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Case Report

Multifocal precursor B-cell lymphoblastic lymphoma in an infant with cardiac involvement: A case report ☆,☆☆

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ABSTRACT

Lymphoma with cardiac involvement is a high-risk lesion, especially in children. We report a rare clinical case of multifocal precursor B-cell lymphoblastic lymphoma in a child with cardiac involvement. A 4-year-old boy presented to the Vietnam National Children's Hospital with a vague headache, but magnetic resonance imaging of the head was normal. After 1 week, the patient showed symptoms of chest pain, fatigue, dyspnea, and abdominal pain. On transthoracic echocardiography and multislice computed tomography of the thorax, a mass was detected in the right atrial wall. Abdominal ultrasound showed a small bowel intussusception, multiple nodules in the intestinal wall, and mesenteric lymph nodes. Histopathology of the bowel confirmed the diagnosis of multifocal precursor B-cell lymphoblastic lymphoma. The patient responded to 3 cycles of chemotherapy for lymphoma. Therefore, combining multiple imaging methods allowed for early diagnosis and improved treatment.

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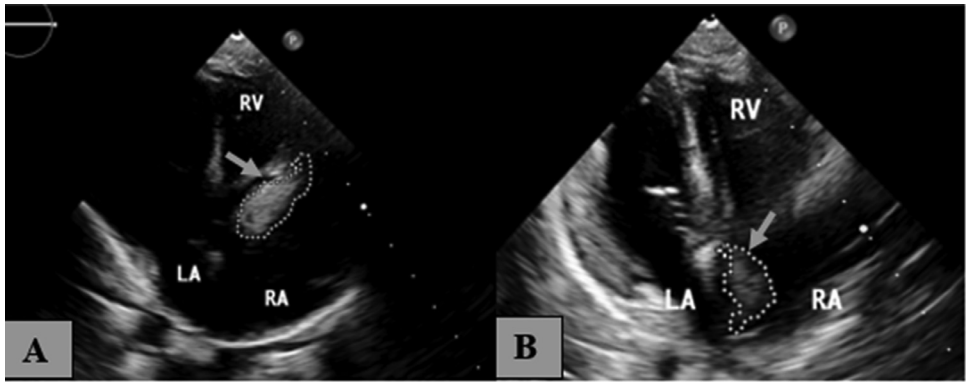

Fig. 1 – Transthoracic echocardiography view showing a mass in the right atrial wall (arrow), which was not yet obstructing the opening and closing of the tricuspid valve (A and B). RV, right ventricle; RA, right atrium; LA, left atrium.

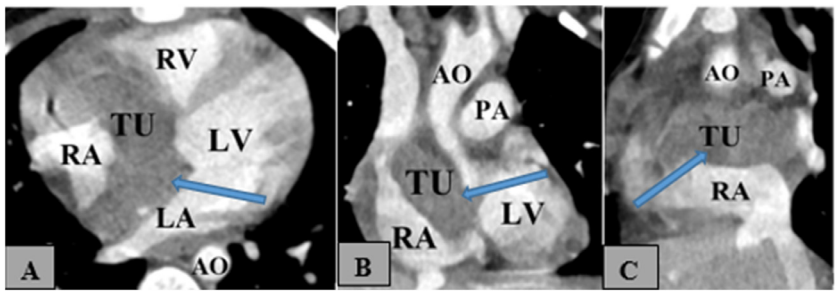

Fig. 2 – Multislice computed tomography (MSCT) was performed following injection with a contrast agent on the axial (A), coronal (B), and sagittal (C) planes. The mass in the atrial wall showed homogeneous density (arrow) and did not compress the superior vena cava or invade the tricuspid valve. TU, tumor; RV, right ventricle; LV, left ventricle; RA, right atrium; LA, left atrium; AO, aorta; PA, pulmonary artery.

Introduction

Lymphoma of the heart and pericardium accounts for approximately 30% of all lymphoma cases [1], but the majority of these cases are diagnosed at autopsy, and very few cases are detected while the patient is alive due to silent symptoms and the rapid progression of injury [2]. Therefore, the early diagnose of this disease is critical to improved outcomes [3,4]. We reported this case to highlight the clinical and imaging features of cardiac lymphoma.

