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Case Report

Clinical presentation of multiple cerebral emboli and central retinal artery occlusion (CRAO) as signs of cardiac myxoma

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Abstract

Cardiac myxomas are benign tumors of endocardial origin that usually occur in the left atrium. Trans-thoracic echocardiography is the diagnostic method of choice, and early surgical removal is the preferred method of treatment.
We present a patient whose history of cerebral emboli and central retinal artery occlusion (CRAO) led to a diagnosis of cardiac myxoma.
Neuroimaging studies showed multiple infarcts in the region of the left middle and anterior cerebral arteries. Ophthalmic examination showed gross retinal pallor compatible with left central retinal artery occlusion (CRAO).
The etiology of stroke was investigated by performing trans-thoracic echocardiography, which showed a mass in the left atrium compatible with cardiac myxoma. Complete removal of the cardiac tumor was performed by open-heart surgery.
Fortunately, after a period of rehabilitation, the patient's hemiparesis almost completely resolved, but the loss of vision OS remained unchanged.
Many cases of myxoma are accompanied by constitutional symptoms, such as anemia, fever and weight loss, which allow for a diagnosis to made before serious complications such as embolism occur. Unfortunately, in some patients, such as ours, the absence of signs and symptoms allows the myxoma to pass completely unnoticed until the first embolic event occurs.

Keywords: Cardiac myxoma, Central retinal artery occlusion, Cerebral emboli, Amaurosis

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Introduction

Cardiac myxomas are benign tumors of endocardial origin[1–3] that tend to be more prevalent in females (2:1).[3–5]

They usually originate in the left atrium (75%) or the atrial septum, close to the foramen ovale.[3,6,7] However, cases have been reported in other locations such as the right atrium and the right and left ventricles.[3]

In a paper published by Pineda et al.,[3] 85% of cardiac myxomas were pedunculated and 15% were immobile. As for the macroscopic appearance of these tumors, 34% were friable with a gelatinous consistency and either an irregular surface or a surface with papillae; 66% had a smooth surface.

Despite the fact that trans-esophageal echocardiography reaches a sensitivity of 100%,[3,10,11] the diagnostic method of choice is trans-thoracic echocardiography, which has a sensitivity of 95%.[3,8,9] CT and MRI play a role in the diagnosis of myxoma because they provide very accurate images that reveal tumor size and adherence to the cardiac septa.[3,12,13] Elevated ESR and anemia are the most characteristic laboratory results found in these patients.[3]

The treatment of choice is early surgical removal of the tumor.[3,14,15] The overall prognosis of these patients after

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surgery is excellent, although there may be a recurrence of between 5 and 14%, according to the literature.[3,16]

We present a patient whose history of cerebral emboli and central retinal artery occlusion (CRAO) led to a diagnosis of cardiac myxoma. This association has been previously described in the literature.[17–19]

Case report

A 53-year-old female, with diabetes and hypertension presented to the neuro-ophthalmology department in July 2013 with a history of sudden right-sided weakness and painless loss of vision in the left eye (OS).

Upon arrival to the emergency department, right-sided weakness and amaurosis OS were noted. Ophthalmic examination, performed 10 hours later, showed gross retinal pallor with narrowing of the retinal vessels.

CT and MRI of the brain were performed. The images revealed multiple small infarcts in the region of the left middle and anterior cerebral arteries compatible with an acute ischemic stroke (Fig. 1).

In establishing the etiology of stroke, carotid Doppler, MRA and CTA of neck were performed and no abnormalities were found. However, trans-thoracic ultrasound showed a mass in the left atrium, 2.2 × 2.5 cm in diameter (confirmed by trans-esophageal echocardiography). Cardiac MRI showed a pedunculated mobile tumor, 2.2 × 2.5 cm in diameter attached to the lateral wall of the left atrium (Fig. 2).

CBC results showed a hemoglobin level of 8 g/dl, indicating a normal thrombotic risk profile.

