

Case Report

Primary cardiac extranodal natural killer/T-cell lymphoma: a case report and literature review

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Abstract: Background: Primary lymphomas of the heart are very rare, especially primary cardiac extranodal natural killer/T-cell Lymphoma. Primary cardiac natural killer/T-cell lymphoma has diversified symptoms and the diagnosis is difficult. Case presentation: We have reported a 35 year old man with palpitation, fever and increasing dyspnea. Transthoracic echocardiography was done and showed a large piece of mass. We resected the mass which was in the right atrium. Pathologic examination of removed cardiac tumor had reported as below: non Hodgkin-NK/T cell lymphoma. The patient received surgery, without chemotherapy, and was died on the 27th day after surgery of follow-up. Conclusion: This disease has a quick progression and a poor prognosis. Only surgery couldn't control primary cardiac natural killer/T-cell Lymphoma very well. Therefore, early detection is becoming more important.

Keywords: Primary cardiac lymphoma, heart tumor, natural killer/T-cell lymphoma

Background

Primary cardiac tumors are rare, they can be found in up to 0.001%-0.28% of autopsy study [1]. Approximately 90% of primary cardiac tumors are benign with atrial myxomas comprising 75% of those. While the rest 10% of primary heart tumors are malignant [2]. Primary cardiac lymphoma (PCL) (5% of primary malignant cardiac tumors) is a very rare disorder [3]. Among cases with cardiac non-Hodgkin's lymphoma in PubMed between 1990 and 2015, diffuse large B-cell lymphoma was the most common histologic subtype, followed by T-cell lymphoma, Burkitt's lymphoma, and small lymphocytic lymphoma [4]. In this article we report a 35-year-old man with an incidental large cardiac mass who was diagnosed primary cardiac extranodal NK/T-cell Lymphoma.

Case presentation

A 35-year-old man, suffering from a 11-day history of palpitation, fever and increasing dyspnea, visited a regional hospital on July 30, 2015. His heart rate was 153 beats per minute, temperature 38.5°C, blood pressure 83/

58 mmHg and his general fatigue and appetite loss had been worsening daily since admission. Studying his medical history, he denied any other medical illness. Initial physical examination had shown plop from tricuspid area and rash over his neck, chest and abdomen, without pain or itch. An electrocardiogram (ECG) revealed sinus rhythm, a ventricular rate of 153 beats per minute. Transthoracic echocardiography was done and showed a large piece of mass, measuring 55 mm×45 mm in the right atrium, obstructing the right ventricular outflow tract and impinging the tricuspid valve. Enlargement of right atrium, pericardial effusion and persistent left superior vena cava also were shown (**Figure 1**). In head, chest and abdominal CT scan, tumor had not been seen. In laboratory assessment, red blood cell count was 3.34 (×10¹² per liter) (normal range, 4.3-5.8), hemoglobin 102 g per liter (130-175), lymphocyte count 0.45 (×10⁹ per liter) (normal range, 1.10-3.20). The lactic dehydrogenase level was 823IU per liter (normal range, 109-245), the hydroxybutyrate dehydrogenase 618 IU per liter (normal range, 95-250), the pro-brain natriuretic peptide 2304 pg per milliliter (normal range, 0-125). The C-reactive protein

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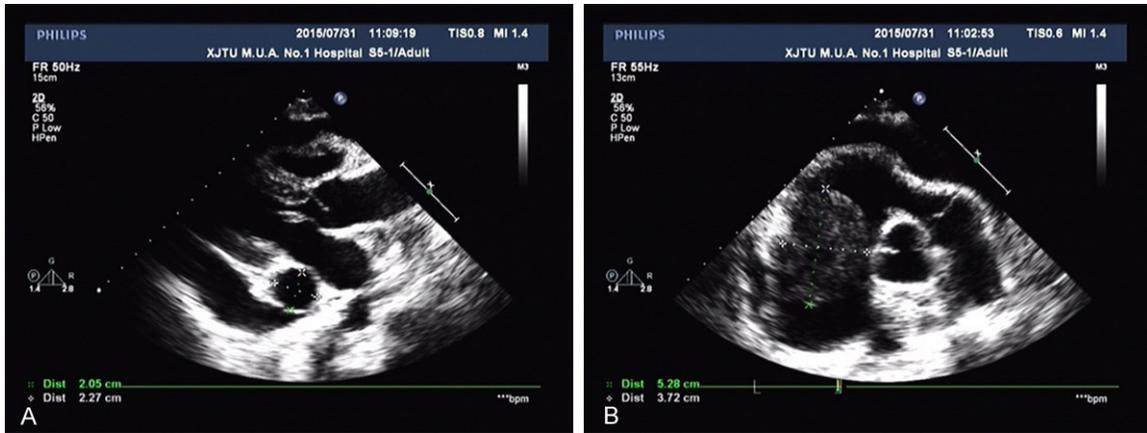


Figure 1. Right Atrial Mass on Echocardiography (A, B).