Case presentation

A 4-year-old male child presented to the hospital with a vague headache. A brain magnetic resonance imaging (MRI) appeared normal. Seven days later, the patient appeared complaining of dull chest pain and dyspnea. On transthoracic echocardiography, a hypoechoic mass was identified in the right atrium along the interatrial septum, measuring 12 × 18 mm, which was not hindering the opening and closing of the tricuspid valve, with a thick 3.3-mm pericardial effusion (Fig. 1). Chest multislice computed tomography (MSCT) revealed an infiltrating mass in the right atrium wall, sized 28 × 46 mm (Fig. 2). This tumor did not compress the superior vena cava or invade the tricuspid valve.

One week later, the patient experienced symptoms of abdominal pain with vomiting. Abdominal ultrasound and MSCT showed a small bowel intussusception in the left hypochondriac region. Multiple nodules were observed in the intestinal wall, the largest of which was 11 × 19 mm in size, surrounded by several lymph nodes. Esophagogastroduodenoscopy showed multiple submucosal masses protruding into the lumen of the duodenum, and a biopsy was performed to obtain a specimen (Fig. 3).

The pathology results confirmed a diagnosis of multifocal precursor B-cell lymphoblastic lymphoma. Immunohistochemistry showed leukocyte common antigen (LCA) (+), CD 20 (+), terminal deoxynucleotide transferase (TdT) (+), 90% Ki-67 (+), CD 10 (+), BC12 (+), BC16 (+), CD99 (−), Myeloperoxidase (MPO) (−), CD 117 (−), and CD3(−) (Fig. 4).The patient was treated with chemotherapy using the lymphoma COG-AALL0232 protocol. After completing the third wave of treatment, the patient no longer experienced atypical chest or abdominal pains. On chest MSCT, only a slightly thickened right atrium wall was observed (Fig. 5).

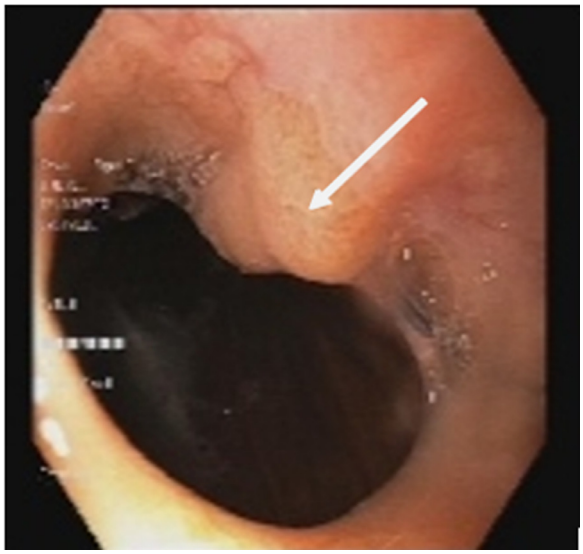

Fig. 3 – Esophagogastroduodenoscopy view showing the submucosal mass protruding into the lumen of the duodenum (arrow).

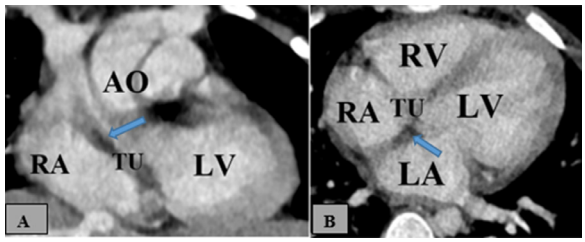

Fig. 5 – After completing the third wave of treatment, MSCT was performed following contrast agent injection. On the axial and sagittal planes, only a slightly thickened atrial wall was observed (arrow), indicating greatly reduced damage compared with before treatment. TU, tumor; RV, right ventricle; LV,left ventricle; RA, right atrium; LA, left atrium; AO, aorta.

Discussion

Lymphoma involving the heart is common among lymphomas; however, most of these cases are diagnosed after death due to rapid progression and fatal complications, [5,6] including arrhythmias, tamponade, and cardiogenic shock. These symptoms are typically nonspecific and often only present when the tumor is very large [7,8]. Therefore, multimodal imaging is the best method for early detection of heart damage in lymphoma cases, and the evaluation of tumor size and extension is essential [9].

