Rapid Growing Recurrent Tumors Causing Peripheral Emboli; are Myxomas Really "Benign"?: Case Report

Periferik Emboliye Yol Açan, Hızlı Büyüyen ve Tekrarlayan Tümör; Miksomalar Gerçekten "Selim" midir?

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ABSTRACT Cardiac myxoma is the most frequent "benign" tumor of the heart that may present diagnostic challenge. The tumor has many clinical manifestations that may mimic other pathologies. Recurrences were reported from as soon as a few months to as long as 8 years after excision of the myxoma. It is estimated that myxomas grow 0.15 mm in a month or 18 mm in a year normally. A case that was operated twice in one year for recurrent myxoma with a growing rate of 1.81 mm/month is presented here. The case which manifested with systemic emboli to the brain is one of the fastest growing recurrent myxomas reported in the literature.

Key Words: Embolism; myxoma; heart ventricles

ÖZET Kardiyak miksomalar tanıda zorluk çıkarabilen, kalbin en sık rastlanılan iyi huylu tümörleridir. Tümör, diğer patolojileri taklit edebilen birçok klinik bulguya sahiptir. Miksomanın cerrahi olarak çıkarılmasından sonra birkaç ay ile 8 yıl sonra bildirilmiş tekrarlayan vakalar mevcuttur. Miksomaların büyüme hızı ayda 0,15 mm ya da yılda 18 mm olarak kabul edilir. Burada ayda 1,81 mm büyüme hızına sahip, bir yılda 2 kere opere edilen tekrarlayan miksoma vakası sunulmaktadır. Beyine sistemik emboli ile başvuran olgu, literatürde bildirilen en hızlı büyüyen tekrarlayan miksoma olgularından biridir.

Anahtar Kelimeler: Emboli; miksoma; kalp ventrikülleri

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Cardiac myxoma is the most frequent "benign" tumor of the heart that may present diagnostic challenge. The tumor has many clinical manifestations that may mimic other pathologies. Systemic emboli is seen in 10-45% of myxoma patients.[1] Although recurrence of myxoma is frequently encountered in familial type, it is extremely rare for the sporadic type to regrow in short time interval and cause life threatening systemic emboli. A case that was operated twice in one year for recurrent myxoma that caused systemic emboli is presented.

CASE REPORT

A 44 -year-old man was admitted to the hospital with effort dyspnea. The patient was a heavy smoker and was diagnosed Buerger disease 6 years ago. In his physical examination the only pathological finding was the loss of

pedal pulses. Echocardiography revealed two pedunculated mobile masses in the left ventricle; first mass attached to ventricle apex with a stalk and the second one attached to anteroapical interventricular septum with dimensions of 2,4x2,7 cm and 0,95x1,59 cm, respectively (Figure 1a). There was also increase in the left atrial and ventricular dimensions along with mild mitral regurgitation. There was no history of cardiac tumor in his family. Coronary angiography was normal. The patient underwent operation.

At the operation, following median sternotomy, cardiopulmonary bypass was established. Left atriatomy was performed and the cardiac mass was visualized through mitral valve. However, the mass was not resectable from this orifice; hence 4 cm long left ventriculotomy in the apical region was also performed. Two masses with dimensions of 4x4 cm and 1x1 cm whose stalks resembled cordae tendinea were completely excised (Figure 2). The ventriculotomy was closed with Teflon felts.

The postoperative term was uneventful except for the paroxysmal atrial fibrillation rhythm disturbance. The patient was discharged home on the 6th postoperative day on warfarin and amiodorone treatment. Histologicaly, the mass consisted of mixoid matrix and rudimentary vessels where globoid and star shaped myxoma cells were scattered in, hence myxoma diagnosis was confirmed. During follow-up, two echocardiographical evaluations on the postoperative 1st and 3rd months didn't reveal any residual tumor. The patient was lost to follow-up then.

