Journal für
Kardiologie
Austrian Journal of Cardiology
Österreichische Zeitschrift für Herz-Kreislauferkrankungen

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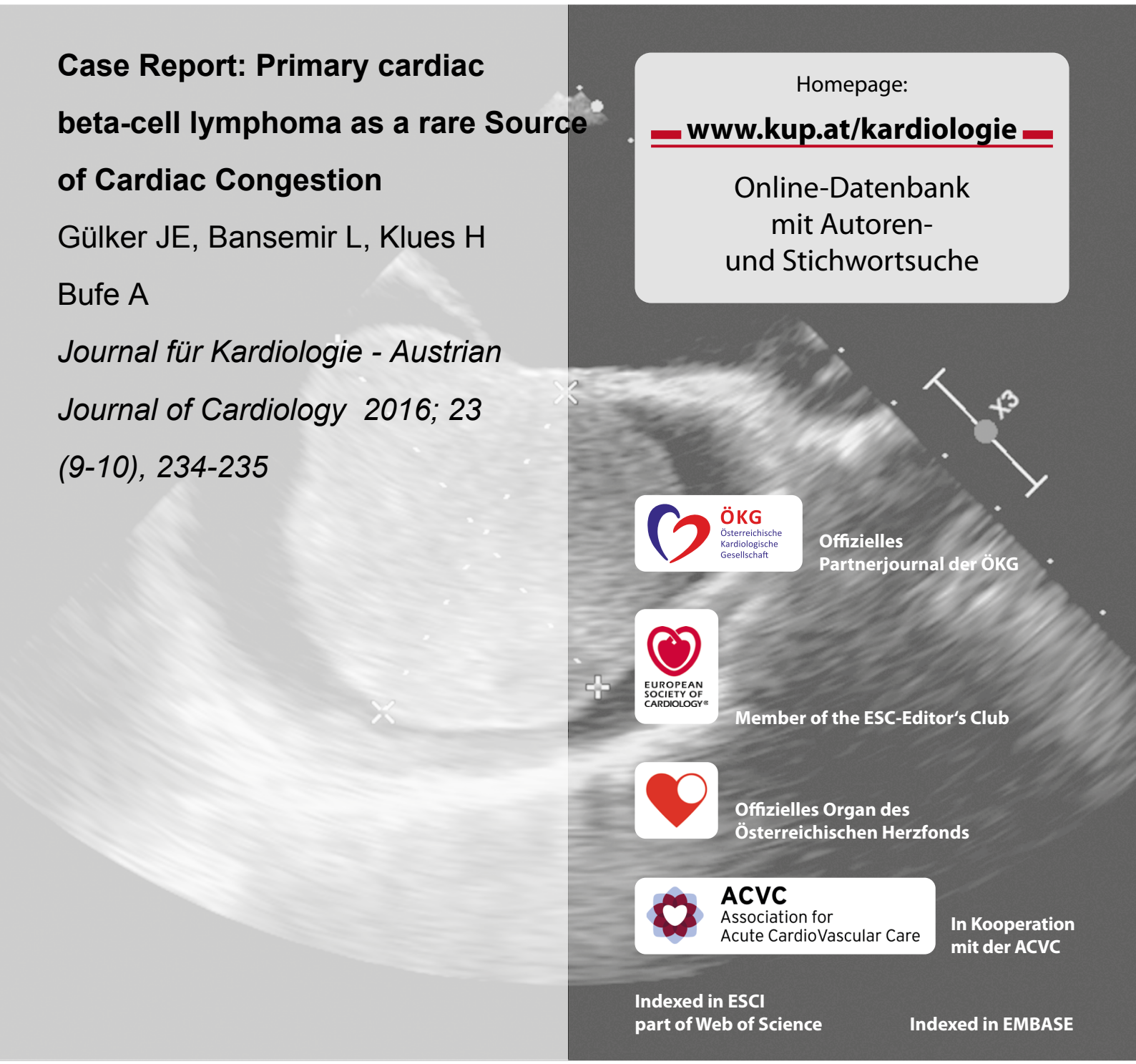
Journal für Kardiologie - Austrian Journal of Cardiology 2016; 23 (9-10), 234-235



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Case Report: Primary cardiac β-cell lymphoma as a rare Source of Cardiac Congestion

J.-E. Gülker, L. Bansemir, H. Klues, A. Bufe

From the Herzzentrum Niederrhein, Medizinische Klinik I, HELIOS Klinikum Krefeld

The occurrence of primary cardiac lymphoma (PCL) is an extremely rare malignancy. Non-Hodgkin's lymphoma (NHL) is either growing on the surface of the heart or infiltrating the heart and/or pericardium [1]. The occurrence of PCL has an estimated incidence of 0.05% in successive autopsies [2].

