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CASE REPORT

A Very Rare Malignant lesion in the Right Heart almost Occluding Whole of Right Atrium and Part of Right ventricle – Case report

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Abstract:

Primary cardiac tumors are rare, with an incidence of 0.056% according to autopsy reports. The most common type is myxoma, while other types, including sarcoma, lipoma, papillary fibroelastoma, rhabdomyoma, fibroma, hemangioma, teratoma, lymphoma and mesothelioma also occur. Primary cardiac tumors usually cause embolization, pericardial effusion and arrhythmia, leading to heart failure. Only 10% of primary cardiac tumors are malignant, approximately 95% of which are sarcomas, while the remaining 5% are cardiac lymphomas and mesotheliomas. This report documents the case of a 44-year-old male with primary cardiac lymphoma.The definitive diagnosis is dependent on Histopathology and Immunohistochemistry. Timely treatment with chemotherapy can be effective.

The goals of this article are to show the difficulties of diagnosing and treating this disease, the role of cardiac surgery in its treatment and to raise awareness of this disease.

Key words: *Heart neoplasm, non Hodgkin lymphoma, primary cardiac lymphoma*

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Introduction:

Primary cardiac lymphoma (PCL) is a rare type of nonHodgkin's lymphoma (NHL) that solely involves the heart, the pericardium, or both and accounts for less than 0.01% of all cardiac tumors.[1–3] The clinical presentation of PCL varies depending on the location, size, and degree of invasion[4]. The rising incidence of PCL is thought to be due to advances in diagnostic radiologic technology, greater exposure to environmental toxins, and a larger number of immunocompromised individuals.[5] Here, we present a case of primary cardiac lymphoma.

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Cardiac tumors are among the least investigated tumor types in oncology. Primary cardiac tumors are rare with an incidence of 0.056% according to autopsy reports.[6] Approximately 90% of primary cardiac tumors are benign, the most common type of which is myxoma, while other types, including lipoma, papillary fibroelastoma, rhabdomyoma, fibroma, hemangioma, teratoma, sarcoma, lymphoma and pericardial mesothelioma also occur. The remaining 10% of primary cardiac tumors are malignant, 95% of which are sarcomas and 5% of which are cardiac lymphomas and mesotheliomas.[7] Primary cardiac lymphoma (PCL) is defined as non Hodgkin lymphoma (NHL) involving only the heart and/or pericardium (strict criteria), or as NHL presenting with cardiac manifestations, particularly when the bulk of the tumor is found in the heart (loose criteria).[8] It is estimated that PCLs account for 1.3% of primary cardiac tumors and 0.5% of all extranodal NHLs.[9] During 1949-2009, only 197 cases of PCL were reported in the literature. By contrast, secondary cardiac involvement of lymphomas is more common, with an incidence of 9-24% in disseminated and terminal-stage NHL cases.[10]

Primary cardiac lymphoma (PCL) constitutes 5-6% of primary cardiac malignant neoplasms.[11,12] Because of its rapid progress, late diagnosis due to its non-specific symptoms and variable patient responses to treatment, the prognosis for patients with PCL remains poor. Complete remissions have been reported, although the longest period of survival has not exceeded 144 weeks.[13] Approximately 80% of patients live less than 12 months after diagnosis.[14]

Case Report:

A 44-year-old Bangladeshi male patient presented with exertional dyspnea for several months. The patient denied any history of systemic disease and physical examination showed no remarkable abnormalities. Laboratory examination showed creatinine levels of creatinine (1.3 mg/dl; normal range, 0.7 1.5 mg/dl for male and 0.5 1.2 mg/dl for female), lactate dehydrogenase (LDH 603 U/l; normal range, 95 213 U/l), aspartate aminotransferase (83U/l; normal range, 5 35 U/l) and alanine aminotransferase (33 U/l; normal range, 0 40 U/l). Chest X-ray (CXR) showed cardiomegaly with mild left pleural effusion. Electrocardiography (ECG) showed sinus bradycardia. Transthoracic echocardiography (TTE) revealed a left ventricular ejection fraction (LVEF) of 65%, and alarge right atrial myxoma with tricuspid regurgitation. CAGwas done which revealed: Single vessel disease (SVD) recommended for CABG with Excision of RA myxoma. After establishment of cardiopulmonary bypass right atrium opened. A huge myxoma like mass was found in RA with broad base over the IAS near triangle of Koch and commissure between septal leaflet and anterior leaflet of tricuspid valve.Surgical resection was performed. RV cavity and pulmonary valve was checked for any residual tumor fragment. Thorough wash given in RV cavity with saline. Patency of tricuspid valve was checked. Good leaflet coaptation present. RSVG was anastomosed to the LAD.

