Cardiac Lymphoma Revealed by Atrial Mass
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Abstract
Primary cardiac lymphoma (PCL) represents a very exceptional clinical entity whose treatment is based on chemotherapy. In this case report, the authors highlight the key role of cardiac surgery in the diagnosis of PCL in a 66-year-old female patient. She was given chemotherapy and so far was on remission.

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Keywords: Lymphoma, Cardiac mass.

Introduction:
Primary cardiac lymphoma (PCL) is the rarest cardiac primitive tumor and represents a very exceptional clinical entity.1,2 The most common histological type of PCL is large B cell lymphoma.3,4 PCL is fatal, unless it is diagnosed and treated in time.5 A case of a patient diagnosed with PCL in our department is reported in this work.

Case report:
MR, a 66-year-old female patient has been suffering from dyspnea and occasional fever associated with cachexia for one year. She had no previous medical history. The cardiovascular risks were smoking, hypertension and dyslipidemia. The clinical examination found a low body mass index 19 Kg/m² and body surface area of 1.37 m². There was no other abnormal finding. Biologically, there was inflammatory syndrome as the CRP was 137 mg/L and white blood cell at 11000 /dL. The renal and hepatic functions were normal. ECG found sinus rhythm and the chest X-Rays, a pleural effusion. Transthoracic cardiac echocardiography (Fig 1) showed a mass located at the infero-lateral wall of the right atrium. It was extended towards the coronary sinus and moderate pericardial effusion could be seen.

The patient was admitted in the department of cardiology for pericardial and pleural drainage. Then a sample of the liquid was taken for analysis and other investigations were prescribed. CT scan revealed a large speculated mass whose size was 71x32 mm and whose appearance suspected a lymphoma. Magnetic resonance imaging (Fig 2) showed thoracic mass with iso-intense signal in T1, T2 and hyper-intense STIR signal at the right atrium extended to atrio-ventricular groove. This mass that presented a contrast but no early hypervascularisation was compatible with a lymphoma. Mass biopsy was decided by the multidisciplinary therapy team. The patient then underwent inferior ministernotomy under general anesthesia. The pericardial liquid was sanguineous; the right atrium was dilated by a stone-shaped mass from inside and the pericardial space infiltrated. Many biopsies of the pericardium and the area of atrio-ventricular groove were done and the sternum was closed. Postoperative course was uneventful. Histology concluded to large B cell lymphoma. He was given R-CHOP chemotherapy (rituximab and cyclophosphamide, doxorubicin, vincristine, and prednisone). Cardiac MRI after the 4th cure of the chemotherapy showed 87% decrease in the mass volume that now sized 32x15 mm (Fig 3). Then, the cancer MDT decided to continue with 4 supplementary and final cures. Since then, the patient is on remission and is asymptomatic.

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Discussion:
To the best of our knowledge, this case of PCL is the only one that is been diagnosed in our department for more than a decade. Indeed, this cancer represents only 1.3% of all cardiac tumors. It can be diagnosed at any time of the life time. However, in one the largest series ever (6 cases) from Fuzellier et al,7 the mean age of diagnosis was 62 years old and 2/3 of patients were men. PCL may be asymptomatic for a relatively long time depending on the site where it starts. It must be distinguished from cardiac extension of non-Hodgkin lymphomas that are more common and in which, disseminated complications are reported in 9 to 27% of cases. Like our patient, the most common type of this tumor is diffuse large B cell lymphoma. Usually, right atrium and right ventricle are involved. PCL is fatal, unless it is diagnosed and treated in time. Cardiac surgery was efficient method to biopsy the mass and subsequently histology. The most effective therapeutic method is chemotherapy which is given according to many protocols. Addition of rituximab to the protocol improves the remission time. As described in our patient, many authors have found that it can consistently reduce the tumor volume. Under some circumstances, particularly if the reduction in the obstructive tumors volume is unsatisfying, surgery may be required to perform atrial resection. Some authors have reported even Fontan procedure to bypass the right-heart. In our patient, the post chemotherapy MRI on month 3 showed effective decrease in the size of the mass. Surgery appeared to be non-beneficial in our case so the plan was to continue with chemotherapy and sub consequently follow-up.

Conclusion:
Cardiac surgery is an effective method to biopsy cardiac masses for histology. Rarely, the diagnosis of these masses are cardiac lymphoma. Chemotherapy remains the key treatment to get a relatively long remission period.

Conflict of Interest - None.

References:

