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# Left ventricular myxoma: Confusing intracardiac mass in a patient with leukemia in remission

**Case Report**

Asli Tanindi[1], Gulten Tacoy[1*], Fatma Hizal[1], Murat Turfan[1], Emrah Ereren[2], Levent Oktar[2], Adnan Abaci[1]

[1] *Gazi University Faculty of Medicine, Cardiovascular Medicine, 06500 Ankara, Turkey*

[2] *Gazi University Faculty of Medicine, Cardiovascular Surgery, 06500 Ankara, Turkey*

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**Abstract:** Cardiac myxomas are the most common primary benign tumors of the heart and appears particularly in the left atrium. Myxomas exhibit a wide spectrum of symptoms from asymptomatic to very serious according to the localisation of the tumours. Only 5% of these tumours occur in the right and left ventricles. We report an unusual case of a 21-year-old woman with a leukemia in remission, in whom routine transthorasic echocardiographic examination demonstrated an incidental left ventricular mass. The patient underwent successful resection of the left ventricular mass under cardiopulmonary bypass through the left atrial approach. Histopathologic examination was diagnostic for myxoma. In this case, the presentation of the mass was atypical for two reasons: the tumour was a cardiac myxoma in the left ventricular localisation and in a patient with leukemia in remission.

**Keywords:** *Myxoma • Left ventricle • Leukemia*



## 1. Introduction

The incidence of primary cardiac tumours has a wide range from 0.02% to 0.28% [1]. 75% of those patients have benign tumours [2,3]. Cardiac myxomas are the most common primary benign tumors of the heart and appear particularly in the left atrium (75%). Although transthorasic (TTE) and transesophageal (TEE) echocardiographic examinations are important tools in diagnosis, the definitive diagnostic method is histopathologic investigation. Concomitant presence of leukemia has not been described previously. The aim of this study is to report the case of a patient with pedunculated left ventricular myxoma and leukemia in remission.

## 2. Case Report

A 21-year-old woman was admitted to the outpatient cardiology clinic for routine echocardiographic assessment after autologous stem cell transplant for acute myeloblastic leukemia. She was diagnosed with acute myeloblastic leukemia in December 2006. She had received high doses of idarubicin and cytosine arabinoside (Ara-C) as first-line therapy. One year after the first diagnosis, the leukemia had relapsed and she had undergone allogeneic stem cell transplantation. One month after the transplantation, bone marrow biopsy showed no residual leukemia or reticulin increase. She was diagnosed to have heterozygous Factor V Leiden mutation when she was searched for any genetic mutations for several clinical conditions. She had received immunosupressant therapy, antibacterial, antifungal and antiviral agents for 9 months after the transplantation.

In the initial assesment she was asymptomatic and her physical examination was completely normal at