Case Report

We present a case of a 78-year old male who was hospitalized to operate laparoscopically an umbilical hernia. The procedure was performed successfully, and the patient was dismissed without any physical problems after three days.

A week later he presented himself with abdominal pain, shortness of breath and peripheral oedema resistent to diuretics. Signs of congestive right heart failure were rapidly progressing.

A pericardial effusion was diagnosed with a maximum diameter of 17 mm. Thorax-CT and Cardio-MTR revealed furthermore a 75 × 49 × 67 mm filling defect (Fig. 1) expanding in parts of the right atrium and right ventricle inducing a functional stenosis of the tricuspid valve. Ultrasound revealed pleural effusions on both sides and ascites fluid. The vena cava inferior and the vena mesenterica superior were overfilled. The echocardiography confirmed an intraluminal mass which extended between the right atrium and the right ventricle. The mass was initially considered a thrombus.

Pleural, pericardial and ascites punction were performed; the punctate was microscopically and histologically analyzed. The analysis revealed numerous CD20-positive B lymphocytes with large nuclei and lymphoblasts.

A coronary angiography showed the mass being fed by small arterial vessels from the right coronary artery (RCA) (Fig. 2). Additionally a whole body FDG-PET was performed (Fig. 3) showing a highly increased utilisation of glucose (SUV 13.3) of the mass in the lumen of the right atrium and right ventricle but revealed no signs of an extracardiac disease, particularly no immunocompromise.

Suspected diagnosis: Primary Cardiac Lymphoma

Subsequently medical treatment using the CHOP scheme, including cyclophosphamide, hydroxydaunorubicin, vincristine and prednisone was applied. Following this scheme the clinical signs of cardiac congestion disappeared completely with-

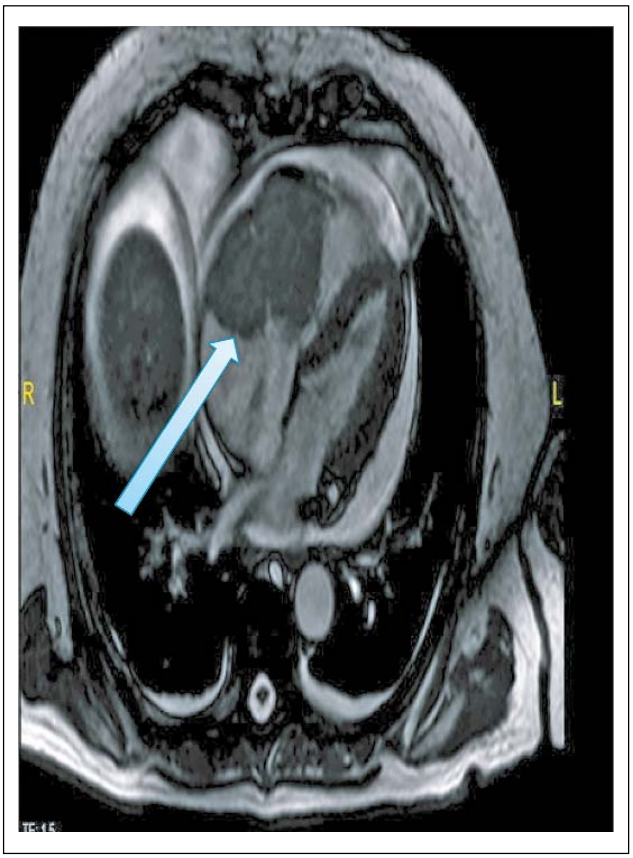

Figure 1: Cardio-MTR with 75 x 49 x 67 mass in right atrium (RA) und right ventricle (RV).

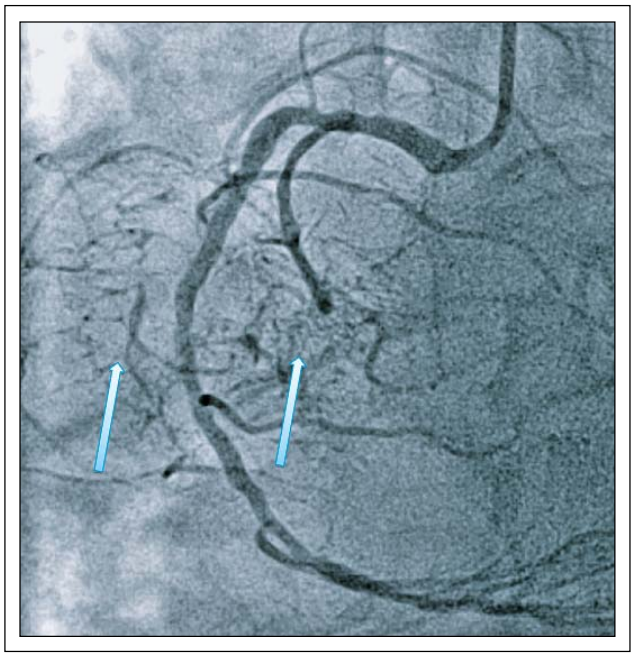

Figure 2: Coronary angiography showed the mass being fed by small arterial vessels from the right coronary artery.

