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CASE REPORT

Cerebrovascular diseases and skin pseudovasculitis masquerading a mass in the heart treated surgically (ophthalmic artery oocclusion due to left atrial myxoma)

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ABSTRACT

Left atrial myxoma is the most common primary cardiac neoplasm, commonly grows on the upper side of atrial septum and has often mimicked connective tissue disease especially some features of vasculitis. The clinical course of the left atrial myxoma is characterized by symptoms resulting from obstructive, embolic, or "pseudovasculitis " manifestations of the tumor. Embolic ischemic manifestations are typically cerebrovascular. We described an unusual clinical presentation of left atrial myxoma in 36 years old man, which initially was assessed as PAN type vasculitis for 4 years. He presented with episodic red skin rashes, blurred vision, abdominal pain, headache and hemiparesia. Based on his symptoms, diagnosis of polyarteritis nodosa type vasculitis was made. Laboratory findings included nothing to indicate systemic vasculitis. Cardiac evaluation revealed no murmur. Brain MRI revealed left parietal cortical ischemic patchy signals. Being unresponsiveness to medical therapy led to echocardiography which demonstrated left atrial mass (2.9 in 2cm). After surgical intervention diagnosis of left atrial myxoma was approved with pathological examination. This case report emphasize that sometimes the symptoms and physical findings of cardiac tumor could simulate systemic vasculitis. Prompt clinical recognition and surgical removal are essential to prevent serious complications.

Key words: cerebrovascular; myxoma; vasculitis; ophthalmic artery

INTRODUCTION

Left atrial myxoma is the most common primary cardiac neoplasm, has often mimicked some features of vasculitis. It may present with obstruction of mitral valve or an embolic phenomenon.[1,2] Embolization can block blood flow and cause several signs and symptoms such as skin rashes, visual loss, GI disorders and many others that may remember us vascular disease.[2,3]

In this study, we describe an unusual clinical presentation of left atrial myxoma in 36 years old man, which initially was assessed as PAN type vasculitis. He presented with unilateral blindness, skin rash, abdominal pain, headache and hemiparesia. Echocardiography demonstrated left atrial mass. After surgical intervention diagnosis of left atrial myxoma was approved.

CASE REPORT

A 36-years-old man faint before going to the bedroom (he had jerky movements and was unconscious for 20 min). He had 4 years history of episodic red skin livedoid and purpuric rash localized in his hand and legs, slowly disappeared after several days. These skin lesions were appeared three more times that were accompanied with blurred vision, recurrent abdominal

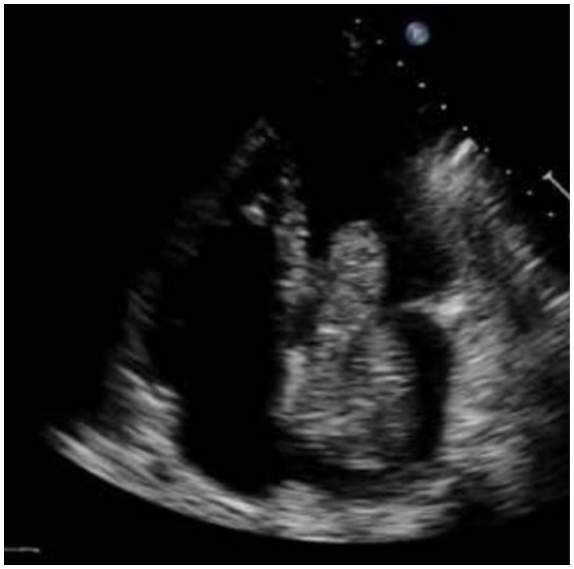

Figure 1, Transthoracic echocardiogram: showing a solid, giant, smooth mass myxoma (2.9×2cm) protrude to left ventricle during diastole.

