Title
Calcified amorphous tumor: A rare cause of central retinal artery occlusion

Authors
Ma, Jeffrey H
Gill, Manjot K

Publication Date
2018-06-01

DOI
10.1016/j.ajoc.2018.01.038

Peer reviewed

American Journal of Ophthalmology Case Reports 10 (2018) 25–27

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: www.elsevier.com/locate/ajoc

Case report

Calcified amorphous tumor: A rare cause of central retinal artery occlusion

Jeffrey H. Ma, Manjot K. Gill*

Department of Ophthalmology, Feinberg School of Medicine, Northwestern University, Chicago, IL, USA

ARTICLE INFO

Keywords:
Central retinal artery occlusion
Calcified amorphous tumor
Mitral annular calcification

ABSTRACT

Purpose: We report the case of a central retinal artery occlusion secondary to presumed embolus from a calcified amorphous tumor of the heart, a very rare non-neoplastic cardiac mass.

Observations: A 60-year-old female presented with acute unilateral vision loss of the left eye. Examination revealed hand motion visual acuity of the left eye and a left relative afferent pupillary defect. Fundoscopy showed whitening of the macula with a cherry red spot, consistent with a central retinal artery occlusion. Initial workup was unremarkable, including hypercoagulability labs, magnetic resonance imaging of the brain, and magnetic resonance angiography of the head and neck. Transthoracic echocardiogram (TTE) showed calcification of the mitral valve but no masses. Subsequently, transesophageal echocardiogram (TEE) was performed, which revealed a mobile calcified amorphous tumor of the heart.

Conclusions: Calcified amorphous tumor of the heart is a very rare cardiac mass that may cause retinal artery occlusion. TEE is a more sensitive imaging modality to assess for potential cardio-embolic sources if TTE is unrevealing.

1. Introduction

While cardio-embolic sources are the second most common cause of retinal artery occlusions after carotid artery disease, calcified amorphous tumor (CAT) of the heart is a very rare cardiac mass with only 2 reports in the literature of its association with retinal artery occlusion.[1,2] We report a case of central retinal artery occlusion due to presumed calcific embolus from a CAT which was initially missed on transthoracic echocardiogram (TTE) and subsequently detected on transesophageal echocardiogram (TEE).

2. Case report

A 60-year-old female presented as a consult to the retina clinic with a history of sudden vision loss of the left eye, which had occurred 2 months prior. She had initially presented to a local hospital where she was diagnosed with a left central retinal artery occlusion (CRAO) that was acutely managed with ocular massage and anterior chamber paracentesis. The patient then transferred care to our hospital. She had a past medical history of ovarian cancer in remission, type 2 diabetes with mild non-proliferative diabetic retinopathy, and morbid obesity. On initial presentation, her best-corrected visual acuity was hand motion in the left eye. Fundus examination of the left eye showed a pale retina with a cherry red spot in the fovea, as well as occlusions of the retinal arteries, consistent with a CRAO. Laboratory tests, including complete blood count, hypercoagulability studies (anticardiolipin antibody, lupus anticoagulant, antithrombin III activity, protein C activity, protein S activity, homocysteine), erythrocyte sedimentation rate, and C-reactive protein, were within normal limits. Magnetic resonance imaging of the brain showed no acute infarcts, and magnetic resonance angiography of the head and neck showed no occlusions or significant stenosis of the carotid or vertebral circulations. TTE with bubble study showed mitral annular calcification but no masses. Subsequently, TEE was performed, which revealed severe caseous mitral annular calcification of the posterior mitral valve, as well as a mobile 0.9 × 1.0 cm calcified endocavitary mass of the mitral valve that was diagnosed as a calcified amorphous tumor of the heart and the presumed source of her CRAO (Fig. 1). The patient was evaluated by the cardiac surgery service whereupon she was deemed a poor surgical candidate for cardiac exploration and mass resection given her morbid obesity. She was started on aspirin and warfarin for anticoagulation, with reconsideration for possible surgery if she were to develop recurrent thromboembolic events or valvular compromise.

