The successful treatment of primary cardiac lymphoma with a dose-dense schedule of rituximab plus CHOP

Primary cardiac lymphoma (PCL) is a rare and often fatal malignancy with a varied clinical presentation [1]. Despite the well-documented clinical course, there is little consensus on management of this lymphoma, including the best modality of medical imaging for diagnosis and monitoring [1]. We report the first case of the combination of rituximab and CHOP (cyclophosphamide, adriamycin, vincristine,
prednisolone) (R-CHOP) given at 14-day intervals as successful therapy for PCL. We also highlight the integral role of cardiac magnetic resonance imaging (MRI) and 18F-fluorodeoxyglucose positron emission tomography (FDG-PET) in guiding diagnosis, management and monitoring of this condition.

A previously well 76-year-old woman presented with syncope and increasing dyspnoea. Investigations for suspected pulmonary embolus with computed tomography (CT) pulmonary angiography revealed a large pericardial effusion and right atrio-ventricular mass. Further investigation with transoesophageal echocardiography and cardiac MRI confirmed an extensive intramyocardial mass involving the right atrium, right ventricular free wall and encircling the pulmonary artery trunk. Surgical biopsy using a xiphisternal approach revealed the histology of a diffuse large B-cell lymphoma (DLBCL) staining CD20- and CD79a-positive. Approximately 80% of the cells were Ki67-positive. Staging including CT scanning of the neck, chest, abdomen and pelvis, cardiac MRI, PET imaging, and bone marrow examination confirmed the lymphoma to be confined to the heart. The anatomical extent and metabolic activity of the lymphoma was clearly demonstrated with the combination of cardiac MRI and PET scanning (Figure 1). Laboratory findings demonstrated an elevated lactate dehydrogenase of 1056 U/l (370–680); however, serum electrolytes, cardiac biomarkers (troponin I and CK-MB), full blood count and blood film were all normal. HIV serology was negative. The patient’s Eastern Cooperative Oncology Group score at presentation was 2, resulting in a high-intermediate International Prognostic Index (IPI) [2].

To assess the potential for cardiac rupture following chemotherapy [3], the muscular integrity of the right atrium and ventricle was assessed using cardiac MRI tagged short axis slices. These sequences revealed the underlying myocardium to be intact and contractile. The patient commenced a 14-day interval dose-dense schedule of R-CHOP [4]. Growth factor support with pegylated granulocyte colony-stimulating factor was administered 24 h following the completion of chemotherapy. To reduce the risk of sudden death following rapid tumour regression, the initial course was administered with a 50% dose reduction of cyclophosphamide and adriamycin. Chemotherapy was completed with a median of 16 days between treatment cycles. Re-staging following the fourth cycle of chemotherapy demonstrated complete resolution of the disease (Figure 1). The patient is currently asymptomatic with no evidence of disease recurrence 11 months following the original diagnosis.

DLBCL is the histological diagnosis in >80% of immunocompetent patients with PCL, and recent evidence suggests that the best therapeutic strategy for DLBCL is likely to be a combination of rituximab and dose-dense chemotherapy [4, 5]. We have demonstrated that six cycles of R-CHOP given in a dose-dense schedule is effective and well-tolerated, even in elderly patients with a high-intermediate IPI. This form of therapy complimented by sensitive medical imaging (cardiac MRI and PET scan) should be considered for all patients diagnosed with this rare malignancy.

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