After informing the patient of the risks and benefits of surgery, complete removal of the tumor was performed by open-heart surgery. Histological analysis was compatible with atrial myxoma.

After gradual resolution of the hemiparesis through rehabilitation, the patient was referred to the neuro-ophthalmology department for consultation.

On examination, visual acuity (VA) was 20/30 OD and NLP (No Light Perception) OS. There was a left relative afferent pupillary defect (RAPD).

Fundus examination OD was unremarkable, however, temporal optic disc pallor OS with multiple vessel occlusions in the macula and inferior arterial arcade were observed (Fig. 3).

Neurological examination showed residual right-sided weakness (4/5 muscle balance of the right arm and leg). The remaining neurological examination was unremarkable.

Goldmann Perimetry was performed OD and shown to be normal. Due to poor vision, no results were obtainable for OS. Electroretinogram (ERG) showed decreased values OS (scotopic and photopic), and macular OCT showed gross retinal atrophy OS (Fig. 4).

All results were consistent with chronic central retinal artery occlusion (CRAO) OS.

Discussion

We present a patient whose history of cerebral emboli and central retinal artery occlusion (CRAO) led to a diagnosis of cardiac myxoma.

Fortunately, after a period of rehabilitation the patient's hemiparesis almost completely resolved, but the loss of vision OS remained unchanged. When questioned, the patient initially denied a prior history of embolism.

Unlike our patient, patients described in previous publications have had cardiac myxomas characterized by the presence of a triad of typical symptoms:

1. Mitral valve obstruction (54–95% of patients), in the setting of congestive heart failure.
2. Systemic embolism, which occurs in 10–45% of patients.[3–5,20] Pineda et al.[3] reviewed 112 patients with cardiac myxoma and detected the presence of embolism in 29% of patients (33 patients). Cerebral embolism was the most frequent (24 patients), even though 15 patients also showed embolism in the extremities; 4 patients had coronary artery embolisms and 1 patient had a central retinal artery occlusion (CRAO). Some patients experienced emboli in different locations simultaneously. Pineda et al.[3] also showed that men are at a statistically greater risk for embolic complications than women. Tumors with papillary or irregular surfaces are more friable, and therefore, associated with an increased risk of embolism. Lee et al.[21] published a series of 59 patients with cardiac myxoma, of which 22% had emboli; cerebral embolism was the most frequently encountered (within which the middle cerebral artery region was most affected). Only one patient in the series presented with CRAO.
3. Constitutional symptoms, such as fatigue, fever and weight loss, occur in up to 90% of patients with cardiac myxoma.[3,4] Women in general, as already mentioned, have a lower risk of embolism, yet present more frequently with constitutional symptoms.[3] Additionally, smooth surface myxomas have a stronger association with the presence of constitutional symptoms.[3,22]

Our patient originally denied any other symptoms prior to the embolism, but admitted upon further questioning to having experienced a period of general fatigue, more noticeable than the usual, in addition to very mild intermittent fevers. These symptoms may reflect the constitutional effects described in other patients with cardiac myxomas. In addition, chronic anemia was detected by laboratory analysis performed upon the patient's arrival at the emergency department.

Therefore, it can be assumed that if the patient had sought medical attention earlier and presented these symptoms, the myxoma may have been discovered before the occurrence of the embolic event.

It is important to include myxoma in the differential diagnosis of any patient with signs of mitral valve obstruction and additional constitutional symptoms (fatigue, fever and weight loss). Contemplating the possible existence of myxoma in these patients may prevent unnecessary consequences, such as in the case of our patient who presented with a massive embolism that resulted in serious health consequences.