On her presentation to the retina service 2 months later, her visual acuity in the left eye remained hand motion, with a left relative afferent pupillary defect. Her fundus exam was significant for perifoveal whitening, arteriolar attenuation, and pallor of the optic nerve with fine neovascularization of the disc (Fig. 2A and B). Fluorescein angiography revealed an enlarged foveal avascular zone with disc leakage and peripheral vascular staining (Fig. 3A and B). She was treated with

* Corresponding author. Department of Ophthalmology, Feinberg School of Medicine, Northwestern University, 645 North Michigan Avenue, Suite 440, Chicago, IL, 60611, USA.
E-mail address: m-gill3@northwestern.edu (M.K. Gill).

https://doi.org/10.1016/j.ajoc.2018.01.038
Received 24 April 2017; Received in revised form 14 January 2018; Accepted 19 January 2018
Available online 31 January 2018

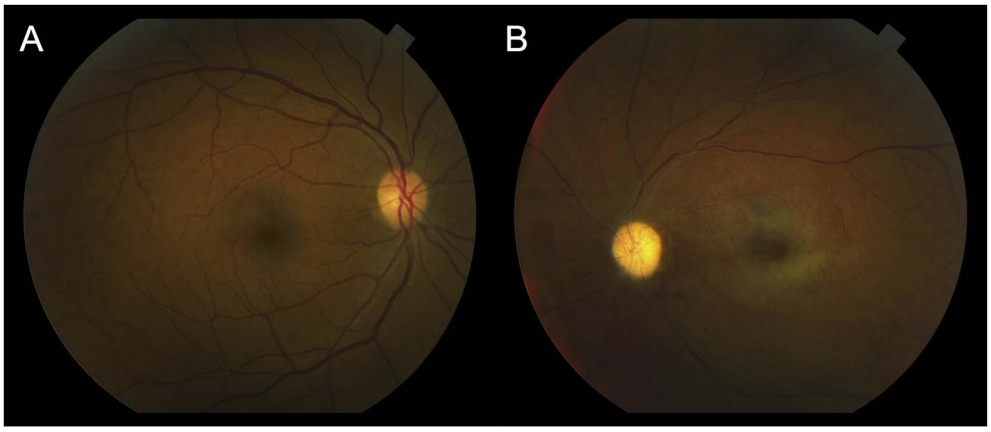

Fig. 1. Fundus photos of the right (A) and left (B) eyes reveals perifoveal whitening, disc pallor and vascular attenuation of the left eye 2 months after initial central retinal artery occlusion.

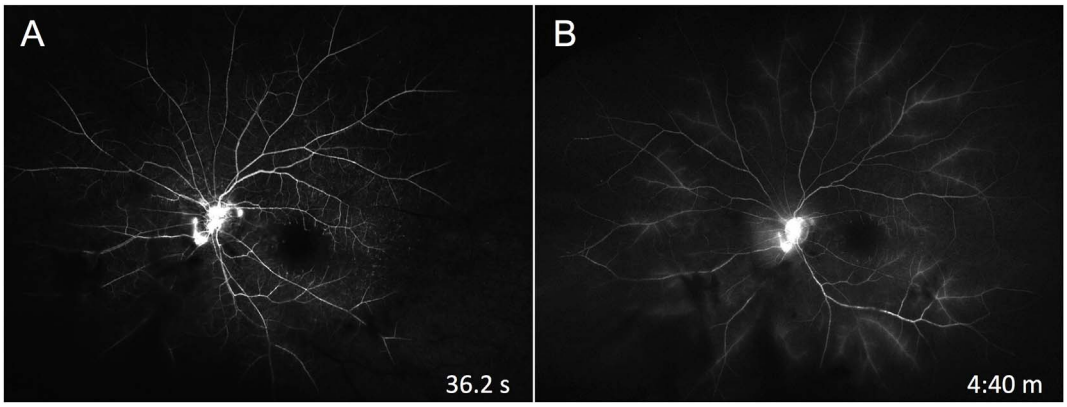

Fig. 2. Fluorescein angiogram of the left eye shows early hyperfluorescence of the disc and an enlarged foveal avascular zone (A), with disc leakage and peripheral vascular staining in the late phase (B).

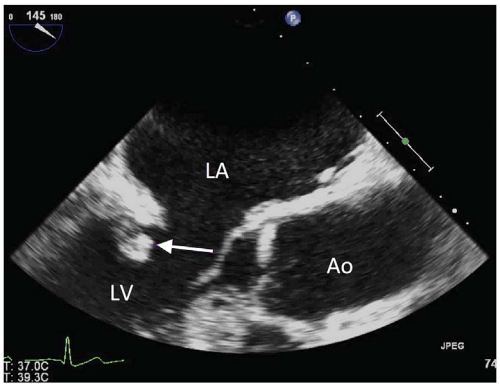

Fig. 3. Transesophageal echocardiogram shows calcification of the posterior mitral valve annulus with an echodense mass (arrow) attached to the mitral valve consistent with calcified amorphous tumor of the heart. LA = left atrium; LV = left ventricle; Ao = aorta.

panretinal photocoagulation of the left eye with regression of neovascularization. She has had no other complications related to her cardiac tumor since then.