* *E-mail: gtacoy@gmail.com*

**Figure 1.** Apical four chamber view demonstrates left ventricular myxoma.

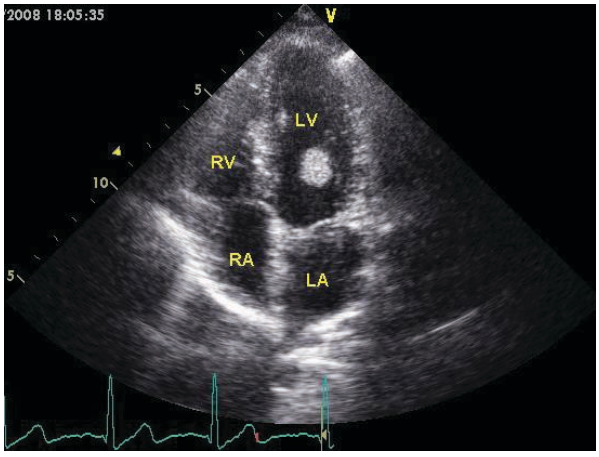



**Figure 2.** Appearance of left ventricular myxoma in parasternal axis view.

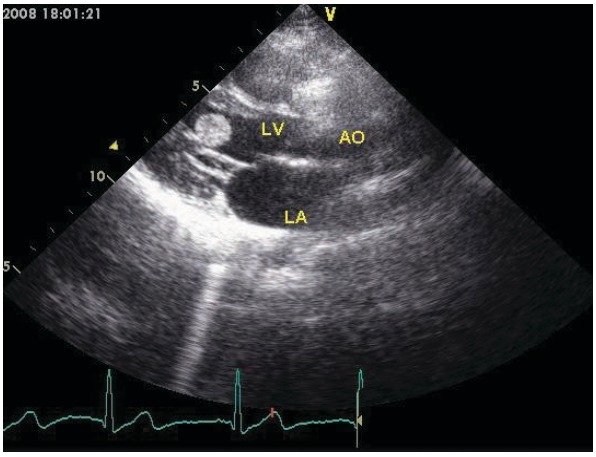



**Figure 3.** Arrowhead shows left ventricular mass in parasternal short axis view.

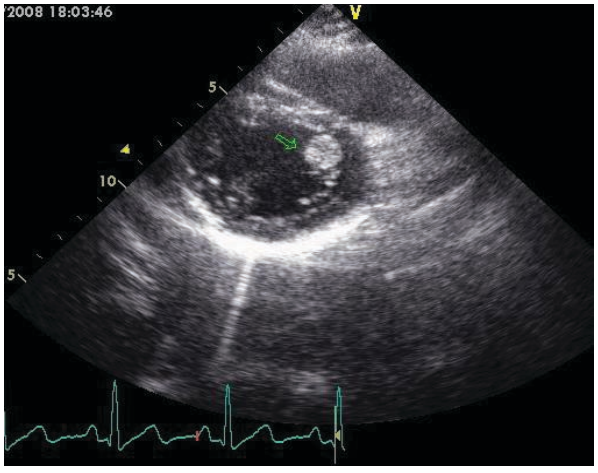
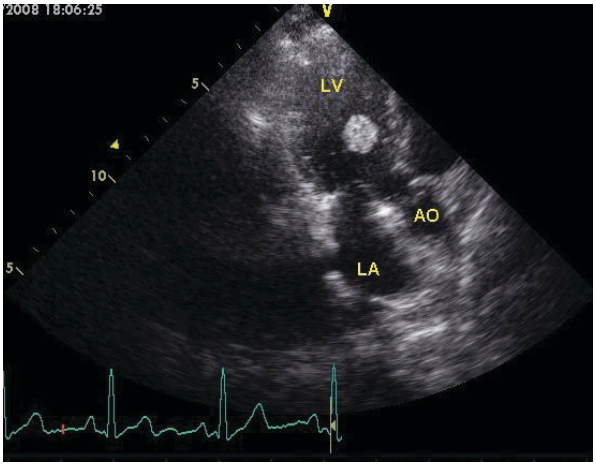



**Figure 4.** İnterventricular septum attached smooth-appearing mass is shown.

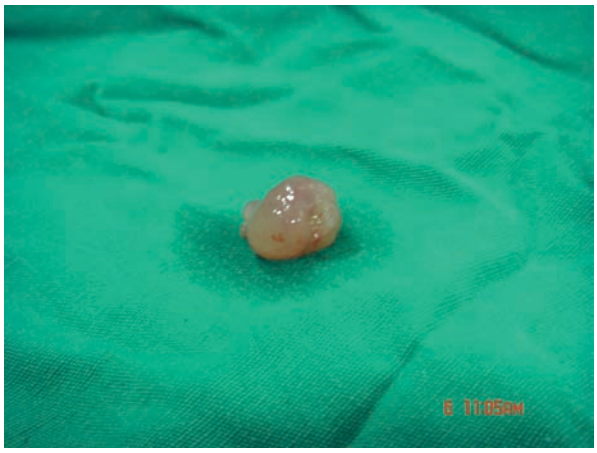



that time. X-ray of the chest and electrocardiography were unremarkable. Complete blood count and biochemistry were all in the normal range. Erythrocyte sedimantation rate and level of C-reactive protein were normal. Transthorasic echocardiography revealed a pedunculated, smooth-appearing mobile mass of 2.2x1.7 cm attached to the interventricular septum (Figure 1-4). The septum was of normal thickness; there was normal contractility of the left ventricle with 60% ejection fraction and normal right heart chambers without pulmonary hypertension. Due to her leukemia history, it had been proposed that the mass might be a trombus. Therefore she has received intravenous heparin therapy for one week. At the end of the week TTE was performed again. There was no decrease in the size of the mass, so it was decided to remove the mass urgently with surgical intervention. The patient underwent successful resection of the left ventricular mass with median sternotomy under cardiopulmonary bypass through the left atrial approach (Figure 5). After resection, intraoperative transesophageal echocardiography revealed no residual tumour. Histopathology revealed benign cardiac myxoma. The patient's postoperative course was uneventful and she was discharged after one week without any complications.

## 3. Discussion

Possible explanations for the etiology of an intracardiac mass could be: non-neoplastic masses such as mural thrombi, vegetation, abscess; or primary neoplasms of the heart and metastatic tumors. First of all we considered mural thrombus rather than a neoplasm since the echocardiography performed 6 months earlier was completely normal. Ventricular thrombi are usually easily differentiated from a tumor since they are associated with abnormal regional wall motion like the presence

**Figure 5.** Macroscopic appearance of resected left ventricular mass.



of dyskinetic or akinetic segments [4,5]. Exceptions to this is the apical thrombus noted in hypereosinophilic syndrome and lymphoma [6,7].

