CASE REPORT: PRIMARY CARDIAC SARCOMA

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ABSTRACT

A 14-years-old female was admitted to the hospital with the chief complaint of having hemoptysis and progressive dyspnea. The imaging studies revealed pericardial, myocardial and mediastinal mass with bilateral lung nodules. The tumor could not be resected through a thoracotomy, only biopsies could be taken. The final diagnosis was cardiac sarcoma. The patient received chemotherapy and three days after that she died from severe hemoptysis and cardiac arrest.

INTRODUCTION

Primary cardiac neoplasms are rare. It is estimated that primary cardiac neoplasms are 100-1,000 times less prevalent than secondary neoplasms of the heart. Even among primary cardiac tumors, the majority is benign and most frequently are atrial myxomas. Sarcomas are the second most common primary cardiac tumor. Reported here is a case of primary cardiac sarcoma in a 14-years-old female.

CASE RERORT

A 14-years-old female was admitted for hemoptysis and progressive dyspnea especially on exertion. Two weeks prior to admission she had dyspnea and weight loss about 2 kg in 2 weeks. She had underlying thalassemia (Hemoglobin E trait). She had no history of asbestos exposure.

On physical examination, she had low grade fever (37-38 c°), dyspnea, and pulse rate was 100 beats/min. She had mild pale. Lung had fine crepitation both lower lungs. Heart showed PMI at 6th intercostal space, mid clavicular line, RV heaving, no thrill, regular rhythm, normal S1, S2, no murmur. JVP was normal.

The laboratory data revealed mild anemia and leukocytosis (Hemoglobin 8.3 g/dl, Hematocrit 25%, WBC 18,100/ml, platelet 211,000/ml). Three days sputum exam for AFB was negative. ESR was 39 mm/hr (0-20).

Electrocardiogram showed inverted T at II, III, AVF, V1-V5 leads.

Chest radiograph showed widening of cardiac shadow, bilateral reticulonodular infiltration with round mass with well defined border at left lower lobe (Figure 1A,1B).

Echocardiogram demonstrated large inhomogeneous mass attach to the right atrium without obstruction during cardiac cycle, suggested cardiac tumor. No pericardial effusion was detected (Figure 2).

A computed tomography scan of the chest revealed heterogeneous enhancing mass involving right atrium, right ventricle, pericardium and mediastinum, representing malignant pericardial tumor with myocardial and mediastinal involvement or myocardial tumor with pericardial and mediastinal involvement. Multiple pulmonary nodules were detected, suggestive of pulmonary metastases (Figure 3).

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The patient was sent to performed percutaneous transpericardial window biopsy but unfortunately it failed to demonstrate the pathology.

Then the patient underwent surgery to obtain biopsy or excision of the tumour. Open thoracotomy was performed. Huge semisolid well-circumsribed tumor at anterior mediastinal space with multiple palpable nodules, varying in size of pulmonary nodules and moderate right pleural effusion were demonstrated. The tumor could not be resected, only biopsy was performed.

Tissue from anterior mediastinum showed multiple pieces of brownish tissue in macroscopic masses measuring 3x2x1 cm in aggregation. The histology revealed sheets of tumor cells which are large polygonal as well as rather elongated cells (Figure 4A). Those tumor cells contained enlarged nuclei with prominent nucleoli. No gland or keratin formation was noted. Immunohistochemical studies revealed the tumor cells were immunoreactive for vimentin, but not with CK, CD3, CD45, CD79a, PLAP, Tdt, or CEA (Figure 4B). The final histopathologic diagnosis was malignant lesion, suggestive of sarcoma.

All treatment options were extensively considered and discussed. With the diagnosis of primary cardiac sarcoma with pulmonary metastasis, she was put on palliative chemotherapy. Three days after chemotherapy she died from severe hemoptysis and cardiac arrest.

