Journal of the Formosan Medical Association (2018) xx, 1–5

Available online at www.sciencedirect.com

ScienceDirect

journal homepage: www.jfma-online.com

Case Report

Fulminant primary cardiac lymphoma with sudden cardiac death: A case report and brief review

Jen-Fang Cheng [a], Sze-Hwei Lee [b], Ron-Bin Hsu [c], Shan-Chi Yu [d], Chia-Tung Shun [d], Pang-Shuo Huang [b], Ying-Hsien Chen [b], Chii-Ming Lee [b,*]

[a] *Department of Internal Medicine, Pingtung Hospital, Pingtung, Taiwan*
[b] *Department of Internal Medicine, National Taiwan University Hospital, Taipei, Taiwan*
[c] *Department of Surgery, National Taiwan University Hospital, National Taiwan University College of Medicine, Taipei, Taiwan*
[d] *Division of Forensic Medicine and Pathology, National Taiwan University Hospital, Taipei, Taiwan*

Received 22 October 2017; received in revised form 12 March 2018; accepted 21 March 2018

KEYWORDS
Heart neoplasms;
Hemophagocytosis;
Death;
Sudden;
Lymphoma;
Cardiac

Primary cardiac lymphoma (PCL) is very rare, with the variable clinical manifestations potentially leading to a delayed diagnosis. PCL is usually detected incidentally through image studies, whereas the diagnosis can be confirmed via analysis of pericardial effusion, endomyocardial biopsy tissue, or surgical specimens. Although no standard therapy has been established for PCL, without treatment, the prognosis is grave, with the estimated overall survival being approximately 1 year.

We report a difficult diagnosis and complicated case of fulminant PCL, which is the first comprehensively reported case of PCL with secondary hemophagocytosis. A man presented with progressive dyspnea for 3 weeks, and then sudden cardiac death with ventricular fibrillation occurred. After resuscitation, echocardiography revealed a thickened left ventricular wall and severe mitral regurgitation, and computed tomography showed a right atrial mass with diffuse myocardial lesions. PCL was confirmed through a pathological analysis of specimens collected during mitral valvuloplasty, which also implied extensive myocardial involvement. Bone marrow biopsy demonstrated no evidence of lymphoma involvement, but secondary hemophagocytosis was noted. Despite aggressive chemotherapy, the patient died of sepsis with multiorgan failure 26 days after the operation.

* Corresponding author. Department of Internal Medicine, National Taiwan University Hospital, 7 Chung-Shan South Road, 100 Taipei, Taiwan. Fax: +886 2 2393 5346.
E-mail address: chiiminglee@ntu.edu.tw (C.-M. Lee).

https://doi.org/10.1016/j.jfma.2018.03.011

Introduction

Primary cardiac tumors are rare with an incidence of approximately 0.02%,[1] and the majority of them are benign. However, both benign and malignant cardiac tumors may cause disastrous embolism or compromised hemodynamics by obstructing blood flow.[1] Image modalities, including echocardiography (UCG), computed tomography (CT), and magnetic resonance image (MRI), are helpful in the detection, etiological diagnosis, hemodynamic evaluation, complication identification, and determination of therapeutic strategies of primary cardiac tumors.[2] Primary cardiac lymphoma (PCL) is very rare, with a prevalence of 1.3%–2% of all primary cardiac tumors.[1] Moreover, PCLs usually present with nonspecific symptoms and are often detected incidentally.[3]

Case report

A 58-year-old man had been well until progressive exertional dyspnea, orthopnea, bilateral leg edema, palpitations, and general malaise developed 3 weeks before presentation at our institution. In response to diuretics prescribed at a chest clinic, the leg edema partially resolved but the palpitations persisted. An electrocardiogram (ECG) showed atrial fibrillation with rapid ventricular response, and chest radiography revealed cardiomegaly with pulmonary congestion (Fig. 1A). He visited the

