Right Cardiac Chambers’ Involvement as the First Manifestation of Recurrent Complex Karyotype Acute Myeloid Leukemia

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Abstract

We describe a case of a 21-year-old male, with a history of acute myeloid leukemia (AML) treated with allogeneic hematopoietic cell transplantation, referred to our department for atypical chest pain and dyspnea. Echocardiography revealed an extensive mass involving right cardiac chambers and tricuspid valve annulus, with increased thickness and impairment of right ventricle. Cardiac magnetic resonance confirmed the presence of cardiac mass involving pulmonary artery trunk, pericardial sleeves, and lung parenchyma. These findings were attributed to a manifestation of recurrent AML involving the right heart.

Keywords: Cardiac magnetic resonance, cardiac masses, echocardiography, right ventricle failure

INTRODUCTION

Cardiac masses are often accidental echocardiographic findings and appear in many clinical scenarios such as heart failure and arrhythmias.

Secondary cardiac tumors are more frequent than primary ones (ratio 20:1). The incidence of cardiac metastases reported in literature is highly variable, ranging from 2.3% and 18.3%. Lymphoproliferative disorders appear in 9.4% of cases with cardiac metastasis (including autopsies of affected patients), especially with myocardial (64%) and epicardial (44%) involvement.

Cardiac metastases usually affect both intracavitary and intramural right cardiac chambers.

Acute myeloid leukemia (AML) is a malignant neoplasm characterized by infiltration of the bone marrow, blood, and other tissues by proliferative, clonal, abnormally differentiated, and occasionally poorly differentiated cells of the hematopoietic system.

Postremission therapy with allogeneic hematopoietic cell transplantation provides the strongest antineoplastic therapy because of cytoreductive conditioning and the immunologic antileukemic graft-versus-leukemia effect.

However, excessive immunosuppression to limit graft versus host disease (GVHD) can magnify the risks of leukemia recurrence and high-risk cytogenetic and molecular subgroups or hematopoietic cell transplantation after the first complete remission all increase the risk of relapse.

Complex karyotype AML is associated with worse outcome cause of its malignant cytogenetic features.

CASE REPORT

A 21-year-old Caucasian male with a history of complex karyotype AML presented to our Emergency Department with atypical chest pain and dyspnea.

He previously underwent allogeneic hematopoietic cell transplantation with complete regression of AML 12 months before.

Immunosuppressive therapy was strengthened (cyclosporine, mycophenolate, and low doses of steroids) after intestinal and dermatological chronic GVHD occurred.

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Immunosuppressive therapy was strengthened (cyclosporine, mycophenolate, and low doses of steroids) after intestinal and dermatological chronic GVHD occurred.
Mild anemia and a stable rise of high-sensitivity troponin T were detected at biochemical analyses. Electrocardiogram showed atrial tachyarrhythmia with right bundle branch block and elevated voltages in the right precordial leads undetected in a previous one [Figure 1a and b]. Echocardiography revealed an extensive inhomogeneous mass involving right cardiac chambers and tricuspid valve annulus, increased thickness of right ventricle (RV), and hyperechogenic pericardium with mild circumferential pericardial effusion [Figure 2a-d and Videos 1-3]. No excursion of RV lateral wall and paradoxical intraventricular septal movement were detectable in M-mode [Figure 2e]. Indeed, during the diastole, ventricular septum appeared flattened as a sign of volume overload. Furthermore, during the systole, the ventricular septum moved away from the dorsal wall of the left ventricle (LV) and contracted in the direction of the RV, probably to improve its emptying.

A preserved LV global systolic function in the presence of moderate diastolic dysfunction was also found out (ejection fraction 60%, E/A 0.7, and E/E’ 9).

Strain imaging with speckle-tracking methods revealed an impairment of RV function (RV global longitudinal strain [RV GLS] - 10%) and a preserved GLS of LV (LV GLS - 25%) [Figure 3].

Cardiac magnetic resonance (CMR) confirmed the presence of cardiac mass involving pulmonary artery trunk, pericardial sleeves, and lung parenchyma [Figure 4a]. It also showed left cardiac chambers’ preservation.

Short TI inversion recovery sequences revealed a wide area of edema on the right cardiac chambers and surrounding structures [Figure 4b]. No perfusion anomalies on first-pass contrast-enhanced sequences and no delay enhancement area were observed [Figure 4c and d].

These findings were consistent with wide myocardial inflammatory infiltration with no evidence of fibrosis. The patient refused endomyocardial biopsy.

During the in-hospital stay, the patient developed fast atrial tachyarrhythmia unresponsive to medical therapy and a rapid worsening of the performance status.

