Epithelioid Leiomyosarcoma with Primary Cardiac Spindle-Celled and is Emergency Surgery too Late?: Case Report

Primer Kardiyak Spindle Hücreli Epiteloid Leiomiyosarkoma, Acil Cerrahi İçin Gecikmiş Olabilir miyiz?

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ABSTRACT Primary cardiac leiomyosarcomas are rare and aggressive tumors with lethal prognosis. We present a patient who underwent an emergent cardiac operation three months following the initial misdiagnosis. The delay for extensive surgery was caused by the earlier magnetic resonance imaging findings suggesting the possibility of a benign mass and subsequent incisional biopsy results suggesting cardiac fibroma. Minimally invasive surgical, echocardiography or CT guided percutaneus needle biopsies may be useful for certain intrapericardial or mediastinal masses. However, the limited amount of tissue collected by these methods may not be available for a specific diagnosis and may misguide the clinicians. The ultimate diagnosis is based on typical pathologic features of the tumor which can be excised during open heart surgery. If there is a suspicion for a malignant intracardiac mass, even without cardiac symptoms, a prompt cardiac operation is the optimal choice for both diagnosis and treatment.

Key Words: Leiomyosarcoma; emergency treatment

ÖZET Primer kardiyak leiomiyosarkomalar nadir görülen, agresif seyirli ve ölümcül tümörlerdir. Bu yazıda, başlangıçta yanlış tanı konan ve ilk tanıdan üç ay sonra acil koşullarda operasyona alınması gereken bir hasta sunulmuştur. Kapsamlı bir ameliyatın gecikmesinin ve hasta için daha güç koşullarda yapılmasının sebebi, o dönemde yapılan manyetik rezonans görüntüleme kitlenin benign izlenimi vermesi ve iğne biyopsisi ile alınan yetersiz materyalin incelenmesinde fibroma tanısının konulmasıdır. Minimal invaziv cerrahi girişimle, ekokardiyografi veya bilgisayarlı tomografiyle yönlendirilen iğne biyopsisi ile mediastinal kitlelerden doku örneği alınabilinir. Ancak alınan doku miktarı kesin patolojik tanı için yeterli olmayabilir. Eğer malignite şüphesi olan bir intrakardiyak kitleyle karşı karşıya isek, kardiyak belirti olmasa bile, hem tanı hem de tedavi için hızlandırılmış bir operasyonun en iyi tercih olduğuna inanıyoruz.

Anahtar Kelimeler: Leiomiyosarkoma; acil tedavi

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Primary cardiac tumors are extremely rare tumors which are shown to have an incidence between 0.001 and 0.28 among all cardiac tumors in the autopsy studies.[1] Primary cardiac leiomyosarcomas represent 8% to 9% of all cardiac sarcomas and less than 0.25% of all cardiac tumours.[2] Primary cardiac sarcomas are highly aggressive lesions, and have an extremely poor prognosis with a median survival of approximately 12-16 months.[3] However, extensive resection and reconstructive surgery

followed by chemotherapy and/or radiotherapy appears to improve survival in patients with primary leiomyosarcomas.[4,5] As these tumors are highly malignant and spread very rapidly, early diagnosis and prompt surgery is essential for better palliation.[6]

CASE REPORT

We report a patient with rapidly growing primary leiomyosarcoma of left atrium, interfering with the mitral valve function, and obstructing both pulmonary vein openings and left atrial outflow tract. Sixty year-old female patient was hospitalized urgently because of signs of mitral valve obstruction and rapidly progressing pulmonary edema. Three months prior to this event, she had been hospitalized in another hospital for an episode of acute cholecystitis. Until that time, she was free of any cardiac or constitutional symptoms. She had no history of embolic phenomena. During routine tests and examinations for cholecystitis, signs of pericardial effusion had been discovered. Transthoracic echocardiography had revealed a left atrial mass on the epicardial surface. Magnetic resonance imaging (MRI) had disclosed a 4.2 x 5.9 x 7 cm epicardial mass infiltrating the left atrial and left ventricular free walls, and extending into the left atrium. Signal intensity of the tumor was isointense or hypointense relative to the myocardium on T2-weighed (cine true-FISP) images. During dynamic contrast enhanced sequences, the mass demonstrated less-nonenhancement with respect to the myocardium, suggesting a poorly vascularised fibrous tissue. There was notable pericardial effusion surrounding the heart. The mass was extending into the left atrium towards the mitral valve, but was not interfering with the function of the valve. Following these investigations, she had undergone a limited anterolateral thoracotomy for an incisional biopsy. The histopathologic examination was suggesting a cardiac fibroma. As she had had no major cardiac complaints, and a benign course of the disease confirmed by her physicians, she had refused further elective cardiac operation. She had been discharged home with no further cardiac workup.