3. Discussion

First described in 1997 by Reynolds et al.,[3] calcified amorphous tumor is a very rare non-neoplastic cardiac mass formed from a degenerating mural thrombus. While the pathogenesis of CAT is poorly understood, histological studies show nodular calcium deposits surrounded by amorphous degenerated fibrin, hyaline and inflammatory cells.[3–5] In a review of all 42 reported cases of CAT from 1997 to 2015, 14% were associated with mitral annular calcification, which is chronic degeneration and calcification of the mitral valve affecting about 10% of the population.[6] CAT may represent the end result of caseous transformation and liquefaction of mitral annular calcification. Patients typically present with dyspnea, syncope and pulmonary embolism. To our knowledge, there have been only 2 previous reports of retinal artery embolism resulting from cardiac CAT.[1,2] We report a case of CRAO due to presumed embolus from a cardiac CAT that was initially missed on TTE, though the possibility of an arteriosclerotic thrombus causing her CRAO cannot be entirely ruled out. Surgical resection is considered the definitive treatment of a cardiac CAT although observation with anticoagulation therapy is sometimes elected if calcium infiltration is extensive or surgical morbidity is high.

Our current case demonstrates the utility of TEE in evaluating for potential cardio-embolic sources that may be missed by TTE. In our patient, TTE demonstrated only mitral annular calcification, while cardiac CAT was only detected on subsequent TEE. Prior studies have reported the higher yield of TEE over TTE in detecting cardiac or aortic abnormalities in patients with retinal artery occlusion.[7–9] In a study by Kramer et al., 11 of 18 patients (61%) with retinal artery occlusion had at least one potential cardiac or aortic source of retinal emboli found on TEE that was missed by TTE.[10] Thus, TEE is a useful imaging modality that should be considered especially in a patient with retinal artery occlusion with an otherwise unrevealing workup for emboli.

4. Conclusions and importance

Calcified amorphous tumor of the heart is a very rare cause of retinal artery occlusion. This case highlights the utility of transesophageal echocardiography as part of a thorough workup for detecting cardiac

sources of retinal emboli, particularly when transthoracic echocardiography is unrevealing.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient.

Acknowledgements and disclosures

Funding

No funding or grant support.

Conflicts of interest

The following authors have no financial disclosures: JM, MG.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Acknowledgements

None.

References

1. Vlasseros I, Katsi V, Tousoulis D, et al. Visual loss due to cardiac calcified amorphous tumor: a case report and brief review of the literature. *Int J Cardiol.* 2011;152:e56–57.
2. Nazli Y, Colak N, Atar IA, et al. Sudden unilateral vision loss arising from calcified amorphous tumor of the left ventricle. *Tex Heart Inst J.* 2013;40:453–458.
3. Reynolds C, Tazelaar HD, Edwards WD. Calcified amorphous tumor of the heart (cardiac CAT). *Hum Pathol.* 1997;28:601–606.
4. Kubota H, Fujioka Y, Yoshino H, et al. Cardiac swinging calcified amorphous tumors in end-stage renal failure patients. *Ann Thorac Surg.* 2010;90:1692–1694.
5. Vaideeswar P, Karunamurthy A, Patwardhan AM, Hira P, Raut AR. Cardiac calcified amorphous tumor. *J Card Surg.* 2010;25:32–35.
6. de Hemptinne Q, de Canniere D, Vandenbossche J-L, Unger P. Cardiac calcified amorphous tumor: a systematic review of the literature. *Int J Cardiol Heart Vasc.* 2015;7:5.
7. Wisotsky BJ, Engel HM. Transesophageal echocardiography in the diagnosis of branch retinal artery obstruction. *Am J Ophthalmol.* 1993;115:653–656.
8. Wiznia RA, Pearson WN. Use of transesophageal echocardiography for detection of a likely source of embolization to the central retinal artery. *Am J Ophthalmol.* 1991;111:104–105.
9. Inatomi Y, Hino H, Hashimoto Y, Furuyoshi N, Misumi I, Uchino M. Transesophageal echocardiography for detection of cardiac diseases in patients with retinal artery occlusion. *Intern Med.* 2001;40:475–478.
10. Kramer M, Goldenberg-Cohen N, Shapira Y, et al. Role of transesophageal echocardiography in the evaluation of patients with retinal artery occlusion. *Ophthalmology.* 2001;108:1461–1464.