Case report

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Diagnosis of rare Loffler endocarditis by cardiac ultrasound: a case report and literature review

Juanjuan Li[1], Ping Li[1], Maogang Gao[2], Xiaoxiao Yin[1], Dongxia Gao[1], Xiaohui Ji[1]

[1]Department of Ultrasound, [2]Department of Medical Thoracic Surgery, The Fourth Hospital of Hebei Medical University, Shijiazhuang, Hebei, China

Abstract

Loffler endocarditis is a rare disease associated with high mortality rates, therefore early diagnosis and prompt treatment are crucial factors in managing this condition effectively. The clinical manifestations are nonspecific which can lead to misdiagnosis easily. Here we report a case of rare idiopathic hypereosinophilic syndrome with Loffler endocarditis as the first presentation, first suspected acute coronary syndrome, diagnosed correctly by cardiac ultrasound. The purpose is to improve our understanding of the ultrasound manifestations of this disease.

Keywords: Loffler endocarditis; cardiac ultrasound; diagnosis

Introduction

Loffler endocarditis is a disease characterized by endocardial or myocardial damage caused by hypereosinophilic syndrome (HES) involving the heart and belongs to the category of restrictive cardiomyopathy [1,2]. The clinical manifestations are nonspecific which can lead to misdiagnosis easily. Cardiac ultrasound is often considered as the initial imaging technique for evaluation and when it reveals endocardial thickening and mural thrombosis, in conjunction with the patient's cardiovascular symptoms and increased eosinophils, this disease should be highly suspected. Here we report a case of Loffler endocarditis, which was diagnosed firstly by cardiac ultrasound.

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Corresponding author: Xiaohui Ji
Department of Ultrasound,
The Fourth Hospital of
Hebei Medical University,
Shijiazhuang, Hebei 050000,
China
Phone: 86-13832342560
E-mail: 47600348@hebmu.edu.cn

Case report

A 30-year-old woman was admitted with palpitations, chest tightness, and shortness of breath. Laboratory data were as follows: troponin I, 0.69 ng/mL; creatine kinase isoenzyme, 20 ng/mL; white blood cells, 19.82×10^9/L; eosinophils, 12.29×10^9/L (62%). The electrocardiogram on admission showed ST segment depression in leads V5-V6 with low and flat T waves. On echocardiography endocardial thickening was noted in the lower middle and apical parts of the left ventricle (LV) with a normal LV ejection fraction and diastolic dysfunction (fig 1). These resulted in reduced volume of LV cavity, apical occlusion, and reduced amplitude of motion of LV walls. Loffler endocarditis was suspected. Global longitudinal strain (GLS) measurements for different views were as follows: A4C: -10.2%, A2C: -9.6%, A3C: -5.6%, average: -8.5% (fig 2). Further enhance relevant examinations: coronary angiography revealed no significant abnormalities; autoimmune examination showed negative; parasite-related tests yielded negative results. Based on the patient's clinical manifestations and comprehensive examination findings, a clinical diagnosis of Loffler endocarditis was made.

A week later, the patient transferred for further management. On repeated echocardiography a crescent-

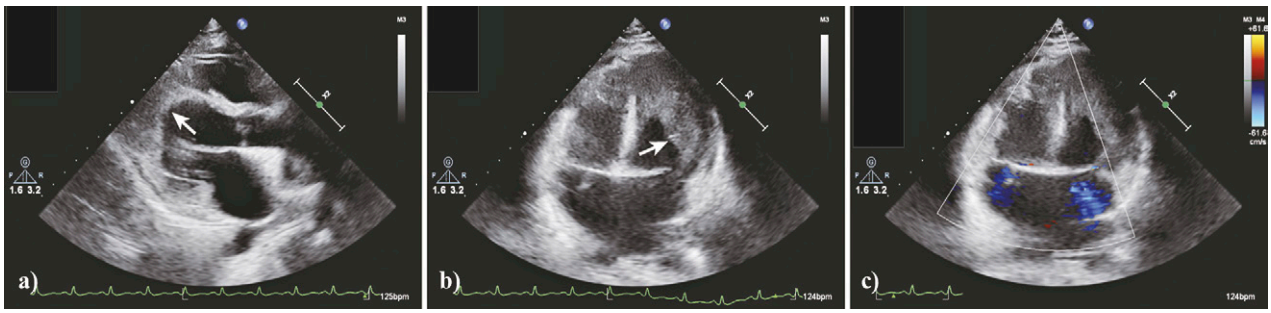

Fig 1. Initial cardiac ultrasound imaging of the patient showing (a) parasternal long axis plane, a diffuse hypoechoic area (arrows) at the apex of the LV; (b) apical four-chamber plane, the hypoechoic area (arrows) in the lower middle and apical parts of the LV measuring approximately 61.8×34.3 mm; (c) apical four-chamber plane, mild mitral valve and tricuspid valve regurgitation.

shaped slightly high echo (28×29 mm, significantly smaller than before) observed in the apex of the LV, consistent with Loffler endocarditis. Laboratory data showed decreased eosinophil count at 1.94×10^9/L (22.74%). Chest computed tomography demonstrated mixed pulmonary edema and presence of some high-density shadows in the lungs that could not be ruled out as inflammation. Bone marrow analysis revealed negative results for FISH screening targeting FLT3, JAK2, ETV6, PDGFRA, PDGFRB and FGFR1 genes; negative MDS gene mutation screening; negative leukemia fusion gene screening. The patient was finally diagnosed with idiopathic hypereosinophilic syndrome, pulmonary eosinophilic infiltration, and Loffler endocarditis. Subsequently, intravenous administration of 40 mg methylprednisolone sodium succinate was initiated once daily for six days. Concurrently, anticoagulation therapy, ventricular rate control, and improvement of cardiac function were implemented. Following comprehensive treatment, the patient experienced subjective symptom relief.