Thrombi usually have broad-based attachment or no attachment site at all, but occasionally there can be a thin stalk mimicking a myxoma. Our patient had no wall motion abnormalities predisposing her to ventricular thrombus formation and the mobile mass was pedunculated thereby attaching it to the interventricular septum; both of these observations indicated against thrombus but they were not enough to rule it out. In the absence of heart disease, mural thrombi most commonly occur in the right atrium rather than the ventricles and in the majority of the patients there is a coagulation defect [8,9]. Antiphospholipid syndrome is the most common coagulopathy associated with mural thrombus but a variety of other conditions are thought to be predisposing [10]. The patient was detected to have heterozygous Factor V Leiden mutation. Factor V Leiden is the most common inherited thrombophilia with an incidence of about 5%. Venous thrombosis risk is increased 50-80 times in homozygotes and 3-5 times in heterozygotes. There are some case reports relating Factor V Leiden mutation and intracardiac thrombosis, but most of the patients that have been reported in these literature have underlying malign disorders or accompanying systemic disorders like vasculitis, in addition to Factor V Leiden mutation [11-14]. Our patient was given intravenous heparin to see if any change would be observed in the size of the mass but there was no difference after one week so we partially excluded the diagnosis of a thrombus.

The type of tumor expected in the heart greatly varies by the location of the mass and age of the patient. If the mass is related to the ventricular chamber wall by a stalk or if there is a broad-based attachment, the most likely diagnosis would be a metastatic tumor, especially for the right ventricle, or an inflammmatory myofibroblastic tumor if thrombus is excluded. Still myxoma, lipoma, sarcoma or a hemangioma could be found in that localization [15].

Microscopic leukemic infiltration of the myocardium and pericardium is a rather frequent finding in patients with acute leukemia, but gross cardiac metastases have been very rarely reported [16]. A higher incidence of cardiac leukemia has been reported with AML than ALL.

Barbaric et al. reported a large echodense mass involving the anterior and lateral walls of the right ventricle at the initial presentation of acute B cell precursor ALL, which resolved with multi-drug chemotherapy [17].

Another report presented a patient with an isolated cardiac recurrence of ALL without any bone marrow or hematopoietic involvement two years after the bone marrow transplant. There was a large tumor in the right atrium with findings of right heart failure. This was the first report of isolated extramedullary relapse of ALL in the heart [18].

Our patient had been disease free for 8 months after stem cell transplant and we were informed by the haemotology department that she was in complete remission but an isolated extramedullary metastasis in the heart was still possible.

One point that kept us away from the diagnosis of leukemic infiltration was the localization of the tumor. About two-thirds of all cardiac metastases are found in the pericardium, one-third in the epicardium or myocardium, and only 5% in the endocardium. Lymphomyeloproliferative diseases are known to involve myocardium preferentially but not the endocardium as noted by Abraham et al. [19].

Fungal infections of the heart are uncommon and generally occur in immunocompromised patients. Although disseminated Aspergillus and Candida infections are common after cardiac transplantation, involvement of the myocardium is rare. In postmortem studies myocardial fungal involvement is characterized by diffuse myocardial abscess as well as coronary artery occlusion by fungal mycele or thrombus [20,21].

We considered if the mass could be an aspergilloma that formed during the time interval when our patient had been seriously immunocompromised. A case report about a patient who received autologous stem cell transplant for acute myeloid leukemia described a left ventricular aspergilloma, which was attached to ventricular septum and treated with antifungal therapy [22]. Tuberculoma or vegetations attached to the ventricular free wall were also possiblities, but the echocardiographic appearance was not constitutive with a vegetation since the mass

was quite smooth-appearing with a thin stalk. In addition there were no clinical signs or symptoms supporting infective endocarditis, and the skin and sputum tests for tuberculosis were negative.

Myxomas account for 30-50% of all primary tumors of the heart. 65% of cardiac myxomas occur in women, and 4.5-10% are familial [23]. A recent meta-analysis showed that 83% occur in the left atrium and 12% in the right atrium. Only 1.7% are in the left ventricle and 0.6% in the right ventricle [24]. In the literature there are case reports describing atypical localizations of myxomas, especially left or right ventricular myxomas that cause outflow tract obstruction [25-27]. Cardiac myxomas are generally pedunculated tumors with a fibrovascular stalk attaching to the subendothelial base.

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