With specific reference to visual symptoms, there are cases of central retinal artery occlusion (CRAO)[23–28] in the literature, and less frequently, cases of ophthalmic artery occlusion.[29] They normally occur in conjunction with cerebral embolism, yet there have also been cases of isolated visual impairment.[30–32]

In our patient, the CRAO occurred simultaneously with the cerebral embolism, most likely because the embolism broke

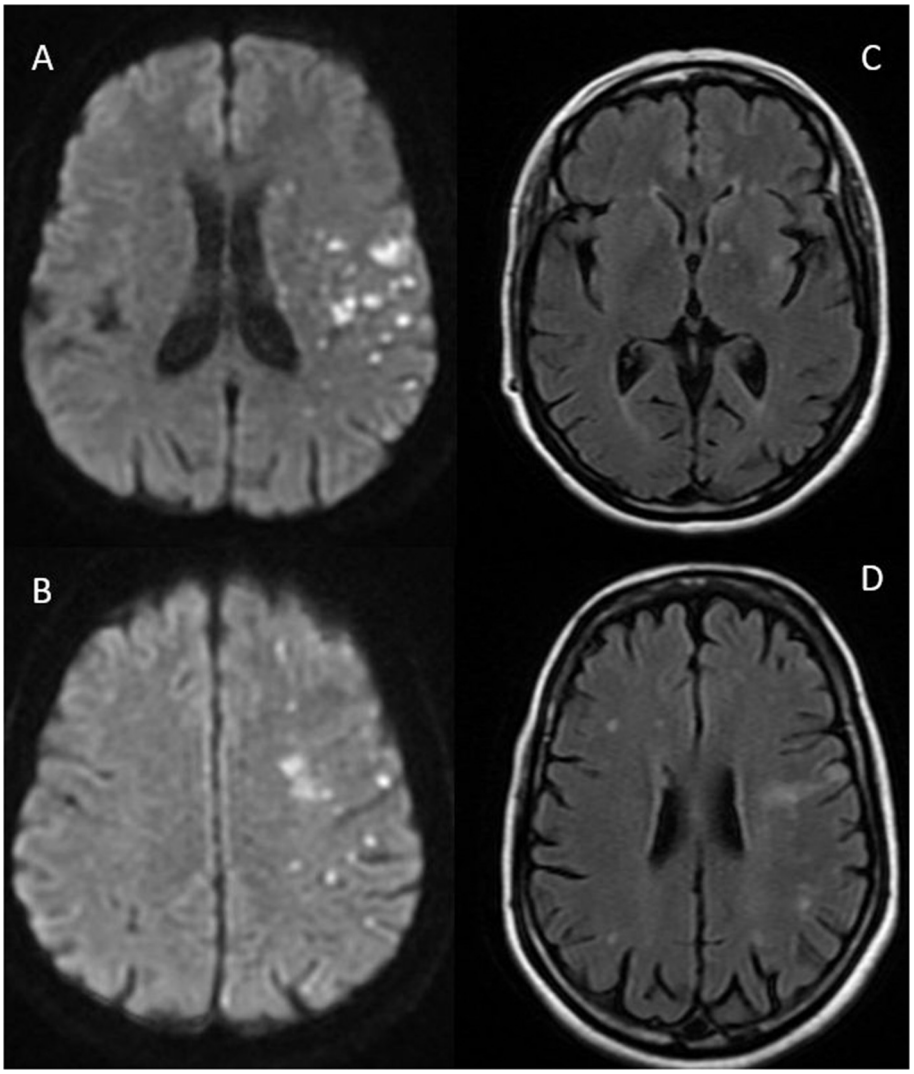

Fig. 1. MRI of the brain (diffusion-weighted MRI) showing multiple small acute infarcts in the region of the left middle and anterior arteries compatible with an infarct of embolic origin (A, B). Axial fluid-attenuated inversion recovery (FLAIR) MRI image in chronic phase demonstrating residual areas of cerebral ischemia (B, C).

