

# Impact of serial echocardiography in the management of primary cardiac lymphoma

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Primary cardiac lymphoma (PCL) is a rare entity [1] and defined as a lymphoma with the main bulk localized to the heart. The prognosis is poor since diagnosis is often delayed and median survival is less than 12 months. More than 90% of PCL is classified as diffuse large B-cell lymphoma (DLBCL).

Combined 18 F-fluorodeoxyglucose positron emission tomography (FDG-PET)/computed tomography (CT) scan has been extensively used in the diagnostic evaluation, staging, and treatment monitoring of different malignant tumors [2]. Immunochemotherapy is nowadays the most effective treatment for PCL [3]. Regarding cardiac lymphoma remission, PET study is suggested to be more accurate than magnetic resonance imaging (MRI) and echocardiography [4].

The aim of our study was to elucidate the usefulness of echocardiographic monitoring in the clinical management through the reports of three

emblematic cases of PCL that were successfully treated, focusing on changes associated with disease regression and identification of potential side effects.

We observed 3 PCL cases among 458 newly diagnosed lymphomas from 2002 to 2014. PCL accounted for 0.6% out of all lymphomas. Diagnosis was performed by heart mass biopsy in two patients and by mediastinal lymph node biopsy in one patient.

At the time of diagnosis, Patients 1, 2, and 3 were aged 46 years (male), 74 years (male), and 70 years (female), respectively. Histology confirmed the diagnosis of DLBCL in all PCL cases. None had a previous history of cardiac disease or acquired human immunodeficiency virus. Clinical presentation included shortness of breath, chest pain, palpitations, and cough; two patients developed systemic manifestations such as fever. All patients underwent combined FDG-PET/CT scan for initial staging, which revealed increased glucose metabolism in correspondence of the cardiac lesions.

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At the time of diagnosis, serial echocardiography was performed (Fig. 1A) prior to and after each course of chemotherapy. The lymphoma was localized at the level of the right chambers in all patients: the soft tissue density was localized at the level of the free wall of the right ventricle (RV), and one patient also presented also a lateral right atrial wall involvement. All patients had right ventricular dysfunction, and the one with pericardial effusion had RV wall hypokinesia (Figs. 1 and 2). Left ventricular ejection fraction and volumes were normal in all the patients. We used sulfur hexafluoride contrast agent (SonoVue, Bracco Imaging, Milan, Italy) to enhance the visualization of the RV cavity, demonstrate tumor infiltration, and evaluate tumor vascularity (Fig. 2, see [supplementary material video](#)). Prompt treatment was initiated in all the patients receiving pretreatment with steroids and vincristine in

order to avoid acute complications, in particular cardiac perforation due to tumor cytolysis. Next, six cycles of R-COMP (Myocet not pegylated liposomal doxorubicin, rituximab, vincristine, and prednisone) were administered. No acute cardiac event occurred during chemotherapy. In all the patients, a significant reduction of the mass dimension (>50%) was appreciable at transthoracic echocardiography (TTE) after the first cycle of COMP.

We also performed tissue Doppler-derived strain and 2D-strain imaging of the RV with evidence of progressive improvement of RV myocardial function (Fig. 1). When no evidence of cardiac involvement was demonstrated by TTE, a second FDG-PET/CT scan confirmed the complete regression of the tumor involvement of the heart. After restaging, all three patients were considered to have had a complete remission (CR). After a