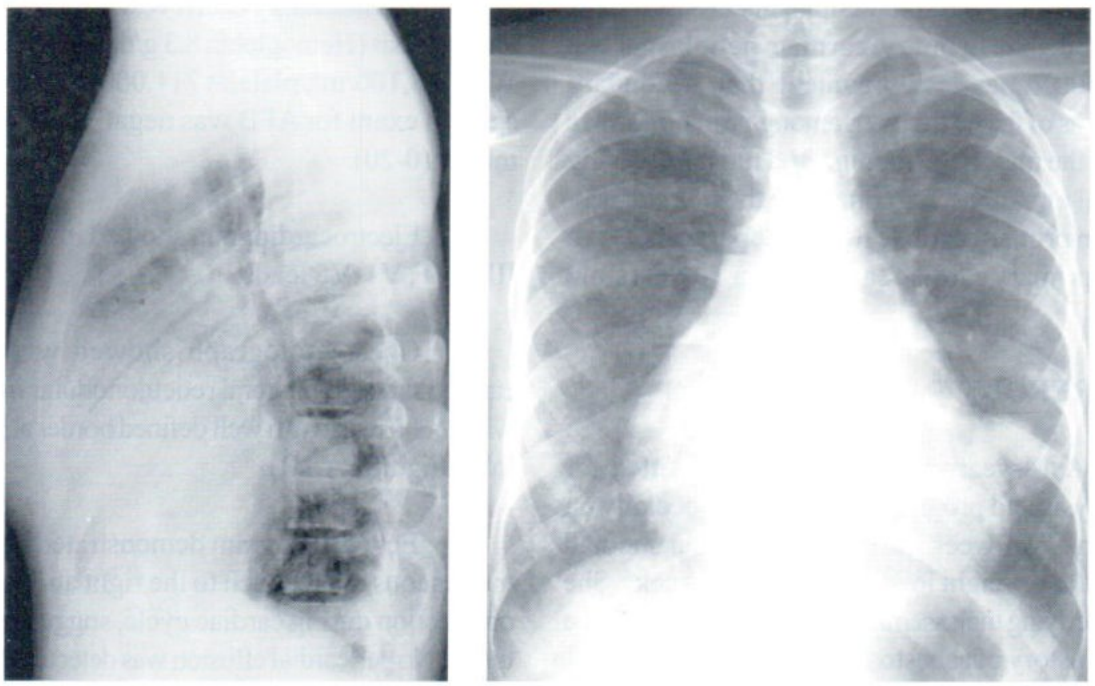

Fig.1A and 1B Chest radiograph (PA, upright and lateral views) showed widening of cardiac shadow, bilateral reticulonodular infiltration with round mass at left lower lobe.

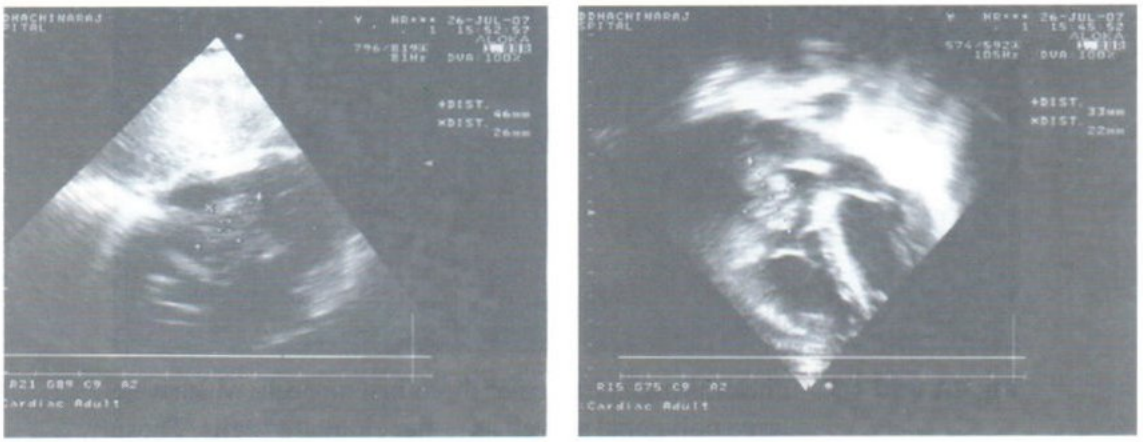

Fig.2 Echocardiogram demonstrated large inhomogeneous mass attach to right atrium without obstruction during cardiac cycle, suggestive of cardiac tumor. No pericardial effusion was detected.