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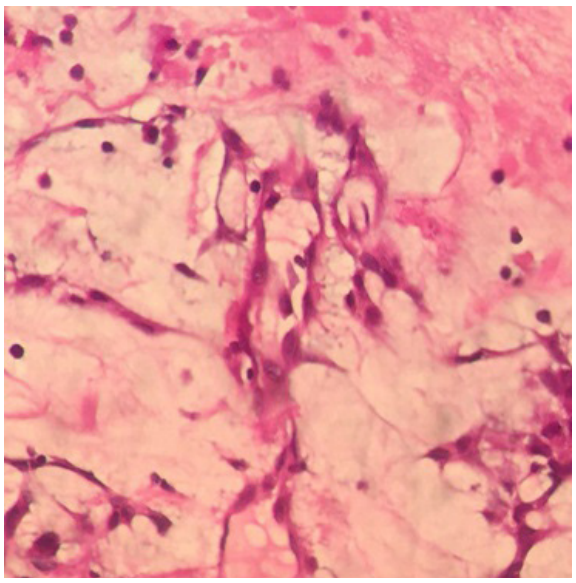

Figure 2, Tumor histology showing myxoma cells in small clusters partially cuffing capillaries Hematoxylin and Eosin (H&E, 40x).

pain and severe headache.

So the physician treated him outpatient with probable diagnosis of systemic vasculitis. At the time of admission he was unconscious and suffered from dysarthria and blurred vision. He lost left eye visual field completely. On physical examination his chest was clear and cardiac evaluation revealed no murmur, click, gallop or another abnormality. Pulmonary and abdominal examination was normal and the pulses in all of his upper and lower extremities was symmetric. Livedoid skin rash and splinter hemorrhage was found. In neurological exam right side hemiparesia was detected. In eye examination diagnosis of Central Retinal Artery Occlusion (CRAO) was detected by ophthalmologist. He denied any history of weight loss, fever, palpitation, dyspnea, cardiac problem, smoking, alcohol abuse and exposure to animals. He had not hypertension, hyperlipidemia, migraine, diabetes, epilepsy and malignancy in his past history. All laboratory findings were normal or negative. Laboratory all findings included normal or negative: CRP was negative, ESR= 4mm/h, PT and PTT was in normal range, Anti cardiolipin IgG 0.5 U/ml and IgM 0.1, B2 glycoprotein IgG 1 and IgM 2.8, Lupus anticoagulant was negative, HBS Ag, HCV Ab was negative, Cryoglobulin not detected, ANA was 0.4 U/ml, Anti ds DNA 1.5 IU/ml, P-ANCA 1 U/ml, C-ANCA 2 U/ml, CH50 122 U/ml, C3 98 mg/dl and complement C4 19 mg/dl, Urinalysis, electrolytes, liver enzymes and blood cell count all was in normal range.

Perimetery test showed complete defect on left eye visual field (also VEP exam indicated prechiasmatic injury of left visual pathways). Electerodiagnostic study of upper and lower limbs was normal. In Doppler sonography intima-media thickness of both carotid was in normal limit and atherosclerotic stenosis was not found. Multiplanar Brain MRI was done that revealed left parietal abnormal cortical ischemic signal change with weighted images abnormal patches in left side. After contrast administration, no abnormal vessels or enhancement was noted. Contrast enhanced wide aortic study Magnetic resonance imaging (MRI) was done with multiplanar reconstruction and revealed normal result. At the electroencephalography (EEG), bursts of bilateral sharp waves were observed on the tracing which is more prominent in the left hemisphere and appropriate neuroimaging and automatic external defibrillator (AED) were advised.

Finally in echocardiography, a large mobile vegetative mass attached to lower part of IAS with thickening of IAS at that site was looked through in echocardiography. The mass size was 2.9×2cm. mass was protruded on MV annular area, but no attachment to mitral valve surface (Fig. 1). Furthermore mild MR, mild TR and classic MVP were reported. Connection site was suggestive for myxoma but mass shape proposes other left atrium (LA) tumors (respect to shape of mass other LA tumors should was been considered).

After LA opening, large fragile and myxomatous tumor in LA was visualized (5×5 cm) with large base adhering to international accounting standards (IAS) that was enucleated from septum with fine layer of endocardium. The pathologist report myxoma (Fig.2).