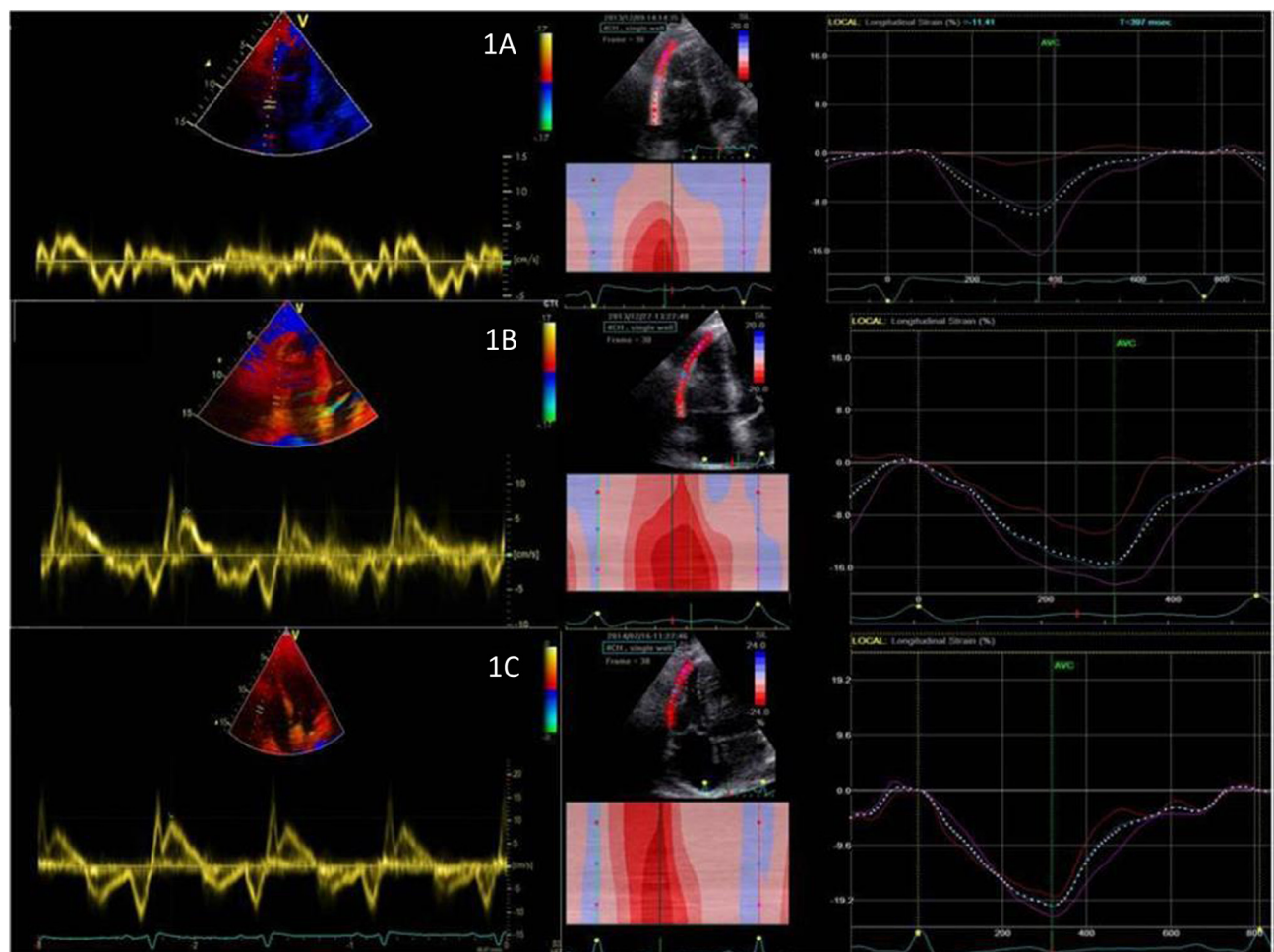


Figure 1. (A) Tissue Doppler of tricuspid annulus and right ventricular strain imaging of Patient 1 at the time of diagnosis showing very low myocardial velocities. (B) Tissue Doppler of tricuspid annulus and right ventricular strain imaging of Patient 1 at the end of the first chemotherapy cycle showing improvement of myocardial velocities. (C) Tissue Doppler of tricuspid annulus and right ventricular strain imaging of Patient 1 at the end of chemotherapy treatment showing normalization of myocardial velocities.

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