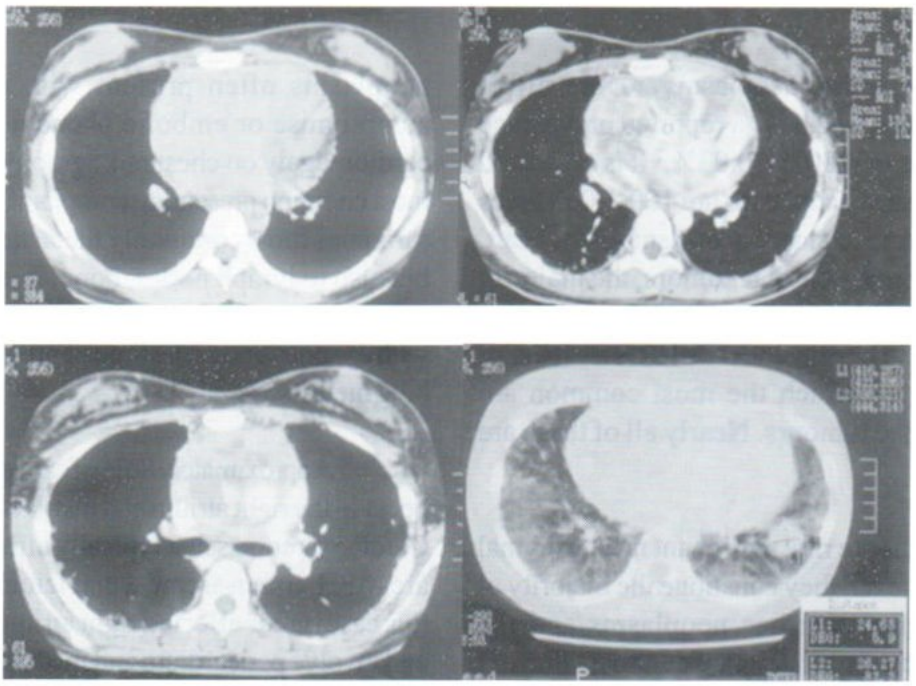

Fig.3 (3A; NECT, 3B and 3C; CECT, 3D; lung window) CTchest revealed heterogeneous enhancing mass involving right atrium, right ventricle, pericardium and mediastinum, which extended from SVC recess into pericardial cavity. Multiple nodular lesions seen in both lungs suggestive of pulmonary metastases.

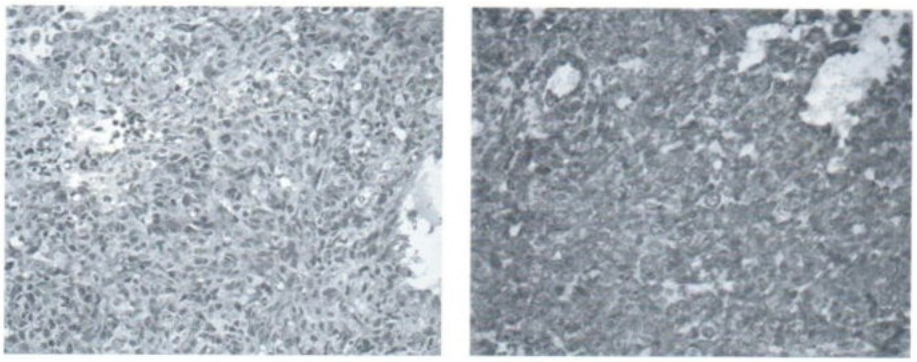

Fig.4A and 4B Photomicrograph revealed sheets of tumor cells which were large polygonal as well as rather elongated cells. Those tumor cells contained enlarged nuclei with prominent nucleoli. No gland or keratin formation was noted (4A). (H&E x40) Immunohistochemical study; the tumor cells revealed cytoplasmic staining with vimentin antibody (4B). (x40)

DISCUSSION

Primary cardiac neoplasms are rare, affect patients of all ages, and have a reported prevalence in autopsy series of 0.001%-0.03%.[1] It is estimated that primary cardiac neoplasms are 100-1,000 times less prevalent than secondary neoplasms of the heart. Even among primary cardiac tumors, the majority (approximate 75%) is benign and most frequently are atrial myxomas.[2] The remainder includes a variety of other tumours, of which the most common are malignant cardiac tumours. Nearly all of these are sarcomas.[3]

Sarcomas are rare malignant mesenchymal neoplasms; however, they constitute the majority of primary malignant cardiac neoplasms and the second most common primary cardiac tumor.[4] Primary cardiac sarcomas, by definition, are confined to the heart or pericardium at the time of diagnosis with no evidence of extracardiac primary neoplasm. Although all types of sarcomas affect the heart, the most common cell types are angiosarcoma (37% of cases), unclassified or undifferentiated sarcoma (24%), malignant fibrous histiocytoma (MFH) (11%-24%), leiomyosarcoma (8%-9%), and osteosarcoma (3%-9%).[5]

Patients affected with cardiac or pericardial neoplasms often present with cardiovascular compromise or embolic phenomena and exhibit cardiomegaly on chest radiography. Dyspnea is the most common presenting complaint. Primary cardiac sarcomas most commonly metastasize to the lungs but also to lymph nodes, bone, liver, brain, bowel, spleen, adrenal .[1,4,7] As in this case, she presented with dyspnea and at the time of presentation, she had lung metastases.

