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Commentary Open Access

Commentary: Primary cardiac lymphoma: Two cases and a review of literature

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Abbreviations

PCL – Primary cardiac lymphoma; TTE – Transthoracic echocardiogram; TEE – Transesophageal echocardiography; CT-computed tomography; MRI – Magnetic resonance imaging; PCTVB – Percutaneous transvenous biopsy

Introduction

Primary cardiac lymphoma (PCL) is a rare malignant potentially lethal disease, which only involves the heart. PCL is diagnosed only in 1.3 % of heart tumor patients. PCL belongs to the non-Hodgkin extranodal lymphomas group and comprise 0.5 % of them. To date about 200 cases are published and the most comprehensive review, which includes 197 cases, is written by Petrich et al. The most common histological type of PCL is diffuse large B cell lymphoma[1–3]. In our article entitled "Primary cardiac lymphoma: Two cases and a review of literature", we have described two patients that were treated at our center[4]. Both of the patients were diagnosed with PCL. Both cases were confirmed histologically to be diffuse large B-cell lymphoma.

While treating our patients and later, when we wrote our article, one of the major difficulties was the fact that there are no unified criteria when this type of tumor can be called PCL. It can be diagnosed if there are no evidences of extrapericardial involvement found during comprehensive clinical examination and instrumental assessment or during an autopsy[5,6].

PCL has no specific symptoms. The patients can present with constitutional complains (fever, weight loss, chills, excessive perspiration), respiratory distress, arrhythmia, superior vena cava syndrome, heart failure, restrictive cardiomyopathy[1,3,7,8]. The lack of specific symptoms makes the diagnosing process more difficult. The patients need to be examined thoroughly. A comprehensive clinical examination is needed. Instrumental assessment should include transthoracic echocardiogram (TTE), computed tomography (CT), though sometimes transesophageal echocardiography (TEE) and heart magnetic resonance imaging (MRI) may be needed. Both CT and MRI are useful in assessing extra pericardial involvement, while TTE is used to asses patients hemodynamic status[9]. Even after thorough examination final diagnoses can be made only after performing a histological examination of the tumor. Usually a least invasive method should be used to get a biopsy. There are

articles which describe successful attempts of performing a percutaneous transvenous biopsy (PCTVB) of the tumor using TEE and fluoroscopy guidance[10–12]. Our first patient was unstable, and a decision to perform an urgent lifesaving procedure was made. A detailed description of the modified bi-directional Glenn shunt that was performed is provided in our original article. During the same procedure an open biopsy of the tumor was made. Our second patient underwent a PCTVB using TEE and fluoroscopy as guidance. However, this approach failed to give any clinically significant data. Due to increasing right heart failure and corrupted pulmonary artery blood flow a decision was made to operate. During the operation an open biopsy of the tumor was made and a modified bi-directional Glenn shunt was formed.

As with all other lymphopoietic disorders PCL should be treated with chemotherapy. Despite that surgery may be needed, firstly to provide a tumor sample for histological examination, secondly to remove the tumor if it is possible, and lastly if the tumor is not removable to reroute the blood flow in order to stabilize the patient's hemodynamic status. Regardless of the fact that PCL itself may be treated only by chemotherapy, due to its involvement of the heart chambers, surgery has its role in treating this disease, by providing the means of diagnosing and time for chemotherapy administration[7,13,14].

The conclusions remain the same as in our previous article. PCL is still a rare disease. The golden standard is still chemotherapy, though palliative surgery may help, especially if the patient is unstable, due to the disturbed venous blood return. However, we would like to emphasize, the necessity of early involvement of hematologic disorders specialist in treating patients with PCL.

Contributors' statement

All authors participated in the design of the case report and coordination and helped to draft the manuscript. All authors read and approved the final manuscript.

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