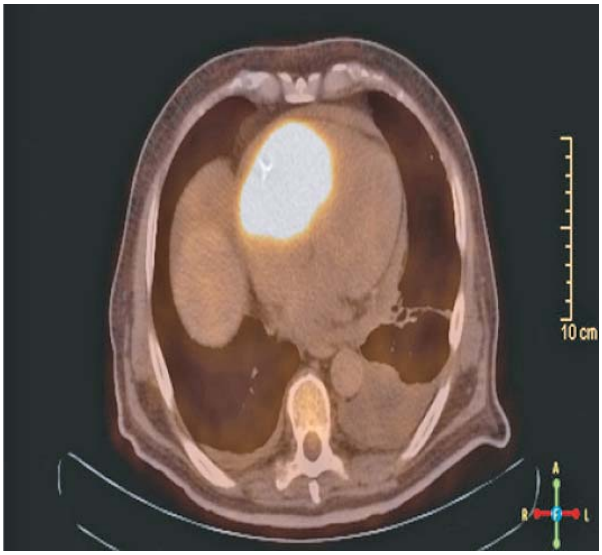

Figure 3: Body FDG-PET with highly increased utilisation of glucose of the mass in the lumen of the right atrium and right ventricle.

in one week. After another two weeks the tumor was significantly reduced with remaining smaller mobile parts (Fig. 4).

Discussion

PCL is a very rare tumor. The tumor favors the right side of the heart. Dyspnoe, refractory to diuretics and chest pain are the most common symptoms. Rapidly progressing right heart failure occurs in almost 50% of cases [3]. Cardiac arrhythmias, particularly atrial arrhythmias may occur. The treatment of choice is chemotherapy applying the CHOP scheme. PCL should be considered in the differential diagnosis of chest pain combined with congestive right heart failure refractory to diuretics. As the expansion of the tumor is often – like in this case – foudroyant, a rapid clarification of the clinical syndrome and subsequent therapy are required. Still 15% of the cases are diagnosed only *post mortem*.

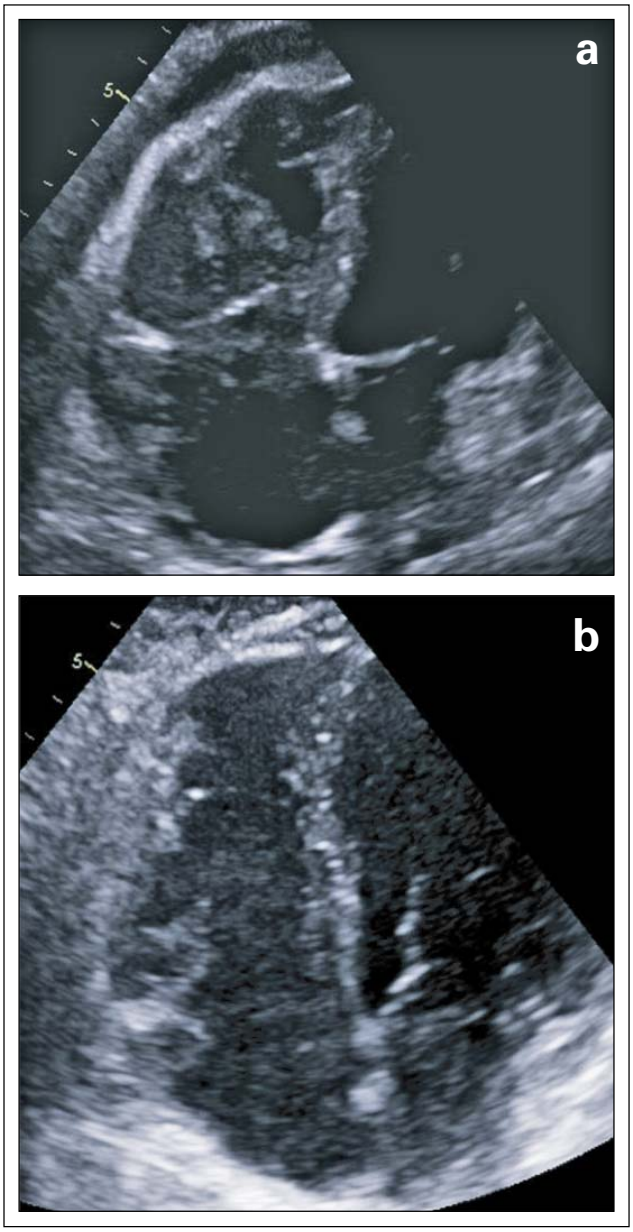

Figure 4: Transthoracic echocardiography before and after chemotherapy with reduced mass within 8 weeks.

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