Cardiac Involvement in CD56 Negative Primary Pancreatic Extranodal NK/T-cell Lymphoma, Nasal Type, Presenting with Ventricular Tachycardia during the Early Stages of Chemotherapy

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Abstract

We herein report the case of a 23-year-old man who presented with recurrent pancreatitis and was diagnosed with primary pancreatic extranodal natural killer/T-cell lymphoma, nasal type, involving the right ventricle. The cardiac involvement was screened and confirmed by transthoracic echocardiography (TTE), cardiac magnetic resonance imaging and fluorodeoxyglucose positron emission tomography. Although the patient did not have any cardiac symptoms or evidence of arrhythmia before chemotherapy, he presented with fatal newly developed ventricular tachycardia during the early stages of chemotherapy. The follow-up TTE after his chemotherapy demonstrated markedly decreased thickness of the invaded myocardium, thus suggesting that the myocardium infiltrated by lymphoma cells might become vulnerable to fatal arrhythmia with tumor regression.

Key words: extranodal NK-T cell lymphoma, nasal type, heart neoplasms, ventricular tachycardia

Case Report

A 23-year-old man was admitted due to intermittent abdominal pain of 2 months duration. He was treated for recurrent pancreatitis and did not have any other previous medical history or alcohol history. The chest X-ray demonstrated mild cardiomegaly. Electrocardiography (ECG) showed ST-segment elevation in the V1, V2 and V3 leads (Fig. 1). The QT interval on the ECG was within the normal range. Telemetry did not reveal any arrhythmia, such as a premature ventricular complex (PVC) or ventricular tachycardia (VT), and the cardiac enzyme was normal.

On an abdominal CT, the pancreas was enlarged with peripancreatic infiltration, and the distal tail portion of the pancreas was not observed. An endoscopic ultrasound guided needle biopsy of the pancreas showed diffuse interstitial infiltration of atypical lymphoid cells with extensive necrosis. Immunophenotypically, the infiltrate was composed predominantly of T-cells, which were cytoplasmic CD3 (cyCD3) positive. Although the cells were CD56 negative, there was nuclear atypia and diffuse positive staining in the

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Figure 1. Baseline electrocardiography (ECG) showed ST-segment elevation in the V1, V2 and V3 leads.
Transthoracic echocardiography (TTE) showed right ventricular (RV) enlargement and a thickened RV wall (arrows) with mild pericardial effusion.

(A) The delayed enhancement image on the cardiac MRI showed a heterogeneous delayed enhancement at the thickened RV focal wall (arrow). (B) Positron emission tomography revealed abnormal fluorodeoxyglucose uptake of the RV (arrow).

Telemetry showed ventricular tachycardia with a left bundle branch block morphology followed by frequent premature ventricular complexes at the time of collapse.

In situ hybridization Epstein-Barr virus (EBV)-encoded RNA and cytotoxic molecules. Transthoracic echocardiography (TTE) was performed due to the ECG abnormality and demonstrated a dilated right ventricle (RV) and hypertrophied mid to basal RV anterior wall with mild pericardial effusion (Fig. 2). The tricuspid regurgitation was trivial, and the estimated pulmonary arterial systolic pressure was 37 mmHg. The cardiac magnetic resonance imaging (MRI) was additionally performed and revealed thickening of the RV focal wall with heterogeneous delayed gadolinium enhancement (Fig. 3A). Fluorodeoxyglucose positron emission tomography (FDG-PET) CT showed an abnormal hypermetabolic lesion in the pancreas without the tail of the pancreas and RV (Fig. 3B).

The patient was diagnosed with CD56 negative primary pancreatic extranodal natural killer (NK)/T-cell lymphoma, nasal type (ENKL). He was placed on an ifosfamide/methotrexate/VP-16/prednisone (IMVP-16/Pd) and L-asparaginase regimen. However, the second day after starting chemotherapy, the patient suddenly collapsed. Intravenous prednisone was used for the first 2 days according to the chemotherapy regimen. The blood test did not show any significant change, in comparison to the baseline’s results, or significant electrolyte imbalance. Telemetry showed VT possibly originating from the RV at the time of collapse.
A follow-up TTE after the first round of chemotherapy showed decreased RV wall thickness (arrows) as compared with the initial TTE.

Discussion

ENKL is one of the types of natural killer-cell lymphoma and extranasal type of ENKL has a highly aggressive clinical course and a poor prognosis (1). ENKL has an immunophenotypical expression of cyCD3+/CD56+ cytotoxic molecules and evidence of EBV infection (2). CD56 is a useful marker for ENKL, but it is not specific for ENKL and can be negative in a few ENKL cases (3). Our patient was diagnosed with CD56 negative ENKL based on the cytolological and immunohistochemical analyses and is a rare case due to its primary pancreatic origin and CD56 negativity (4).

When the heart is occupied by a tumor, the clinical symptoms can be nonspecific and cardiac involvement is frequently undetected before death (5). In our case, the cardiac involvement was screened and characterized by TTE and the cardiac MRI. Additionally, PET-CT confirmed that the lymphoma was confined to the pancreas and heart. The patient presented with fatal VT shortly after starting his first round of chemotherapy, although he did not have any cardiac symptoms or evidence of an arrhythmia before chemotherapy. The association of VT with malignant lymphoma was previously reported in patients with primary or secondary cardiac lymphoma (6-8). Most of them were reported as presenting with VT at the time of admission or diagnosis. The myocardium infiltrated by the tumor cells may provide an electrically heterogeneous substrate which can be vulnerable toward developing an arrhythmia. Ventricular conduction delay or triggered activity by the myocardium invaded by the lymphoma cells may be one of the mechanisms of VT in this population (9, 10). However, our patient did not have any arrhythmia before chemotherapy and presented VT on the second day of chemotherapy. The follow-up TTE showed markedly decreased thickness of the invaded myocardium, suggesting that the tumor cells responded very effectively to the chemotherapy. The myocardium infiltrated by the lymphoma cells may become vulnerable to fatal arrhythmia following tumor regression by effective chemotherapy, and VT may be triggered from this situation.

In conclusion, we herein described a case of cardiac involvement in a patient with CD56 negative primary pancreatic ENKL presenting with VT during chemotherapy. Clinicians should therefore be alert for the potential occurrence of fatal arrhythmias, and additional cardiac vigilance is warranted during chemotherapy in this population.

The authors state that they have no Conflict of Interest (COI).

References


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