

Primary cardiac B cell lymphoma

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ABSTRACT

Primary tumors of the heart are very uncommon and approximately 25% of them are malign. Primary cardiac lymphomas account for approximately 4% of the primary cardiac tumors. Both B- and T-cell lymphomas have been reported. We report the case of a 75-year-old male who was referred to our hospital complaining of fatigue, dyspnea and swelling in the abdomen. Transthoracic echocardiography revealed a mobile 5.0×4.3 cm mass in the right atrial cavity impinging tricuspid valve. Surgery was performed and the mass was resected. The pathological and histological examination of the removed mass showed diffuse large B-cell lymphoma.

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Introduction

Primary cardiac lymphomas (PCLs) (which, by definition, arise from the heart and/or the pericardium without evidence of extracardiac involvement) are extremely rare and account for about 1% of the primary cardiac tumors and 0.5% of the extranodal lymphomas [1, 2]. Therewithal, disseminated lymphoma with cardiac involvement can occur in up to 20% of patients with lymphoma [2]. Recently, the incidence of PCLs has been increasing since they have been diagnosed as a manifestation of immunodeficiency states and with the help of multiple diagnostic modalities have been developed. Clinical and anatomic features of PCL remain uncertain. There is no standardized therapy for PCL. Tumor resection and standard chemotherapy combination regimens are widely used as therapeutic modality. In this article, we report a 75-year-old man with large cardiac mass who

was diagnosed primary cardiac extranodal diffuse large B cell lymphoma.

Case Presentation

A 75-year-old immunocompetent man with coronary artery disease medical history presented a 1-month history of dyspnea on exertion, shortness of breath and swelling in the abdomen. An electrocardiogram revealed sinus rhythm without any feature of myocardial ischemia. Transthoracic echocardiography disclosed a large intracardiac mass (5.0×4.3 cm), which extended from the right atrium, impinging tricuspid valve, a moderate pericardial effusion, and a preserved ejection fraction (Figure 1A). After then, the patient underwent transesophageal

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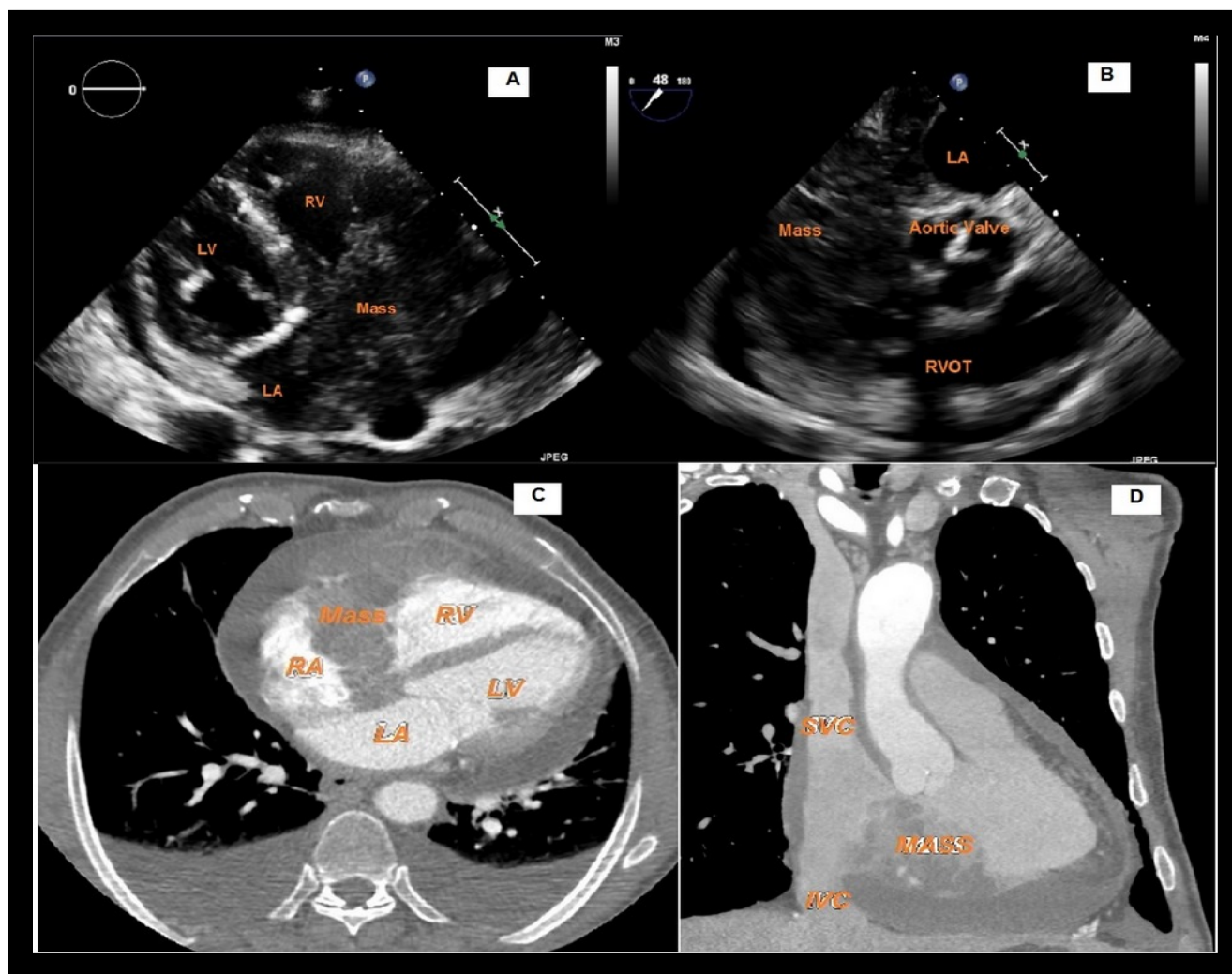


