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Case report

Amaurosis fugax due to pleomorphic sarcoma in the left atrium

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ABSTRACT

Purpose: This report describes a case of amaurosis fugax due to a rare primary cardiac sarcoma.**Observations:** A patient who was recently diagnosed with left atrial pleomorphic sarcoma presented with a chief complaint of multiple episodes of intermittent vision loss in the right eye during the course of radiation therapy.**Conclusions and importance:** The authors postulate emboli from the left atrial sarcoma entered systemic circulation and subsequently caused brief episodes of transient occlusion to retinal, ophthalmic and/or ciliary arteries leading to momentary retinal hypoxia. We believe this is a novel finding, previously unreported in the literature, of transient embolic occlusion without permanent visual sequelae due to a malignant primary cardiac pleomorphic sarcoma.© 2016 The Authors. Published by Elsevier Inc. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Primary cardiac sarcoma is a rare malignancy with a poor prognosis.¹ Due to this rarity, a case of amaurosis fugax caused by cardiac sarcoma is unusual.

2. Case report

A 49 year old male with a past medical history of Hodgkin's lymphoma at age 20, hypothyroidism, and recently diagnosed cardiac sarcoma of the left atrium, presented to our office with a four month history of visual changes in the right eye. Previously, the patient received treatment for Hodgkin's lymphoma, first with chemotherapy with carmustine, cyclophosphamide, vinblastine, prednisone, and procarbazine. Following an incomplete response in his neck, the patient received mantle field and periaortic radiotherapy for a total dose of 40 Gy.

Regarding ophthalmologic symptoms, he described a "curtain going over the right eye on and off" resulting in a full graying out of vision; these episodes occurred occasionally, and occurred for a duration of four months. This "graying out" would last

approximately two minutes in the right eye, then slowly fade away back to normal vision. Closure of the right eye during the episode confirmed that the episode was only in the right eye and not a right homonymous hemianopic field defect. He reported the episodes had been increasing in frequency. He was evaluated by a community ophthalmologist at initial onset of symptoms. At that time, no retinal abnormalities were identified. The only additional symptoms included shortness of breath and intermittent left sided chest pain; no further symptoms such as claudication were identified. He was diagnosed with retinal migraines.

After experiencing visual symptoms for four months and noticing increasing ophthalmologic symptoms, he was found to have a left atrial sarcoma. Cardiac MRI with and without contrast revealed an enhancing mass in the posterior left atrium with extension into the inferior left pulmonary vein without involvement of the mitral valve. The lesion had intermediate signal intensity which measured 4.8 cm in maximum transverse dimension and 2.1 cm in AP dimension. An MRI brain obtained to further evaluate for intracranial metastasis showed no evidence of acute intracranial pathology or metastatic disease.

A trans-septal intracardiac biopsy was performed and surgical pathology revealed atypical spindle cell proliferation, consistent with undifferentiated pleomorphic sarcoma. A transthoracic echocardiogram completed at an outside institution revealed sarcoma involving the left atrial wall with chronic obstruction of the left inferior pulmonary vein, with narrowing of the right inferior

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pulmonary vein, and extension to the left superior pulmonary vein without obstruction. Functionally, the sarcoma did not involve the mitral valve.

The rest of the findings were otherwise within normal limits. PET/CT scan from skull base to mid thorax further staged the sarcoma. Metabolic evidence of malignancy correlated to the left atrial sarcoma with regional subcarinal lymph node involvement. There was no metabolic evidence of distant metastasis or lymphoma. The non operable cardiac sarcoma was presumed to be the result of previous radiation treatment for Hodgkin's lymphoma.

Following the initiation of daily cardiac radiation for the pleomorphic atrial sarcoma, the episodes of amaurosis fugax became a daily occurrence. At that time, he was referred to our academic ophthalmology department for evaluation. On the initial exam with academic ophthalmology, the visual acuity was normal and the clinical exam revealed no retinal or optic nerve abnormalities; these findings were consistent with the exam performed by the outside community ophthalmologist four months prior. To further evaluate, a fluorescein angiogram was done to assess for areas of non-perfusion or delayed circulation time. The angiogram showed normal circulation time, with no areas of non-perfusion or retinal ischemia seen in either eye. A complete systemic work up was done to rule out alternative explanation for the amaurosis fugax. Carotid Doppler showed no evidence of atherosclerotic disease. Blood work including ESR and CRP was normal.