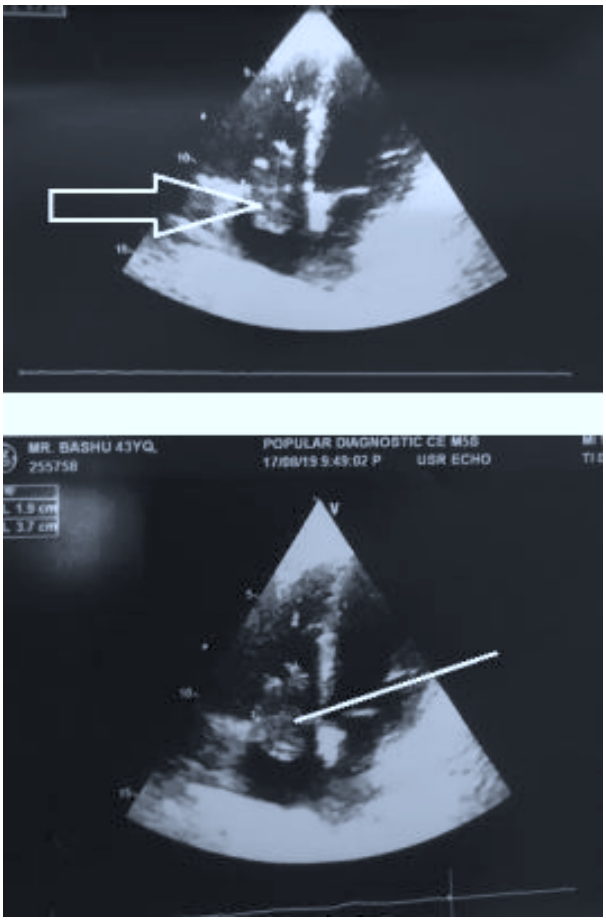
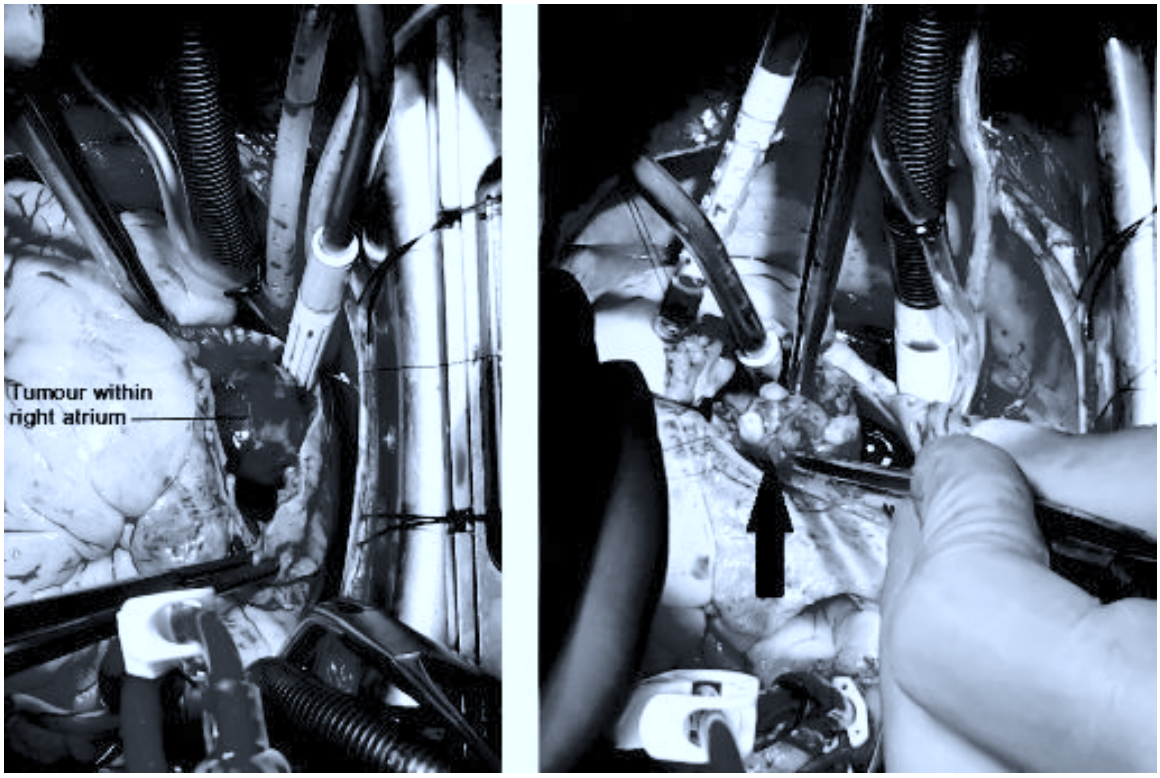