Fifteen days after the admission, leukocyte count increased and blast cells were detected in the peripheral blood as recurrence of the previous myeloproliferative disease.

Figure 1: Electrocardiographic features: Atrial tachyarrhythmia with right bundle block and elevated voltages in the right precordial derivations (b) undetected in a previous electrocardiogram (a)

Figure 2: Echocardiographic features: cardiac mass involved lateral wall of the right ventricle in parasternal long-axis view (a) and short-axis view (b and c). Apical 4-chamber view showing the involvement of the right ventricle free wall, right atrium, and tricuspid valve annulus (d). No excursion of right ventricle lateral wall and a paradoxical intraventricular septal movement were detectable in M-mode modality (e)

These data were confirmed by morphological, immunophenotypic, and molecular analysis of blood and bone marrow biopsy.

Chemotherapy was immediately started, but the patient died few days after the cause of respiratory failure.

DISCUSSION

This report presents the echocardiographic and CMR detection of an extensive mass involving right cardiac chambers in a young patient with complete remission of an aggressive AML, after the allogeneic hematopoietic cell transplantation. Clinical signs of heart failure and the development of electrocardiographic disturbances are all first manifestations of relapsing leukemia. Atypical chest pain may be due to pericardial sleeves inflammation, whereas electrocardiographic changes and arrhythmias could depend on myocardial and conduction system infiltration.[6]

CMR features were consistent with relapsing leukemia because of the extensive involvement of cardiac chambers and surrounding structures.[7,8]
Probably, no fibrotic areas were detected because of an inability of the immune system of self-restraint due to a concomitant systemic immunosuppressive therapy.

CMR assessment of cardiac mass localization and morphological properties was crucial to define patient risk stratification and clinical management strategy.[7]

Indeed, few cases of cardiac masses as the first recurrence of AML are reported in literature,[8-10] and our echocardiographic and CMR findings are extremely suggestive of the malignant nature of cardiac involvement.[7]

To the best of our knowledge, our case was the first reported in literature of extensive cardiac mass as primary manifestation of relapsing complex karyotype AML.

Chest pain, heart failure, and electrocardiographic changes were all earliest clinical signs of relapse. Despite the massive parenchymal and vascular infiltration and a rapid clinical condition worsening, no blast cells were identified on admission blood analyses.

Therefore, infiltration of thoracic structures and development of electric disturbance could be poor prognostic factors in recurrent AML.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Basso C, Rizzo S, Valente M, Thiene G. Cardiac masses and tumours. Heart 2016;102:1230-45.
2. Bussani R, De-Giorgio F, Abbate A, Silvestri F. Cardiac metastases. J Clin Pathol 2007;60:27-34.
3. Döhner H, Weisdorf DJ, Bloomfield CD. Acute myeloid leukemia. N Engl J Med 2015;373:1136-52.
4. Pasquini MC, Devine S, Mendizabal A, Baden LR, Wingard JR, Lazarus HM, et al. Comparative outcomes of donor graft CD34+ selection and immune suppressive therapy as graft-versus-host disease prophylaxis for patients with acute myeloid leukemia in complete remission undergoing HLA-matched sibling allogeneic hematopoietic cell transplantation. J Clin Oncol 2012;30:3194-201.
5. Walter RB, Buckley SA, Pagel JM, Wood BL, Storer BE, Sandmaier BM, et al. Significance of minimal residual disease before myeloablative allogeneic hematopoietic cell transplantation for AML in first and second complete remission. Blood 2013;122:1813-21.
6. Cates CU, Virmani R, Vaughn WK, Robertson RM. Electrocardiographic markers of cardiac metastasis. Am Heart J 1986;112:1297-303.
7. Zhu D, Yin S, Cheng W, Luo Y, Yang D, Lin K, et al. Cardiac MRI-based multi-modality imaging in clinical decision-making: Preliminary assessment of a management algorithm for patients with suspected cardiac mass. Int J Cardiol 2016;203:474-81.
8. De Lazzari M, Pedrigo M, Perazzolo Marra M, Calabrò F, Tarantini G, D’Amore EG, et al. Relapsing leukemia infiltrating the heart. Circ Heart Fail 2015;8:1133-4.
9. Kara IO, Sahin B, Paydas S, Kara B. Granulocytic sarcoma of the heart: Extramedullary relapse of acute myeloblastic leukemia after allogeneic stem cell transplantation successfully treated by chemotherapy alone. Leuk Lymphoma 2005;46:1081-4.
10. Makaryus AN, Tung F, Liu W, Mangion J, Kort S. Extensive neoplastic cardiac infiltration in a patient with acute myelogenous leukemia: Role of echocardiography. Echocardiography 2003;20:539-44.