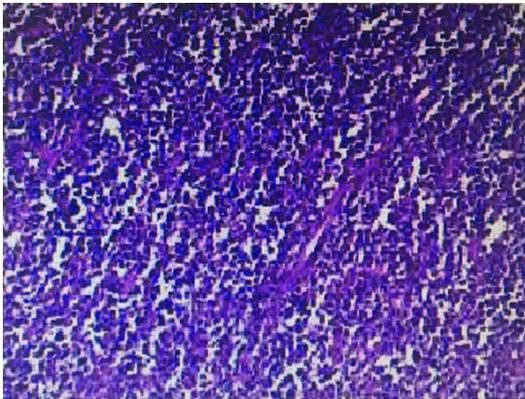


Figure 2. Extranodal NK/T-cell Lymphoma (H and E, $\times 10$).

level was 25.5 mg per liter (normal range, 0-10), the procalcitonin 2.51 ng per milliliter (normal range, <0.5), and staphylococcus hominis was found from a positive blood culture. Additional laboratory findings included a ferritin (one of tumor markers) level of 1530 ng per milliliter (normal range, 30-400), and a carbohydrate antigen level of 49.34 IU per milliliter (normal range, 0-35). This case was discussed in the multidisciplinary meeting and decided for resection of the right atrium on August 12, 2015. Induction of anesthesia was carried out carefully. After initiation of cardiopulmonary bypass, the form of right atrium was abnormal and the mass, distributing widely and encroaching on right ventricle, was approached through right atriotomy. On palpation, it was very hard. Palliative operation was essential because of the high risk of this cardiac surgery. We just resected the mass which was in the right atri-

um. Pathologic examination of removed cardiac tumor had reported as below after immunohistochemistry study: CD3+, GRB+, TIA-1+, Ki67+, EBER in situ hybridization: positive. Compatible with non-Hodgkin lymphoma in favor of NK/T-cell origin (**Figure 2**). After resection of the mass, uncomfortable symptom, such as palpitate and dyspnea, had diminished. However, the patient and his family insisted on leaving hospital after the wound healing despite of the need for further clinical medications. During follow-up period, the patient died on September 8, 2015.

Discussion

Extranodal NK/T-cell lymphoma is more clearly defined in the World Health Organization classification of lymphomas [5]. It accounts for <1% of all lymphomas in Western countries, and 3% to 9% of all lymphomas in Asia [6, 7]. Primary cardiac extranodal NK/T-cell lymphoma is a subtype of cardiac non-Hodgkin's lymphoma (NHL) [8]. Because of the rarity of primary cardiac NK/T-cell lymphoma and the diversity of symptoms, the diagnosis is difficult and the prognosis is often poor [9]. While the median survival was only 3 months for all patients with cardiac NHL [4]. In this case, the patient died in 50 days since symptoms occurred.

Primary cardiac tumours present with different symptoms of the classic triad: cardiac symptoms resulting from intracardiac obstruction; signs of systemic embolisation; and systemic or constitutional symptoms [10, 11]. In this case, the patient suffered from increasing dyspnea,

palpitation and fever. Dyspnea from pulmonary venous hypertension or frank pulmonary oedema are the commonest cardiac symptoms [12]. Fever, weight loss, and fatigue are forms of systemic manifestations. They always cause confusion with infective endocarditis, especially transthoracic echocardiography shows a piece of mass and blood culture is positive. Initially, based on echocardiography, blood culture as well as clinical symptoms, such as fever, rash, fatigue, anemia, we thought the patient might be with infective endocarditis. However, his fever was on going after using a few days of antibiotics. Meanwhile, tumor markers were strong positive. A presumed diagnosis, which was confirmed by surgery and pathologic biopsy, was cardiac tumor with infection endocarditis.

The imaging methods frequently used for the diagnosis and evaluation of cardiac masses are transthoracic echocardiography, CT and CMRI [12]. A transthoracic echocardiogram is the most easily available and useful tool to identify at least the presence of cardiac mass. CT and CMRI enable optimal assessment of the location and soft tissue features of the mass and may possibly provide an accurate identification [13]. Up to now, only pathologic biopsy, which is not accepted by all of patients, can serve as a gold standard for diagnoses. Most Chinese do not have a habit of regular physical examination, thus, in most cases, the disease often reaches advanced stage once feeling uncomfortable. This case gives us some inspiration: maybe early detection can prolong survival period despite diagnostic methods are limited.

Definitive treatment of primary cardiac extranodal NK/T-cell lymphoma is still not established [14]. Several articles reported successful treatment of primary cardiac lymphoma with chemotherapy alone [15-17], while others reported successful treatment by surgery and chemotherapy [18, 19]. However, Ceresoli et al. [17] reported that there was no evidence of survival improvement with surgery. In this case, the patient, without chemotherapy, died on the 27th day after the resection of the right atrium. We can't rule out surgery as an option completely, but from this case the benefits for patient is very limited. Because the patient and his family refused chemotherapy, we don't know whether it is more effective. We

have no idea which therapeutic method is better, chemotherapy alone or surgical resection followed by chemotherapy might have been effective before the tumor invaded the right ventricle.

Conclusion

We report a case of primary cardiac NK/T-cell Lymphoma, which is a very rare disorder. The disease, with fever, right atrium vegetation, positive blood culture, is easily confused with infective endocarditis, and it has a quick progression and a poor prognosis. Only surgery couldn't control primary cardiac NK/T-cell Lymphoma very well. Therefore, early detection is becoming more important. In addition, with the further understanding of molecular biology and genomics, we believe that more available biomarkers for early diagnosis will be found, and more effective treatment options will update the current clinical therapeutic regimens.

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Disclosure of conflict of interest

None.

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