11 months after the operation, the patient was admitted to the emergency ward with sensorial aphasia and amnesia. The cranial diffusion magnetic resonance imaging detected an acute middle cerebral artery territory infarction which recovered after heparin treatment. Echocardiography showed two mobile masses localized at the anteroseptum and anterior apex of the left ventricle with dimensions of 1,9x2 cm and 1,8x1,6 cm (Figure 1b).

At the second operation, same surgical technique that had been used earlier was performed. The two pedunculated, mobile masses 3x1,5 and 1x1,5 cm were excised. The origins of each stalk were cauterized. Histopathological evaluation disclosed the tumor to be a myxoma. At the postoperative 6th month follow up, there was no residual tumor echocardiographically and he had a complete neurologic recovery.

DISCUSSION

The incidence of primary cardiac tumors varies from 0.02% to 0.28% and they are one of the least common causes of cerebrovascular accidents.[2,3] Almost all of these tumors are sporadic benign myxomas, of which 7% are recurrent tumors.[3] Re-

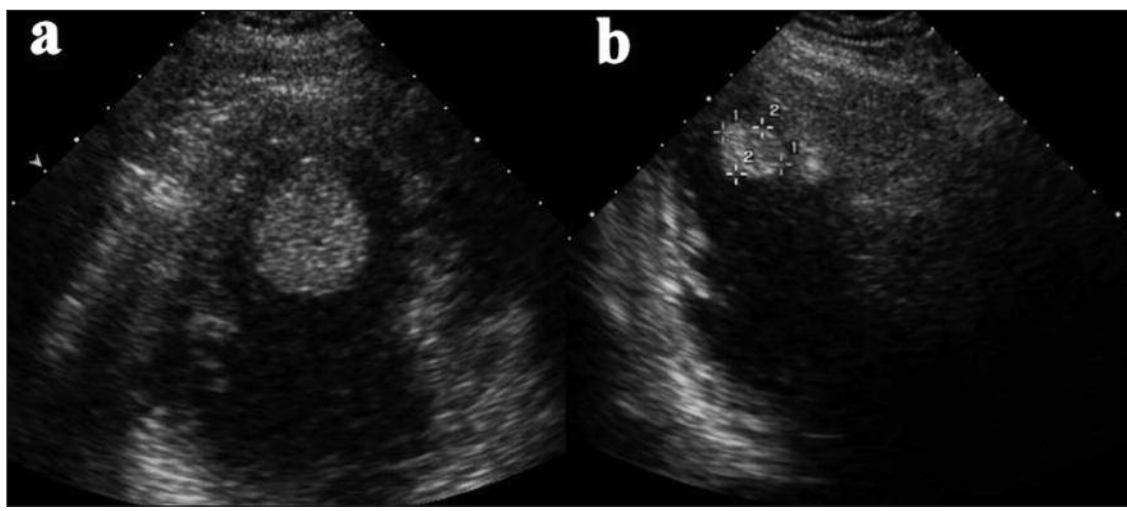

FIGURE 1: a) Preoperative transthoracic echocardiogram of left ventricle showing cardiac myxoma and its estimated size. **b)** Preoperative transthoracic echocardiogram of left ventricle showing recurrent myxomas.

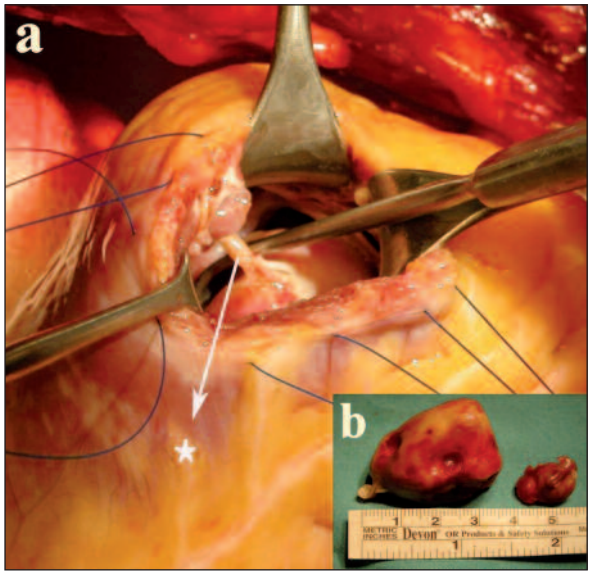

FIGURE 2: a) Operational view of apical myxoma (1st operation) * indicates the stalk of the tumor. **b)** Macroscopic view of the two tumors (1st operation). (See color figure at http://cardivascular.turkiyeklinikleri.com/)