Currently, the patient's condition is stable and continuous monitoring along with regular follow-up treatment are being conducted.

Discussion

Loffler endocarditis is a rare disease characterized by endocardial or myocardial damage caused by hypereosinophilic syndrome (HES) involving the heart and belongs to the category of restrictive cardiomyopathy [1, 2]. It was named after first reported by Löffler in 1932. The cause is unknown, and the prognosis is usually poor. Cardiovascular damage is the main cause of death in HES patients [3]. Potential causes include allergies, rheumatic diseases, myeloproliferative disorders, cancer, and idiopathic eosinophilic syndrome. Endocardial biopsy serves as the gold standard for diagnosing Loffler endocarditis; however, due to its invasiveness and associated complications its application is limited. In 1975, Chusid et al proposed diagnostic criteria for HES: 1) peripheral blood eosinophil count ≥1.5×10^9/L for six months; 2) evidence of organ involvement; 3) exclusion of parasitic infection and allergic diseases that can induce eosinophilia [4]. Specific organ damage exists in HES with skin, heart, and nervous system impairments being more prevalent. Eosinophil-specific granules exhibit cytotoxicity while their metabolites can also harm cells. Additionally, eosinophils produce various factors that promote inflammation and fibrosis including transforming growth factor α and β (TGF-α, TGF-β), tumor necrosis factor α (TNF-α), among others. The combined effects of these factors contribute to multiple organ damage with cardiac impairment being a typical manifestation observed in Loffler endocarditis. The pathological features of Loffler endocarditis involve infiltration and deposition of eosinophils within subendocardium and myocardial tissue leading to endothelial cell injury as well as cardiomyocyte damage ultimately resulting in endocardial thrombosis formation along with fibrosis [5]. When the heart valves are affected, they may present as valve contracture, stiffness, and insufficiency. Loffler endocarditis can be classified into three stages based on pathological changes: acute necrosis, thrombosis, and fibrosis stage. The acute ne-

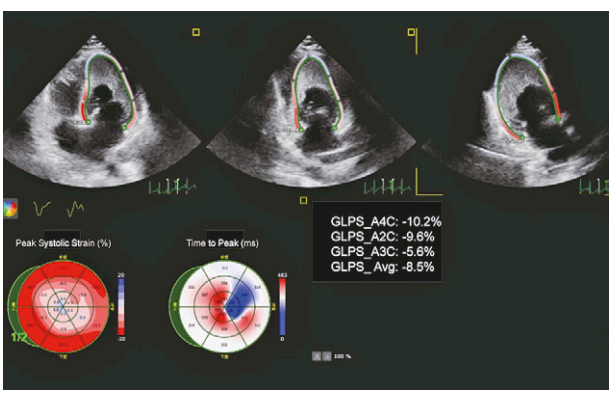

Fig 2. GLS left ventricular myocardial strain image of the same patient.

crosis stage is characterized by endocardial eosinophil infiltration, degranulation, inflammation, and necrosis; the thrombosis stage involves thrombus formation on the endocardial surface; while the fibrosis stage is marked by endocardial fibrosis and restrictive filling disorder.

Clinical symptoms of this disease are often nonspecific. Electrocardiogram findings typically show ST segment depression in leads V5-V6 with low and flat T waves along with elevated serum troponin I levels indicative of early-stage inflammation and necrosis in Loffler endocarditis. Typical echocardiographic manifestations include ventricular inflow part thickening mostly located at the apex resulting in reduced cardiac chamber volume or even occlusion. In the acute phase, there may be an unclear boundary between thickened endocardium and myocardium accompanied by mural thrombus formation at affected sites leading to diastolic dysfunction. Once it progresses to the fibrotic phase later on prognosis becomes poor [6]. The decreased amplitude of wall motion in the lower middle segments of the LV may be attributed to eosinophil involvement in the LV endocardium, myocardium, and coronary arteries. Cardiac magnetic resonance imaging (CMRI) is a more sensitive modality for detecting cardiac damage and has become the gold standard for non-invasive evaluation of cardiac morphology, structure, and function. CMRI manifestations typically include restricted non-dilatation or small chambers, endocardial thickening and fibrosis primarily occurring in the apical region. Additionally, specific subendocardial gadolinium delayed enhancement can also be observed.

It is important to differentiate Loffler endocarditis from other heart diseases such as myocardial infarction, intraventricular thrombosis and hypertrophic cardiomyopathy.

Treatment for Loffler endocarditis is determined based on the underlying cause, disease stage, and symptom severity. Adrenocortical hormone therapy is considered first-line treatment, followed by immunosuppressive therapy if necessary. Additionally, PIK3-related genes may serve as potential therapeutic targets [7]. Other treatments for cardiac conditions should be tailored to individual patients based on their specific clinical manifestations or signs. This may include anticoagulation therapy and heart failure treatment among others. In cases of severe valve damage, valve replacement surgery or even heart transplantation may be required [8].

Conclusion

In summary, Loffler endocarditis is a rare disease associated with high mortality rates, therefore early diagnosis and prompt treatment are crucial factors in managing this condition effectively. The clinical manifestations are nonspecific which can lead easily to misdiagnosis. The ultrasound examination is a non-invasive and convenient diagnostic tool, often considered as the initial choice for evaluation. When cardiac ultrasound reveals endocardial thickening and mural thrombosis, in conjunction with the patient's cardiovascular symptoms and increased eosinophils, this disease should be highly suspected. Treatment strategies are determined based on the underlying cause, disease stage, and symptom severity.

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