Case report

Recurrent cardiac chloroma presenting as acute chest pain

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Learning Point for Clinicians

Granulocytic sarcoma should be a part of the differential diagnosis of patients who present with mass lesions after allogeneic transplant for chronic myeloid leukemia, even in the absence of hematologic relapse.

Case

A 41-year-old man presented to the hospital with sudden onset of chest tightness. His past medical history was notable for diagnosis of chronic myeloid leukemia (CML), made at 33 years. He was treated with imatinib, dasatinib and allogeneic peripheral blood stem cell transplant (PBSCT) with HLA mismatched, unrelated donor and he was still in complete hematologic response.

On admission, his vital signs and cardiac enzymes were normal. Right ventricular hypertrophy was newly developed compared with his previous electrocardiography. He got an urgent coronary angiography due to ongoing chest pain, and the result showed no significant stenosis but anomalous tumor feeding vessels from left circumflex artery. A huge branch of left circumflex artery was supplying the tumor, which seems to be consistent with development of chest discomfort, relative myocardial ischemia.1

Transthoracic echocardiography showed a 3.6 x 3.7 cm sized, irregular shaped mass in the right atrium (Figure 1A). The cardiac magnetic resonance image also revealed an intracardiac lobulating mass, which was considered as lymphoma at first, followed by chloroma and primary cardiac sarcoma.

To relieve his chest pain, he underwent en bloc resection of the mass. Cardiac mass revealed a neoplastic population consisting of myeloid blasts and megakaryocytes. Immunohistochemistry was positive for myeloperoxidase, CD34, C-kit, bcl-2, chloroacetate esterase stain (Figure 1B) but negative for CD3, CD10. These findings confirmed the mass as myeloid sarcoma.2 As there was no evidence of leukemic infiltration in other organs; the final diagnosis was isolated extramedullary relapse involving heart in CML patient after allogeneic PBSCT.

Because of no standard treatment guidelines for isolated extramedullary relapse of CML,3 dasatinib therapy was started with 100 mg/day after the operation. He had to reduce the dose to 60 mg/day, against the recurrent bilateral pleural effusion as the side effect of dasatinib.

Seventeen months after the operation, he visited the emergency department with the same pattern of chest discomfort. Echocardiography found a 2.3 x 3.1 cm sized, recurrent right atrial mass. His peripheral blood had no evidence of leukemia again this time. Considering the cardiac mass as a recurrent extramedullary relapse of CML, we escalated the dose of dasatinib upto 140 mg/day and planned to follow up the mass.
Discussion

Chloromas, also known as granulocytic sarcoma or myeloid sarcomas, are rare extramedullary tumors composed of immature myeloid cells. Although most cases of granulocytic sarcoma occur in acute myeloid leukemia as extramedullary relapse, it may occur during the course of myeloproliferative diseases. Extramedullary relapse following allogeneic stem cell transplantation was a rare phenomenon with a reported frequency of 0.45% in a large European multicenter survey, especially for CML. Most chloroma cases were described with bone marrow relapse commonly involving the central nervous system and skin. Cardiac relapse without bone marrow leukemia is exceedingly rare; there has been only one case report of pericardial involvement.

Imatinib is known to be nearly incapable of penetrating the extramedullary tissue; however, second generation tyrosine kinase inhibitors (TKIs) seem to influence extramedullary manifestations. A recent study showed long-lasting responses to dasatinib in patients with CNS involvement due to imatinib resistant CML.

Given the rarity of isolated extramedullary disease in particular, following allogeneic stem cell transplantation, the best treatment strategy remains unclear. Treatment options include systemic chemotherapy, donor lymphocyte infusion, repeat transplantation, or irradiation of localized lesion. However, long-term prognosis is generally poor.

This case has the following features: (i) this is the first report of isolated extramedullary relapse involving heart in CML patient after allogeneic PBSCT; (ii) the detection of recurrent cardiac chloroma was due to the coronary-tumor feeding vessel steal syndrome, which caused relative myocardial ischemia; and (iii) the second relapse was found while treating with second generation TKIs, dasatinib.

Conflict of interest: None declared.

References