Verbal consent was obtained from the patient to describe the findings for the literature.

3. Discussion

Amaurosis fugax is often caused by emboli from atherosclerotic carotid plaques. Hemodynamic abnormalities from vasculitis, such as giant cell arteritis, vasospasm, atherosclerotic stenosis, hyperviscosity, or hypercoagulability can lead to amaurosis fugax.² In this case, we hypothesize an atrial mass was releasing cellular matter into the retinal arterial circulation, resulting in transient episodes of vision loss. Cardiac vegetations are a less common source of systemic emboli, and may originate from intracavity thrombus, cardiac myxoma, and infective endocarditis, as well as valvular abnormalities including mitral valve prolapse and aortic valve calcification.³ Systemic emboli from left heart lesions, in particular left atrial myxoma, are much more common than pulmonary emboli with right heart lesions. Cardiac myxoma, a benign tumor of the heart, occurs in the left heart in approximately 75% of cases.⁴ However, malignant cardiac sarcomas are reported with increased frequency in the right heart, typically in the right atrium.⁴

A literature search for previous reports of systemic emboli and their sequelae in the setting of primary cardiac sarcoma revealed a two case reports of cerebral ischemia secondary to cardiac tumor and one case of cardiac chondrosarcoma with visual manifestations.^{5–7} Cardiac myxomas have been reported to present with retinal emboli. Reported ophthalmologic manifestations include unilateral retinal artery occlusion, as well as autopsy proven myxomatous involvement of choroidal arteries and posterior ciliary arteries.^{8,9} Cardiac sarcomas typically occur secondary to metastases; metastatic cardiac tumors are 30 times more likely than a primary neoplasm.^{10,11} A study of 75 patients with primary sarcomas of the heart cited common signs and symptoms of sarcoma as dyspnea, pain, shortness of breath secondary to pericardial effusion, chest pain, syncope, or hemoptysis.¹² Amaurosis fugax as a symptom of primary cardiac pleomorphic sarcoma is previously unpublished in the literature and thus we report a novel finding.

In our patient, presentation of transient monocular vision loss occurred without retinal findings such as Hollenhorst plaques,

cotton wool spots, or flame hemorrhages suggestive of emboli from atherosclerotic disease.¹³ Carotid Doppler imaging in this patient was negative for significant atherosclerotic stenosis as a potential cause of amaurosis fugax.^{14,15} Transient monocular vision loss due to vascular inflammation such as GCA was not consistent with the presentation in this patient. Although metastasis to the occipital cortex via the cerebral circulation from the primary cardiac tumor is possible, no evidence of intracranial pathology or anatomic abnormality was found on brain MRI to explain the transient visual loss in this patient.

Retinal migraines is a diagnosis of exclusion, and was the initial diagnosis provided to our patient. The patient did not provide a history of personal migraines, and his episodes were typically painless. While acephalgic migraines are possible, the lack of personal history makes retinal migraine unlikely. Moreover, a study of 142 patients with headache and transient monocular vision loss concluded that the diagnosis of "retinal migraine" was incorrect in all but 16 patients, with most patients having a secondary cause of vision loss.¹⁶

The findings on cardiac MRI and biopsy suggest that emboli from the sarcoma is the most credible cause for amaurosis fugax in this patient. The increased frequency of episodes following the initiation cardiac radiotherapy may suggest that with involution of the tumor, the release of cellular material occurred with increased frequency. We postulate that emboli from the left atrial sarcoma entered the systemic circulation. These emboli subsequently caused transient occlusion to the arteries supplying the optic nerve inner retina, outer retina and choroid, or watershed zones; this presented as "graying out" of vision.¹⁷ The process of transient embolic occlusion through the above mentioned process, leaves no visible evidence of retinal or optic nerve damage on clinical examination, and has no permanent visual sequela. Clinically, a retinal artery occlusion has clinical findings of retinal ischemia in the form of retinal whitening in the area of vascular distribution, and angiographic evidence of non-perfusion. Furthermore, there is irreversible vision loss corresponding to the areas of ischemia.

Our case report details the first published incidence of a cardiac pleomorphic atrial sarcoma as source of emboli to the systemic circulation to present with ophthalmologic symptoms. The absence of evidence pointing to common etiologies for transient vision loss strongly suggests that the sarcoma was the most likely cause of amaurosis fugax in this patient.

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