Isolated Extramedullary Relapse of Acute Myelocytic Leukemia After Bone Marrow Transplantation Affecting Left Ventricle and Right Atrium in an Adult

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A 60-year-old woman was admitted to cardiac care unit for 10 days of precordial compressing pain and abnormal electrocardiogram (Panel A) which demonstrated elevated ST segment and T wave inversion at the lateral wall leads and corresponding depressed ST segment at the inferior wall leads. She had been diagnosed with acute myelocytic leukemia-M5 in 2011 and received allogeneic hematopoietic stem cell transplantation 5 months thereafter. Positron emission tomography-computed tomography (PET-CT) performed 5 months prior (Panel B) and bone marrow puncture 2 months before this attack were both reported to be negative. Physical examination was unremarkable with a heart rate of 95bpm, BP 100/75mmHg, and a temperature of 36.5℃. Elevated cardiac troponin (cTNI 0.56ng/mL, reference range 0.0-0.9ng/mL) confirmed myocardial infarction. Peripheral blood analysis was in normal range (white blood cell: 6.56×109/L, Ne%: 70.4%, Mo%: 5.3%, Eo%: 0.2%; red blood cell 3.75×1012/L, hemoglobin 113g/L and platelet 78×1012/L). Serum creatinine and ESR were largely normal (101.7μmol/L and 32mm/h respectively). Bedside echocardiography revealed segmental wall motion abnormality at the left ventricular lateral wall consistent with electrocardiogram, notably with marked asymmetric left ventricular hypertrophy and a large sessile echogenic mass attached to the right atrial lateral wall (Panel C-E, Video 1 and 2 in supplementary material on web-site) which produced no obstruction to blood flow across the inferior vena cava or tricuspid valve. Cardiac magnetic resonance (CMR) cine confirmed a thickened and dysfunctional left ventricular anterior and lateral wall, and a large multilobular mass with fuzzy margin in the right atrium, both of isointensity to the intercostal muscle, indicating their same origin similar to soft tissue (Panel F, Video 3). First-pass perfusion images showed lower intensity of the involved left ventricle and right atrium than that of the unaffected myocardium (Panel G) suggesting decreased coronary microvascular perfusion. Late gadolinium enhancement demonstrated patchy contrast residual at both lesions (Panel H), suggesting infiltrated myocardium wherein. The patient was commenced on conservative therapy for

Highlights

Isolated extramedullary relapse (IEMR) of acute leukemia after bone marrow transplantation is rare, particularly in the heart. We report a case in a female patient which involved both the right atrium in the form of a mass and the left ventricle as asymmetric infiltrative hypertrophy.

Keywords: leukemia; cardiac; extramedullary; relapse; transplant

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acute myocardial infarction. Considering the acute coronary event and the infiltrated and widely attached masses presenting no chance for surgery, no intervention for the extramedullary relapse of acute leukemia was initiated. She died one month later of sudden death.

Hematological malignancy can involve any site of the body and ranks 4th among the malignant tumors most commonly involving the heart.\(^1,^2\) The manifestation of acute leukemia affecting the heart is variable and mostly insidious, including arrhythmia, heart failure, pericardial effusion, mural thrombosis or less commonly as intra-cardiac mass or diffuse infiltration.\(^3,^4\) Isolated extramedullary relapse (IEMR) of acute myelocytic leukemia after bone marrow transplantation, with a prevalence of 5.1%-12%, is rather rare involving the heart.\(^5,^6\) Although histopathology has been the diagnostic gold standard, it remains a challenge in the living patient. Echocardiography serves as an efficient screening tool. Enhanced CMR provides a unique value for its noninvasive determination of tissue characterization. IEMR is considered a precursor of full-blown bone marrow relapse and portends a poor prognosis as reflected from this patient. No standard therapy has been established.\(^5,^7\)

**Declarations of Interest:**
The authors declare that there is no conflict of interests

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**References**

**Figure 1.** Electrogram and cardiac imaging presentations of the patient. LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.