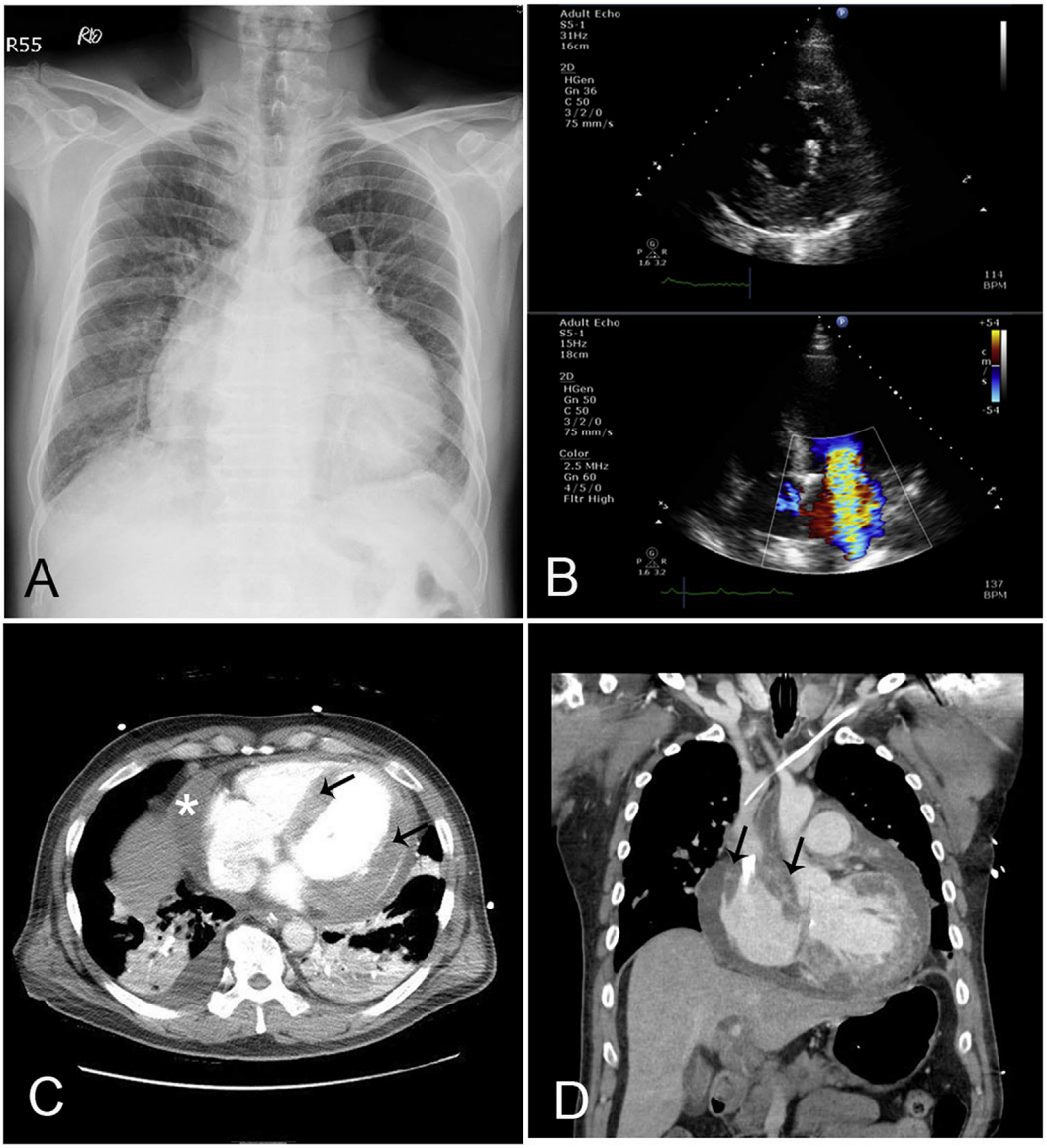

Figure 1 Chest X-ray, echocardiography, and CT. Chest X-ray revealed cardiomegaly and pulmonary congestion (A). Echocardiography showed hypertrophy at the posterior lateral wall of the left ventricle, impaired left ventricle contractility, and severe mitral regurgitation (B). Computed tomography (CT) revealed heterogenous enhancement in the left ventricular myocardium (arrow) and a moderate amount of pericardial effusion (asterisk) (C). The reconstructed coronal view of the chest CT revealed a right atrial mass (arrow) (D).

Please cite this article in press as: Cheng J-F, et al., Fulminant primary cardiac lymphoma with sudden cardiac death: A case report and brief review, Journal of the Formosan Medical Association (2018), https://doi.org/10.1016/j.jfma.2018.03.011