Figure 1. Modified apical four chamber (A) and short axis (B) echocardiographic view showing right atrial mass; transverse (C) and sagittal plane (D) of tomographic view showing cardiac mass. IVC = inferior vena cava, LA = left atrium, LV = left ventricle, RA = right atrium, RV = right ventricle, RVOT = right ventricular outflow tract, SVC = superior vena cava

echocardiography which confirmed the mass, arising from the right atrium, associated invasion of the right ventricle and the interatrial septum, showed causing mild tricuspid valve obstruction and a moderate pericardial effusion (Figure 1B). A contrast cardiac computed tomography (CT) showed a large infiltrating mass (11×7×4.5 cm sized), localized in the right atrium, arising from posteroinferior wall of the right atrium and diffuse infiltration of almost the full of the right atrium, contiguous to the right ventricle, extending to the pericardium from lateral wall of the right atrium, giving early and delayed heterogeneous contrast enhancement and involving the proximal segment of right coronary artery (Figure 1C and 1D). Presence of extracardiac involvement was excluded with the global assessment of the chest and abdomen with CT and scintigraphy. This case was discussed in the multidisciplinary meeting and decided for resection of the mass, because of mechanical

complication as tricuspid valve stenosis. A median sternotomy was performed, the mass was excised with the use of cardiopulmonary bypass with moderate systemic hypothermia, deep topical hypothermia, and cardioplegic cardiac arrest. A right atriotomy was employed for resection of mass. Right heart mass was partially resected. Right atrial mass was resected however the right ventricular mass resected partially because the mass has completely invaded the right ventricular free wall. The tricuspid valve was normal function and had not mass invasion, so that tricuspid valve was protected. The pathologic examination of removed cardiac mass had reported as below after immunohistochemistry study: intense cytoplasmic staining with LCA(+), CD79a(+), and Bcl-2(+), intense membranous staining with CD20 (+), nuclear staining in 40-60 % with Bcl-6(+), and focal staining with Vimentin(+) and CD5(+). Staining with CK19, CD31, CD10, Cyclin D1(Bcl-1), EMA, CD21