Approximately 80% of cardiac angiosarcomas occur in the right atrium and involve the pericardium. Cardiac sarcomas that typically affect the left atrium are MFH, osteosarcoma, and leiomyosarcoma.[5] In this case tumor affected right atrium, right ventricle and involve pericardium and mediastinum.

The pathologic features of cardiac sarcoma are extremely varied. The vast majority of cardiac sarcomas are large, invasive masses at the time of diagnosis. The most common cell types are angiosarcoma , unclassified or undifferentiated sarcoma and malignant fibrous histiocytoma (MFH)[5,6] Unclassified and undifferentiated sarcomas are lack of specific histologic, ultrastructural ,or immunohistochemical features.[1,4] In this case histologic type was unclassified sarcoma.

The most common radiographic abnormality in patients with cardiac sarcoma is cardiomegaly. Other findings include heart failure, pleural effusion, focal cardiac mass, pulmonary consolidation, and pericardial effusion.[4,5]

Echocardiography remains the initial imaging modality of choice for evaluating cardiac masses. It accurately delineates cardiac anatomy in multiple aspects. However, echocardiography has limited capability to demonstrate tumor infiltration and cannot delineate mediastinal and extracardiac involvement.[8] In this case, echocardiogram showed large inhomogeneous mass attach to the right atrium without obstruction during cardiac cycle, suggestive of cardiac tumor.

Because CT is beneficial in the evaluation of cardiac sarcomas as it demonstrates the broad-based tumor attachment; myocardial, pericardial, and mediastinal invasion; as well as extension into the great vessels and pulmonary metastases. In this case, CT scan revealed heterogeneous enhancing mass involving right atrium, right ventricle, pericardium and mediastinum, which may be pericardial tumor with myocardial and mediastinal involvement or myocardial tumor with pericardial and mediastinal involvement. Multiple nodular lesions were suggestive of pulmonary metastases.

MR imaging typically demonstrate large, heterogeneous, broad-based masses that frequently occupy most of the affected cardiac chamber or multiple chambers. Pericardial and extracardiac invasion, valvular destruction, tumor necrosis, and metastases are frequently seen and are all characteristic features of malignant lesions. Pericardial invasion is characterized by disruption, thickening, or nodularity. In this case MRI was not performed.[9,10]

Primary cardiac sarcomas are highly aggressive lesions that are uniformly fatal. The mean survival of affected patients is from 3 months to 1 year.[1,4] The patient outcome is almost invariably poor due to unfavorable location, their rapid growth and early metastasis. However, the mainstay of treatment is surgical resection because the surgical resection can substantially alleviate symptoms and yield a modest improvement in survival.[2,8] Even after complete tumor excision, however, local recurrence and metastatic disease occur frequently and early, usually within 1 year.[4] Chemotherapy and radiation therapy have not proved to be beneficial for the treatment of affected patients.[11,12] Death in these patients usually results from postoperative complications, cardiopulmonary failure from progressive tumor growth, and metastatic disease.[4] Heart transplantation has been performed in some patients with unresectable cardiac sarcoma with satisfactory results.[11,13] Success of molecular targeted therapy in gastrointestinal stromal tumor, Kapposi's sarcoma and angiosarcoma, has given an exciting new prospect for treatment of cardiac sarcoma. However, in this case palliative approach was preferred.

CONCLUSION

Primary cardiac sarcoma is a rare tumor and has a very poor prognosis due to very aggressive behavior, most often presents with advanced disease and early metastasis. The initial diagnosis is usually suggestive at echocardiogram, but identification of mediastinal invasion and extracardiac metastasis is best achieved with CT and MRI. The mainstay of treatment is surgical resection. However, local recurrence and metastatic disease occur frequently and early. Chemotherapy and radiation therapy have not proved beneficial for the treatment of affected patients. So the mean survival of affected patients is from 3 months to 1 year.

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