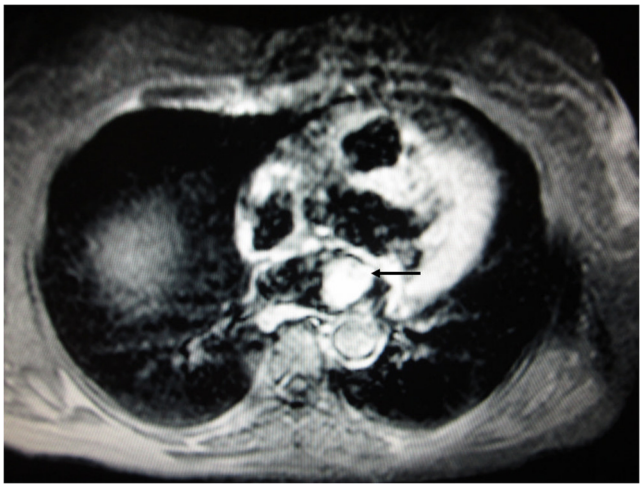

Fig. 2. Cardiac MRI showing a mobile pedunculated tumor 2.2 × 2.5 cm in diameter (arrow), joined to the lateral wall of the left atrium.

up and affected several arteries, including the middle cerebral artery (MCA), the anterior cerebral artery (ACA) and the central retinal artery.

According to previous publications, the incidence of CRAO in patients with myxoma is approximately 0.8–1.6%.[3,21] However, there are publications that estimate the incidence to be as high as 3%.[24]

Published cases of ophthalmic artery occlusion tend to show a variable clinical presentation. Orbital MRI demonstrates increased signal in the orbital fat, the optic nerve sheath and the extra-ocular muscles, compatible with ischemia secondary to arterial occlusion. The fundus has a whitish bone-colored appearance due to the absence of chorioretinal perfusion. Fluorescein angiography shows decreased retinal and choroidal flow with late phase transudation (leakage) secondary to impaired function of the retinal pigment epithelium (RPE).[29] In some publications, large choroidal arterial occlusions producing what are known as Amalric triangles have been described,[24] while in other cases, only small vessel occlusions producing Elschnig spots were observed.[30]

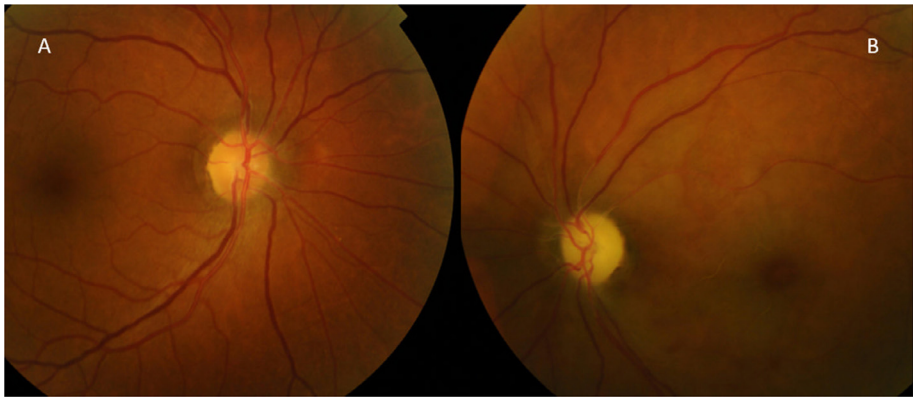

Fig. 3. Fundus photo showing a normal right eye (OD) (A). In the left eye (OS) there is temporal optic disc pallor with multiple occluded vessels in the macula and inferior arterial division of the retina (B).