Fig.-1*: Transthoracic echocardiography showing a echogenic mass (3.7 x 1.9) cm within the right atrium (white arrow and line) extending into right ventricle.*



(a) (b)

Fig.-2(a &b): *RA-tomy showing an irregular mass with variegated appearance arising from right atrium (Black line & arrow).*

Histologic sections revealed anintermediate/diffuse proliferation of atypical large lymphocytes. Frequent atypical mitosis and prominent starry sky pattern are evident. Immunohistochemical studies revealed that the malignant cells stained positively with antibodies directed against CD20, BCL2, C-Myc, MUM1.Histomorphological and IHC features are compatible with diffuse large B-Cell Non-Germinal Centre type.The final diagnosis was primary cardiac lymphoma, diffuse large B-cell type. The patient was referred to hematologist for further management.

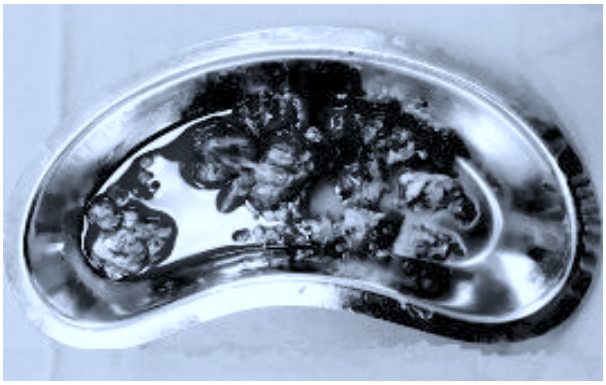


Fig.-3: *Resected cardiac tumor.*

Discussion:

The major clinical features of PCL include dyspnea, heart failure, precordial pain, life-threatening arrhythmia due to tumor invasion of the conduction system or irritation of myocardium, pleural effusion, pericardial effusion and shock due to cardiac tamponade or obstruction of blood flow.[15] Constitutional symptoms, including fever, chills, sweats and weight loss, are also common. In some PCL cases, patients may develop pulmonary or cerebral embolism. The most common ECG abnormalities include atrial arrhythmias and AV block, with up to 61% of patients experiencing complete AV block. Other ECG features are right bundle branch block, inverted T waves, low voltage and life threatening ventricular tachyarrhythmia.[16] CXR usually reveals cardiomegaly or pleural effusion. TTE has a sensitivity of 55 60% for primary cardiac tumors and TEE has a sensitivity of 97 100%. The sensitivity of cardiac magnetic resonance imaging is superior to that of CT (90 92 vs. 71 73%).[17] In 92% of PCL cases reported, the right heart chambers were involved, predominantly the RA.

By contrast, in only 7% of cases, the left heart was involved without right heart invasion. In 25% of PCL cases, the superior vena cava was also affected. Diffuse large B cell lymphoma is the most common sub type of PCL (113 cases reported); the remaining sub types include Burkitt's lymphoma, T cell lymphoma, small lymphocytic lymphoma and plasmablastic lymphoma.[18] At present, no definite guidelines exist for the management of PCL. Early systemic chemotherapy appears to be the only effective therapy. The major regimen is the same as that for other types of NHL, namely cyclophosphamide/hydroxydaunorubicin/oncovin/ prednisone (CHOP) and since 2001, CHOP + rituximab. It is difficult to perform a complete surgical resection of PCL, which provides no survival benefit. Chemotherapy followed by radiotherapy for PCL may enhance survival, although its efficacy remains to be determined[19]. The overall response rate of patients with PCL to chemotherapy is 79% and the complete remission rate is 59%. The median overall survival of patients with PCL is ~12 months. Poor prognosis[19] is associated with extra-cardiac disease, immunocompromised status, LV involvement and, notably, absence of arrhythmia.[20] The pathology of diffuse large B-cell lymphoma also represents the most common cell type of PCL. Primary cardiac tumors are rarely encountered by clinical physicians and PCL is even rarer. Although the development of modern imaging technology has improved the detectability of this disease, invasive tissue biopsy and awareness of clinicians of the disease are required for early pathological diagnosis, which is essential for effective treatment. Despite the poor overall survival of affected patients and limited current knowledge regarding specific treatments, early systemic chemotherapy yields a high response rate and improves the possibility of survival. Lymphoma should be considered by clinicians if cardiac tumors are encountered. Although PCL is associated with poor prognosis and life-threatening complications, timely and appropriate treatment can be beneficial.

Conclusions:

In conclusion, prompt diagnosis and treatment management maximize prolonged survival in PCL. Therefore, the appropriate choice of diagnostic procedure influences the patient's prognosis. Unfortunately, the range of modern diagnostic approaches may be seriously limited in an advanced stage of PCL and when a patient's condition on admission is critical.

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