On admission to our hospital, she had signs of pulmonary edema. Chest roentgenogram revealed cardiomegaly and bilateral pleural effusions. Transthoracic two-dimensional echocardiograhic findings were almost the same as the previous one with exception of severely impaired mitral valve function. A few hours following the admission, the patient was transferred to the operating room because of severe hypotension being unresponsive to medical therapy. At emergent surgery via median sternotomy, cardiopulmonary bypass was established by bicaval cannulation. There were massive pericardial and bilateral pleural effusions. Both the pericardium and the parietal pleurae were markedly thickened, and a hard mass was palpated in the left atrial roof and left ventricular basal region. Posterior pericardial cavity was widely invaded by the tumor, extending from left atrial appendage to the diagraphmatic regions of right ventricle and left atrium. The origin of the tumor seemed to be the basis of the left atrium. Inside the left atrium, the tumor invaded pulmonary vein openings and mitral annulus, limiting intracardiac blood flow. An aggressive tumor resection was performed, in order to allow normal blood flow through the left atrium. In addition, the left atrial tissue around the left atrial appendage was resected. This part of the atrium was reconstructed primarily. The patient was weaned from cardiopulmonary bypass without any haemodynamic compromise.

She had a routine intensive care unit (ICU) course and weaned from the respirator at 24 hours. There was no need for inotropes. Histopathologic examination showed an epithelioid and spindle-celled cellular tumour that invaded into epicardial fat and myocardium, and involved surgical resection margins. Most of the tumor cells showed minimal pleomorphism, exhibiting small nuclei and sparse cytoplasm. However, these cells were strongly positive for both desmin and smooth muscle actin.

Informed consent was obtained from patient before she was discharged.

DISCUSSION

Malignant cardiac neoplasms are classified by tissue type as mesenchymal (sarcoma), lymphoid (lymphoma), and mesothelial (mesothelioma).[2] Cardiac sarcoma represents the second most common primary cardiac neoplasm after myxomas.[4] However primary cardiac leiomyosarcomas are extremely rare malignant mesenchymal tumors with high degree of invasiveness. To our knowledge, our case is the second reported case from Turkey.[7] These tumors are highly aggressive and cause the death of the affected patient within 3 months to 1-year after diagnosis.[4] The tumor may progress so rapidly that within a month after the onset of symptoms, it may cause an outflow tract occlusion, requiring urgent surgical resection.[8] However, there are reports of survival up to 7 years with combined approach of early extensive surgery, medical and radiation therapy.[9] However, complete macroscopic resection is possible in only one third of the cases.[3] In contrary, cardiac fibroma is a congenital neoplasm that typically affects children.[4] However, approximately 15% of cardiac fibromas occur in adolescents and adults.[10] Cardiac fibromas may manifest as a discrete mural mass or focal myocardial thickening with nonenhancing areas at MRI imaging.[4] Cardiac fibromas may remain stable in size for years or may even regress.[11]

Two-dimensional echocardiography is currently the most widely used diagnostic procedure to detect heart tumors.[1,2] Gated cardiac MRI plays a significant role in the evaluation of cardiac masses and is of greatest value when echocardiographic findings are equivocal or suboptimal or when the lesion has an atypical location or appearance. MRI can also demonstrate with precision the tumoral mass and pericardial or great vessel extension as experienced in the case reported. These two diagnostic modalities are recognized to be correlative. However, iso-hypointensity of the mass relative to the myocardium on T2-weighted images and less-nonenhancement of the contrast relative to the myocardium were suggesting poorly vascularised fibrous tissue (Figure 1). Furthermore, histopathologic examination of the previous biopsy specimen collected at the time of left anterolateral thoracotomy confirmed a cardiac fibroma. Since the tumor

