Case report - Cardiac general

Unusual presentation of primary cardiac lymphoma*

Izhar N. Bagwan*†, Saral Desai‡, Andrew Wotherspoon*, Mary N. Sheppard*

*Department of Histopathology, The Royal Brompton Hospital, London, UK  †Department of Histopathology, The Royal Marsden Hospital, London, UK

Received 4 February 2009; received in revised form 23 March 2009; accepted 24 March 2009

Abstract

Cardiac lymphomas are rare neoplasms and account for a minor proportion of primary cardiac malignancies. Secondary involvement of the heart and pericardium by systemic lymphoma is well documented, but primary lymphomas of heart and pericardium are extremely rare, accounting for ~2% of all primary cardiac tumours. Most cases are diagnosed at autopsy, but nowadays, with modern imaging technologies, early diagnosis and treatment is possible. Here, we present two unique incidental presentations of primary cardiac lymphomas (PCL), one in an atrial myxoma and other involving a valvular homograft and discuss the potential pitfalls and prognosis of this rare entity.

Keywords: Primary cardiac lymphoma

1. Introduction

Primary cardiac lymphomas (PCL) are extremely rare, accounting for ~2% of all primary cardiac tumours and <1% of extranodal lymphomas [1, 2]. The most common presenting symptoms of PCL are congestive cardiac failure, pericardial effusion, superior vena cava syndrome and arrhythmia [3]. Their incidence is increasing in immunocompromised patients, either HIV related or iatrogenic and hence early diagnosis and treatment has become crucial. This is now possible with modern imaging technologies especially magnetic resonance imaging (MRI), which allows visualisation and characterisation of tumour masses [4, 5]. In spite of this, the appearance of PCL can be very subtle and the diagnosis can be easily overlooked on imaging, giving rise to their presentation as incidental findings.

2. Case report

2.1. Case 1

A 50-year-old man presented with aortic regurgitation (AR) and history of aortic valve replacement (AVR) with a mechanical prosthesis and closure of a ventricular septal defect in 1989. He subsequently developed endocarditis on this mechanical valve and underwent a repeat procedure later in the same year. This was followed by redo AVR first in 1993, then in 2004 and finally in 2005 with porcine stentless freestyle bioprosthesis for severe AR. Coronary arteriogram revealed normal coronary arteries.

During the operation ‘vegetations’ were seen to be completely covering the right and the left coronary leaflets of the aortic valve, but sparing the non-coronary leaflet (Fig. 1a). Histologically the excised homograft revealed that instead of infection the ‘vegetations’ were composed of large pleomorphic malignant cells with necrosis, apoptosis and abundant mitosis (Fig. 1b and c). These cells invaded and destroyed two of the valve leaflets. They were positive for CD45 (Fig. 1d), CD20 (Fig. 1e), CD79a and CD10 with heterogeneous staining for BCL-2 and BCL-6. Approximately 80% of the tumour cells showed Ki67 expression. Immuno-staining for EBV-LMP-1 was negative and there was no over expression of p53. A diagnosis of diffuse large B-cell lymphoma was made. As staging revealed no evidence of disseminated disease or bone marrow involvement, this was considered to be primary cardiac diffuse large B-cell lymphoma. The patient received chemotherapy (R-CHOP: Rituximab-cyclophosphamide, doxorubicin, vincristine and prednisone), but succumbed six months later to rupture of the bioprosthesis with dehiscence and fistula formation found at autopsy. There was extensive necrosis in the area of dehiscence, but no macroscopic or histological evidence of any residual lymphoma at autopsy (Fig. 1f).

2.2. Case 2

An 81-year-old female was referred with a history of multiple strokes associated with what looked like an atrial myxoma on echocardiography. A CT-scan of the brain revealed multiple bilateral middle cerebral artery infarctions. The left atrial myxoma looked exactly as expected at surgery, attached to the left atrial wall in the region of

*Corresponding author. CRY Centre for Cardiac Pathology, NHLI, Imperial College London, London SW3 6NP, UK. Tel.: +44-2073518426; fax: +44-2073518425.

E-mail address: izhardrster@gmail.com (I.N. Bagwan).

© 2009 Published by European Association for Cardio-Thoracic Surgery.
The diagnosis of PCL is made when there is involvement of heart and/or pericardium only without any evidence of nodal or other extra-nodal site of involvement [2]. The rarity of these tumours is illustrated by the finding of only 77 cases reported in the literature up to 2007 [4]. In the series described by Nascimento et al. [4], all five cases of PCL were symptomatic with echocardiographic abnormalities and presented as mass lesions involving the right side of the heart. In these cases in which there are obvious mass lesions the diagnosis can be made with a combination of imaging and intra-myocardial biopsy. However, in our cases the findings were purely incidental with no previous clinical suspicion of lymphoma.
These case reports describe a number of unusual features that are important and challenging from the clinical, diagnostic and pathophysiologic perspectives. PCL are more commonly seen in immunocompromised states, either iatrogenic including those associated with solid organ or bone marrow transplantation or HIV related and these are usually associated with EBV infection. Lymphomas in immunocompetent individuals are extremely rare. In our first case, where the lymphoma was involving the homograft, there was no indication of immunosuppression being involved in the process, and EBV was not present. Patients with homograft valves do not receive immunosuppressive therapy and no previous case of lymphoma involving a cardiac homograft has been reported.

There have been case reports of PCL mimicking atrial myxoma and presenting as an atrial mass [6]. Primary presentation of cardiac lymphoma occurring within an atrial myxoma has not previously been described.

Most PCL are B-cell lymphomas which are seen also in our two cases, but T-cell neoplasms have been described [4, 7]. The differential diagnosis in cases of a poorly differentiated malignant neoplasm involving the heart usually include metastatic carcinoma, melanoma, angiosarcoma and lymphoma. Immunohistochemistry is essential to solve the diagnostic challenge of these tumours. The prognosis is variable depending upon the aggressiveness of the disease, with some showing excellent results with long-term survival while others have a poor prognosis. Both our cases highlight unexpected significant finding in surgical cardiac material and emphasise that all surgical cardiac specimens should be sent for histological analysis.

References