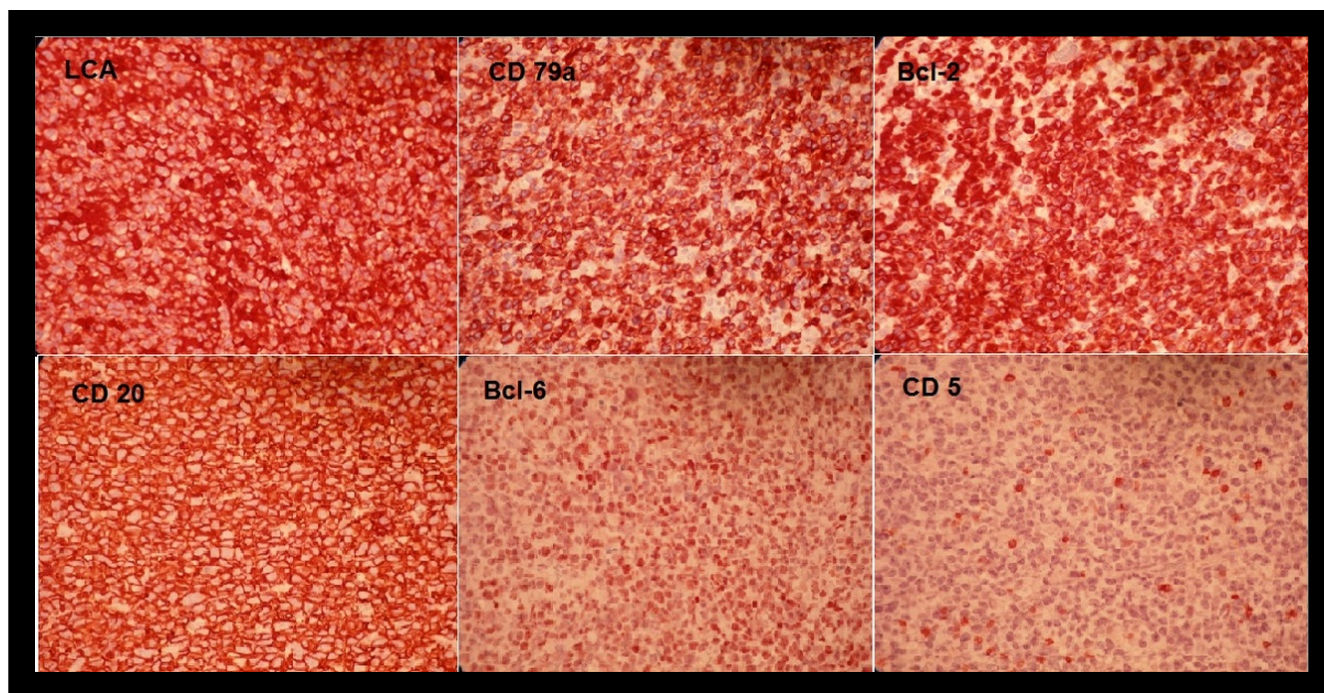


Figure 2. In the immunohistochemical study, proliferating cells were stained diffuse cytoplasmic with LCA, CD79a and Bcl-2, stained diffuse membranous with CD20, stained nuclear with Bcl-6 and stained focal with CD5.

and CD30 were not demonstrated. In the light of immunohistochemistry evaluation, the final pathologic report confirmed the diagnosis of diffuse large B cell lymphoma. We planned consulting with hematology service for chemotherapy. Unfortunately, the patient died at postoperative 7th day due to post-surgical cardiac complications.

Discussion

PCLs are extremely rare and primary lesion arises from the heart or the pericardium. Therewithal, myocardial or pericardial invasion of systemic lymphomas could be seen in up to 20% along the course of the disease [3]. The most common sub-type of PCL was diffuse large B-cell lymphomas and PCL are mostly right-side involvement [4]. The fundamental characteristics of PCL included clinical, imaging and pathologic features, diagnosis and treatment strategies remain being challenging. PCL patients present different and non-specific symptoms such as dyspnea, edema, arrhythmia, cardiac tamponade, palpitations and heart failure [5]. In our case most relevant symptom was dyspnea. Most of available literature about PCL consist of case reports [6]. Carras *et al.* [3] reported on the largest monocentric series of patients with PCL analyzed

from initial symptoms to pathological diagnosis and treatment with chemotherapy. They examined a total of 13 patients. No patient was immunocompromised. Lymphoma aroused from the right atrium at 10 patients (77%). They made pathological diagnosis (diffuse large B-cell lymphoma in 12 cases and Burkitt in 1 case) on cardiac surgical biopsies and by intravascular procedure. All patients received first-line chemotherapy and complete response rate was 62%. Recurrences occurred in 55% of patients, mostly at extracardiac extranodal sites. During 1975-2017, only 249 cases of PCL were reported in the literature. Diffuse large B-cell lymphoma is the most common sub-type of PCL; the remaining sub-types include Burkitt's lymphoma, T-cell lymphoma, small lymphocytic lymphoma and plasmablastic lymphoma [4]. Unfortunately, at present, no definite guidelines for the management of PCL. Complete surgical resection of PCL provides no survival benefit. Early systemic chemotherapy appears to be the only effective therapy. The major regimen is the same as that for other types of non-Hodgkin lymphoma, namely cyclophosphamide/ hydroxydaunorubicin/ oncovin/ prednisone (CHOP) and since 2001, CHOP + rituximab. General 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 associated with poor prognosis [4].

Transesophageal echocardiography is the firstly choice diagnosis tool for PCL. Transthoracic and transesophageal echocardiography have a sensitivity of 55-60% and 97-100%, respectively for primary cardiac tumors. Cardiac magnetic resonance imaging is superior to CT which are enhanced techniques visualize morphology and tumor localization [6]. Usually location of PCL is atrial and infiltrating to atrial or ventricular walls. The earliest sign of PCL can be sometimes pericardial thickening and effusion which is also commonly seen in other diseases [7].

Definite diagnosis of PCL is biopsy of cardiac mass. Biopsy can be taken by intracardiac or surgically. Patients with pericardial effusion, cytological analysis of pericardial fluid also make the diagnosis. Treatment is usually chemotherapy with combining surgery if necessary. Surgery alone has no effect outcome [8]. Surgery can enable time for chemotherapy and resolve hemodynamics problems due to obstructions [9].

Conclusions

Although PCL is associated with poor prognosis and life-threatening complications, we believe that, timely and appropriate treatment can be beneficial, and more effective treatment options will update with clinical therapeutic regimens.

Informed consent

Written informed consent was obtained from the patient's family for the publication of this case report.

Conflict of interest

The authors declared that there are no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

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