Echocardiography is a readily available, non-invasive method for evaluating heart diseases and is the first choice of imaging modalities in many cases. Cardiac lymphoma may appear as hypoechoic mass infiltrating the heart wall, often accompanied by pericardial effusion. According to studies by Ceresoli et al. [12], transthoracic echocardiography allows for the identification of intracardiac masses in 55% of cases, whereas Sanna et al. [13] reported that transesophageal echocardiography allows for the identification of masses in 100% of cases.

Although in many cases, the initial diagnosis of an intracardiac tumor can be performed using echocardiography alone, cross-sectional imaging can be important for further defining the tumor features and relevance. Both MSCT and MRI significantly improve the resolution and contrast of lesions [14,15].

On MSCT, lymphoma typically presents as an infiltrate of the myocardium or pericardium and sometimes as masses in the wall of the myocardium, which usually decrease in proportion to the size of the heart muscle [10,14]. After the administration of an intravenous contrast agent, lesions often enhance heterogeneously [13].

MRI is useful for detecting infiltrates of the myocardium and pericardium [1,14,16,17]. Lymphoma often presents as hyperintense on T2-weighted imaging and as hypointense on T1-weighted imaging, but it can also appear with heterogeneous signal intensity [15,17]. After the injection of contrast agent, either homogeneous or heterogeneous enhancement can be observed [1,11].

Fluorodeoxyglucose (^{18}F-FDG) positron emission tomography-computed tomography (PET-CT) is a sensitive modality that can be used to detect abnormal lesions and contributes information regarding the metabolic activ-

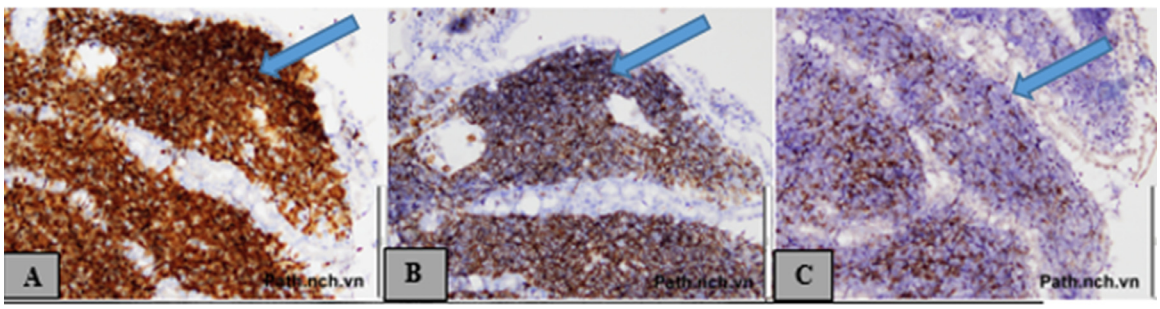

Fig. 4 – Histopathology of the nodule under the duodenal mucosa confirmed the diagnosis of multifocal precursor B-cell lymphoblastic lymphoma. Immunohistochemical staining (× 200) showed that tumor cells were positive for terminal deoxynucleotidyl transferase (TdT, A), leukocyte common antigen (LCA, B), and CD20 (C).

ity of the lesions, which can play an important role in the identification and follow up of lymphoma cases [15].

In our patient's case, the lesion of the heart was detected early using chest MSCT and transthoracic echocardiography. This patient was diagnosed with gastrointestinal B-cell lymphoblastic lymphoma by biopsy during gastroscopy, and the heart lesion responded to chemotherapy treatment, resulting in a final diagnosis of multifocal precursor B-cell lymphoblastic lymphoma with heart involvement. Due to the early diagnosis and active treatment, the patient demonstrated a good response, and all lesions decreased in size significantly after 3 cycles of chemotherapy.

Conclusion

Multifocal lymphoma with cardiac involvement is associated with a high risk of death, especially among children, due to dangerous complications. Because early detection rates are low, multimodality imaging should be performed in all suspected lymphoma patients.

Informed consent

Informed consent for patient information to be published in this article was obtained.

Ethical statement

Appropriate written informed consent was obtained for the publication of this case report and accompanying images.

Author contributions

Tran PN and Nguyen MD contributed to this article as co-first authors. All authors have read the manuscript and agree to the contents.

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