardiogra-

phy at 126° showing the mitral-aortic intervalvular fibrosa

with a pseudoaneurysm (arrow) communicating with the left

ventricle outflow tract and a free thrombus inside.

Supplementary 2. Video 2: transoesophageal echocardiogra-

phy with 3D reconstruction showing a highly mobile mass

suggestive of a small thrombus.

2.5. Treatment. In the acute setting in the emergency depart-

ment, treatment consisted of the intermittent digital massage

of the affected eye and the use of ocular hypotensive drugs

and intravenous acetazolamide. The patient had a partial

recovery of right eye visual acuity to 6/12. After 3 months

of INR-adjusted warfarin treatment for target 3.0–3.5, the

patient was submitted to a surgical pseudoaneurysm closure

with a synthetic patch.

2.6. Outcomes. Since surgical pseudoaneurysm closure, and

after the 1-year follow-up, the patient had no other embolic

event. Visual acuity had no further improvements.

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Supplementary 1

Supplementary Materials

See Supplementary 1

See Supplementary 2

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See Supplementary 1
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