DISCUSSION

Myxoma is the common primary tumor of the heart, with one of three kind symptoms: obstruction, embolisation and systemic involvement such as fever, weight loss or symptoms resembling vascular disease.[3, 4] The occurrence of rash and ophthalmic complication are suspicious of atrial myxoma, but has also been seen in systemic vasculitis. There is considerable symptomatic overlap between atrial myxoma and systemic vasculitis5. The diagnosis can made by simple transthoracic echocardiogram (TTE).[6]

Various disease can lead to retinal artery occlusion. They may be caused by an emboli originating from the heart or carotid artery. Valve vegetation of the heart has also caused vascular retinal occlusion.[7]

Ocular disturbance due to emboli of cardiac tumor are rare [7, 8] noted in a few 9-18 reported cases in literature [7, 19], actually the eye is the extremely rare target organ of myxomatous emboli. The first case of the central retinal artery emboli due to myxoma with blindness was reported in 1934 by Reichlin[3], but until now a few studies have been reported.[7,9-12,17] Ophthalmoscopy is important so can detected central retinal artery occlusion.[16]

CONCLUSION

This case report emphasize that the signs and symptoms of cardiac tumor could simulate vascular disease. On the other hand, sometimes retinal artery occlusion such as embolic event can caused by a cardiac tumor. So atrial myxoma should be considered in the differential diagnosis of any embolic disease and ev-

ery one with focal neurological signs should have a thorough cardiac examination. As a result, clinical recognition and surgical removal are essential to prevent serious complications.

CONFLICT OF INTEREST

None.

REFERENCES

1. Mouine NN, Asfalou II, Raissouni MM, Benyass AA, El Mehdi EZ. Giant left atrial myxoma mimicking severe mitral valve stenosis and severe pulmonary hypertension. International archives of medicine. 2013 Apr 19;6(1):13.
2. Mizuno R, Hayata Y, Taniguchi S, Saito Y, Okamoto Y, Fujimoto S. Giant left atrial myxoma causing severe pulmonary hypertension. Journal of echocardiography. 2011 Dec 1;9(4):151-3.
3. Tonz M, Laske A, Carrel T, da S, V, Real F, Turina M. Convulsions, hemiparesis and central retinal artery occlusion due to left atrial myxoma in child. Eur J Pediatr 1992 Sep; 151(9):652-4.
4. Lee VH, Connolly HM, Brown RD, Jr. Central nervous system manifestations of cardiac myxoma. Arch Neurol 2007 Aug;64(8):1115-20.
5. Thomas MH. Myxoma masquerading as polyarteritis nodosa. J Rheumatol 1981 Jan; 8(1):133-7.
6. Dhawan S, Tak T. Left atrial mass: Thrombus mimicking myxoma. Echocardiography. 2004 Oct 1;21(7):621-3.
7. 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(7):296-304.
8. Rafuse PE, Nicolle DA, Hutnik CM, Pringle CE. Left atrial myxoma causing ophthalmic artery occlusion. Eye (Lond) 1997; 11:25-9.
9. Cogan DG, Wray SH. Vascular occlusions in the eye from cardiac myxomas. Am J Ophthalmol 1975 Sep; 80(3 Pt 1):396-403.
10. Matamoros N, BenEzra D. Bilateral retinopathy and encephalopathy. Graefes Arch Clin Exp Ophthalmol 1989; 227(1):39-41.
11. Donaldson RM, Emanuel RW, Earl CJ. The role of two-dimensional echocardiography in the detection of potentially embolic intracardiac masses in patients with cerebral ischaemia. Journal of Neurology, Neurosurgery & Psychiatry. 1981 Sep 1; 44(9):803-9.
12. Furlong BR, Verdile VP. Myxomatous embolization resulting in unilateral amaurosis. The American journal of emergency medicine. 1995 Jan 1; 13(1):46-9.
13. Dahrling BE. The histopathology of early central retinal artery occlusion. Arch Ophthalmol 1965 Apr; 73:506-10.
14. Byrd WE, Matthews OP, Hunt RE. Left atrial myxoma presenting as a systemic vasculitis. Arthritis Rheum 1980 Feb; 23(2):240-3.
15. Boussen K, Moalla M, Blondeau P, Ben AH, Lie JT. Embolization of cardiac myxomas masquerading as polyarteritis nodosa. J Rheumatol 1991 Feb; 18(2):283-5.
16. Taylor RH, Deutsch J. Myxoma mix-up. A case report. J Clin Neuroophthalmol 1992 Sep; 12(3):207-9.
17. Manschot WA. Embolism of the central retinal artery originating from an endocardial myxoma. Am J Ophthalmol 1959 Sep; 48:381-5.