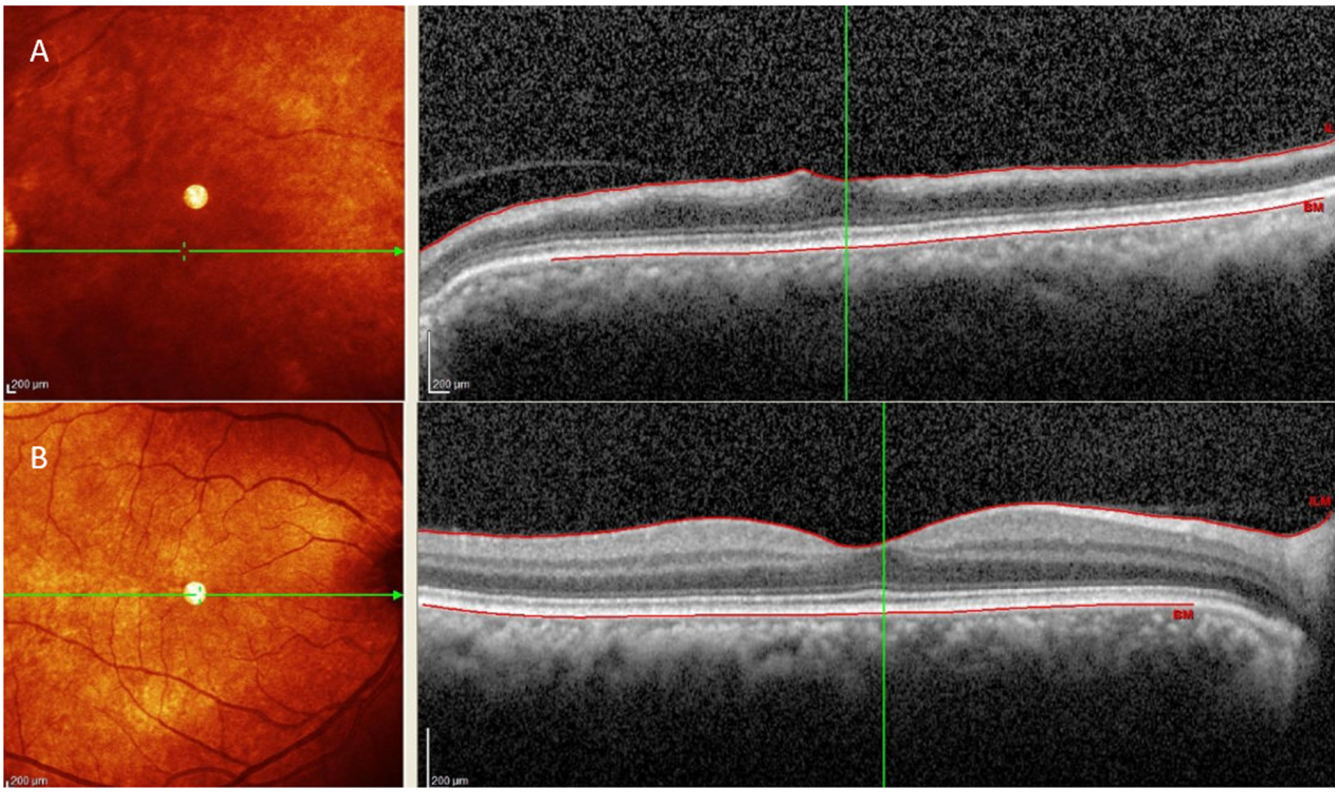

Fig. 4. Macular OCT showing gross retinal atrophy OS (A), and a normal OD (B).

In our patient, everything indicates that this is an isolated CRAO without any occlusion of the ophthalmic artery per se. Although we did not evaluate the patient in the acute phase, the description of the fundus at that time indicated the presence of whitish retinal edema, without any concurrent choroidal ischemia. Likewise, no change in orbital fat or extra-ocular muscle signal was noted on head and orbital MRI.

In some patients with CRAO, the presence of multiple arterial occlusions caused by a yellowish material have been described. These are in fact embolized tumor fragments.[23]

In our patient, only a marked and generalized attenuation of the retinal vessels during the acute phase was observed, without the presence of obstructive material. This indicates that the occlusion was most likely located in the retrolaminar region.