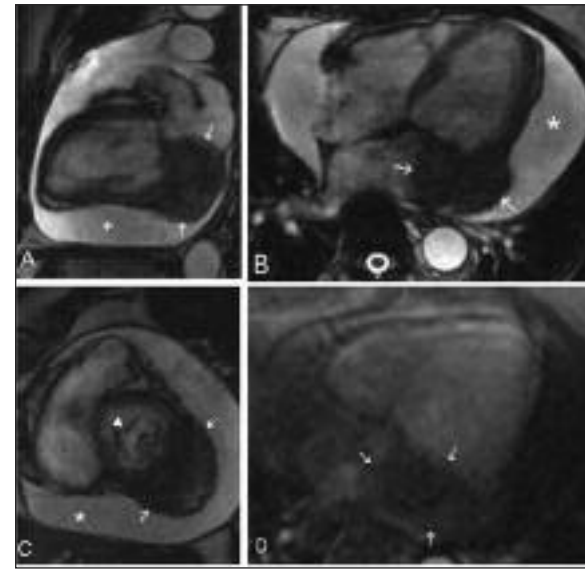

FIGURE 1: Magnetic resonans imaging showing the left atrial tumour with pericardial effusion (stars). **A)** Vertical long axis true-FISP. **B)** Horizontal long axis true FISP. **C)** Short axis true-FISP. **D)** Last image of the myocardial perfusion sequence. Iso-hypointense tumoral mass (arrows) invades the left ventricle free wall and left atrium, prodrude to the left atrial cavity. No contrast enhancement is detected **(D)**. It doesn't obstruct the mitral valve (arrow head) and causes massive.

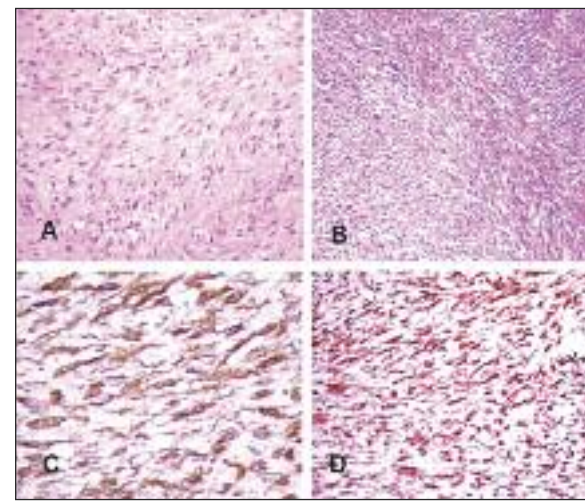

FIGURE 2: In some areas odematous fibrous tissue containing many capillaries were detected. Some vessels contained trombus and recanalisation mimicing organisation of a trombus. In some areas bland looking spindle cells arranged as intersecting bundels. Some spindle like cells showed little pleomorphism **(A)**. Immunohistochemically tumour cells express diffuse cytoplasmic vimentin, desmin **(B)** and smooth muscle actin. Mostly pleomorphic cells arranged in compact bundles. Giant cells were also seen. High mitotic rate and necrosis were detected **(C)**. In a small localized area histologic changes like biopsy material were also detected. Neoplastic cells express vimentin, smooth muscle alpha actin and desmin **(D)**. The final diagnosis was leiomyosarcoma.

enhancement on MRI and the subsequent incisional biopsy caused a misdiagnosis, an early elective surgery was abandoned. Unfortunately our patient presented a rapidly growing tumor. Three months later she required an emergency operation for intracardiac obstruction.

Minimally invasive surgical, echocardiography or computer tomograhpy (CT) guided percutaneus needle biopsies may be useful for certain intrapericardial or mediastinal masses. However, the limited amount of tissue collected by these methods may not be available for a specific diagnosis and may misguide the clinicians.[12] When examining a biopsy from a cardiac tumor, it must be kept in mind that tumors from this location may show heterogeneous morphological patterns as observed in our case. In the case of sarcomas histological classification continues to be confusing.[6] The biopsy material may not be representative of the condition or may cause misdiagnosis. Especially, as demonstrated in our case, fibroblast-rich areas or organizing thrombus may be misinterpreted (Figure 2). When a tumor with spindle cells forming bundles like fibromas or leiomyomas are detected in the heart, immunohistochemical evaluation with smooth muscle markers should be performed. Demonstration of smooth muscle cell derivation virtually confirms malignancy.

CONCLUSION

In summary early diagnosis and radical surgery provides the best chance for palliation of cardiac leiomyosarcomas. The ultimate diagnosis is based on typical pathologic features of the tumor which can be excised during open heart surgery. If there is a suspicion for a malignant intracardiac mass, even without cardiac symptoms, a prompt cardiac operation is the optimal choice for both diagnosis and treatment. Complete resection is best performed with total cardiopulmonary bypass and cardiac standstill. The time window between the diagnosis of an intracardiac mass and extensive surgery should be minimized and adjuvant therapy should be considered for a better outcome.

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