currences are reported from as soon as a few months to as long as 8 years after excision of the myxoma.[1] It is estimated that myxomas grow 0.15 mm in a month or 18mm in a year.[4,5] In our case both tumors grew nearly 2 cm in 11 months (1,81 mm/month), being one of the fastest growing recurrent myxomas reported in the literature.

The overall risk of recurrence for familial and Carney complex myxomas is about 12% and 22%, respectively, whereas it is only 1-3% for sporadic tumors.[1,4,6] Although we didn't perform any genetic screening, we consider our case to be a sporadic myxoma since there is no familial history nor concomitant endocrine tumor and skin lesions. The cause of recurrence is not clear, but probable risk factors include inadequate or incomplete resection, intracardiac implantation, intraoperative displacement of tumor material, embolization, multicentricity of the tumor, and/or a reserve of tumor precursor cells in the subendocardium.[7]

The treatment of choice for myxomas is surgical removal. The most common approach is through either right or left atrial depending on the location of the tumor.[8] However ventricular approach may be necessary when atrial approach does not provide sufficient exposure for total resection. Minimal invasive and robotic surgical excisions of ventricular myxomas have been reported recently.[9,10] Chemotherapy and radiation therapy are disappointing. Radical excisional therapy and total cardiac transplantation for the unresectable tumors have been reported.[4] Once the diagnosis is made, urgent excision of the tumor is essential. Embolic complications or sudden death are always possible, even in asymptomatic patients.

Regular echocardiography after surgical resection of benign cardiac myxomas is utmost important in detecting recurrence and avoiding potential complications. Genetic analysis combined with echocardiographic surveillance of family members would enable exclusion of familial syndromes.

REFERENCES

1. Pinede L, Duhaut P, Loire R. Clinical presentation of left atrial cardiac myxoma. A series of 112 consecutive cases. Medicine (Baltimore) 2001;80(3):159-72.
2. Knepper LE, Biller J, Adams HP Jr, Bruno A. Neurologic manifestations of atrial myxoma. A 12-year experience and review. Stroke 1988;19(11):1435-40.
3. Rathore KS, Hussenbocus S, Stuklis R, Edwards J. Novel strategies for recurrent cardiac myxoma. Ann Thorac Surg 2008;85(6):2125-6.
4. Hermans K, Jaarsma W, Plokker HW, Cramer MJ, Morshuis WJ. Four cardiac myxomas diagnosed three times in one patient. Eur J Echocardiogr 2003;4(4):336-8.
5. Liesting C, Ramjankhan FZ, van Herwerden LA, Kofflard MJ. Systemic embolisation as presentation and recurrence of cardiac myxoma two years after surgery. Neth Heart J 2010;18(10):499-502.
6. Mendoza C, Bernstein E, Ferreira A. Multiple recurrences of nonfamilial cardiac myxomas: a report of two cases. Tex Heart Inst J 2007;34(2):236-9.
7. Moyssakis I, Anastasiadis G, Papadopoulos D, Margos P, Votteas V. Second recurrence of cardiac myxoma in a young patient. A case report. Int J Cardiol 2005;101(3):501-2.
8. Gajjar TP, Shah GB, Desai NB. Giant ventricular myxoma obstructing right ventricular outflow tract. Rev Bras Cir Cardiovasc 2011;26(4):663-6.
9. Hassan M, Smith JM. Robotic assisted excision of a left ventricular myxoma. Interact Cardiovasc Thorac Surg 2012;14(1):113-4.
10. Modi P, Hassan A, Chitwood WR Jr. Minimally invasive transaortic thoracoscopic resection of an apical left ventricular myxoma. J Thorac Cardiovasc Surg 2009;138(2):510-2.