The most frequent causes of CRAO are cardiac embolism, coagulation disorders, migraine, hemoglobinopathies, sickle cell anemia, rheumatic disease, trauma, contraceptive use and intravenous drug abuse.[30]

In this case, we should reiterate that CRAO secondary to myxoma is rare, but should be considered if accompanied by symptoms of mitral valve obstruction and other embolic phenomena or the presence of fever, fatigue and anemia.[30]

These symptoms (which make up the classic triad of cardiac myxoma) should indicate the possible existence of myxoma. The onset of severe clinical complications can be prevented if a timely diagnosis is made.

However, in patients such as ours, the absence of typical symptoms allows a myxoma to pass completely unnoticed until the occurrence of the first embolic event. In such cases,

primary prevention of these complications is almost impossible.

Conflict of interest

The authors declared that there is no conflict of interest.

References

1. Johansson L. Histogenesis of cardiac myxomas. An immunohistochemical study of 19 cases, including one with glandular structures, and review of the literature. *Arch Pathol Lab Med* 1989 Jul;**113**(7):735–41.
2. Loire R. Is there a carcinologic malignant potentiality of cardiac myxoma? *Arch Mal Coeur Vaiss* 1991 Mar;**84**(3):395–9.
3. Pinede L, Duhaut P, Loire R. Clinical presentation of left atrial cardiac myxoma. A series of 112 consecutive cases. *Medicine (Baltimore)* 2001 May;**80**(3):159–72.
4. Markel ML, Waller BF, Armstrong WF. Cardiac myxoma. A review. *Medicine (Baltimore)* 1987 Mar;**66**(2):114–25.
5. Reynen K. Cardiac myxomas. *N Engl J Med* 1995;**333**:1610–7.
6. Chopra P, Ray R, Airan B, Talwar KK, Venugopal P. Appraisal of histogenesis of cardiac myxoma: our experience of 78 cases and review of literature. *Indian Heart J* 1999;**51**(1):69–74.
7. Fine G, Morales A, Horn Jr RC. Cardiac myxoma. A morphologic and histogenetic appraisal. *Cancer* 1968 Dec;**22**(6):1156–62.
8. Nomeir AM, Watts LE, Seagle R, Joyner CR, Corman C, Prichard RW. Intracardiac myxomas: twenty-year echocardiographic experience with review of the literature. *J Am Soc Echocardiogr* 1989;**2**(2):139–50.
9. Obeid AI, Marvasti M, Parker F, Rosenberg J. Comparison of transthoracic and transesophageal echocardiography in diagnosis of left atrial myxoma. *Am J Cardiol* 1989 Apr 15;**63**(13):1006–8.
10. Engberding R, Daniel WG, Erbel R, Kasper W, Lestuzzi C, Curtius JM, et al. Diagnosis of heart tumours by transoesophageal echocardiography: a multicentre study in 154 patients. European Cooperative Study Group. *Eur Heart J* 1993 Sep;**14**(9):1223–8.
11. Lukács L, Lengyel M, Szedö F, Haán A, Nagy L, Thomka I, et al. Surgical treatment of cardiac myxomas: a 20-year follow-up. *Cardiovasc Surg* 1997 Apr;**5**(2):225–8.
12. Freedberg RS, Kronzon I, Rumancik WM, Liebeskind D. The contribution of magnetic resonance imaging to the evaluation of intracardiac tumors diagnosed by echocardiography. *Circulation* 1988 Jan;**77**(1):96–103.
13. Casey M, Mah C, Merliss AD, Kirschner LS, Taymans SE, Denio AE, et al. Identification of a novel genetic locus for familial cardiac myxomas and Carney complex. *Circulation* 1998 Dec 8;**98**(23):2560–6.
14. Castells E, Ferran V, Octavio de Toledo MC, Calbet JM, Benito M, Fontanillas C, et al. Cardiac myxomas: surgical treatment, long-term results and recurrence. *J Cardiovasc Surg (Torino)* 1993 Feb;**34**():49–53.
15. Meyns B, Vancleemput J, Flameng W, Daenen W. Surgery for cardiac myxoma. A 20-year experience with long-term follow-up. *Eur J Cardiothorac Surg* 1993;**7**(8):437–40.
16. Cooley DA. Surgical treatment of cardiac neoplasms: 32-year experience. *Thorac Cardiovasc Surg* 1990 Aug;**38**(Suppl 2):176–82.
17. Kim JH, Youn HJ, Jung MH, Oh CY, Ahn SH, Cho WH, et al. Retinal artery occlusion by left atrial myxoma misdiagnosed as thrombus. *Springerplus* 2016 Mar;**22**(5):352.
18. Yu Y, Zhu Y, Dong A, Su Z. Retinal artery occlusion as the manifestation of left atrial myxoma: a case report. *BMC Ophthalmol* 2014 Dec;**23**(14):164.
19. Sabater N, Alforja S, Rey A, Giralt J. Delayed diagnosis of ophthalmic artery obstruction due to atrial myxoma]. *Arch Soc Esp Oftalmol* 2013 Aug;**88**(8):313–5.
20. St John Sutton MG, Mercier LA, Giuliani ER, Lie JT. Atrial myxomas: a review of clinical experience in 40 patients. *Mayo Clin Proc* 1980;**55**():371–6.
21. Lee SJ, Kim JH, Na CY, Oh SS. Eleven years' experience with Korean cardiac myxoma patients: focus on embolic complications. *Cerebrovasc Dis* 2012;**33**(5):471–9.
22. Larsson S, Lepore V, Kennergren C. Atrial myxomas: results of 25 years' experience and review of the literature. *Surgery* 1989 Jun;**105**():695–8.
23. Lewis JM. Multiple retinal occlusions from a left atrial myxoma. *Am J Ophthalmol* 1994 May 15;**117**(5):674–5.
24. Lee SY, Loo JL, Ang CL. Ischemic oculopathy as a complication of surgery for an atrial myxoma. *Arch Ophthalmol* 2004 Jan;**122**():130–1.
25. Thompson J, Kapoor W, Wechsler LR. Multiple strokes due to atrial myxoma with a negative echocardiogram. *Stroke* 1988 Dec;**19**():1570–1.
26. Ong CT, Chang RY. Intravenous thrombolysis of occlusion in the middle cerebral and retinal arteries from presumed ventricular myxoma. *Stroke Res Treat* 2010 Dec;**2**(2011):735057.
27. Schmidt D, Hetzel A, Geibel-Zehender A. Retinal arterial occlusion due to embolism of suspected cardiac tumors – report on two patients and review of the topic. *Eur J Med Res* 2005 Jul 29;**10**():296–304.
28. Sachers F, Guignard P. Cardiac myxoma with central retinal artery occlusion, hemiparesis and epileptic seizure in a 9-year-old patient. *Klin Monbl Augenheilkd* 1992 Aug;**201**(2):112–5.
29. Rafuse PE, Nicolle DA, Hutnik CM, Pringle CE. Left atrial myxoma causing ophthalmic artery occlusion. *Eye (Lond)* 1997;**11**(Pt):25–9.
30. Porrini G, Scassellati-Sforzolini B, Mariotti C, Giuliani M, Giovannini A. Plurifocal cilioretinal occlusion as the presenting symptom of cardiac myxoma. *Retina* 2000;**20**(5):550–2.
31. Yasuma F, Tsuzuki M, Yasuma T. Retinal embolism from left atrial myxoma. *Jpn Heart J* 1989 Jul;**30**(4):527–32.
32. Salehian O, Demers C, Patel A. Atrial myxoma presenting as isolated unilateral blindness: a case report and review of the literature. *Can J Cardiol* 2001 Aug;**17**(8):898–900.