

Fibrinous and hemorrhagic pericarditis with cardiac tamponade due to acute myeloid leukemia

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Conflict of interest

The Authors declare no conflict of interest.

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Dear Editor,

a 64-year-old man died after four days of fever, chest pain and dyspnea. Seventeen years before, he had received intensive consolidation chemotherapy and allogenic hematopoietic stem cell transplantation (allo-HSCT) for acute myeloid leukemia (AML). More recently, three years before death, he developed, on full donor marrow, a Jak2-positive myelodysplastic/myeloproliferative neoplasm, unclassifiable (MDS/MPN-U), for which he was receiving hydroxyurea. Two days before death, blood examination showed a high white blood cell count (111,700/mm³) with low platelet count (62,000/mm³) and low hemoglobin level (7.9 g/dl). The bone marrow biopsy revealed AML. At post-mortem examination, abundant hemorrhagic fluid was present in the pericardial sac. The heart was enlarged and the pericardium diffusely covered by villus-like projections (Fig. 1, left). Death was attributed to cardiac tamponade associated with hemorrhagic and fibrinous pericarditis. Acute myocardial infarction, trauma, surgery, uremia, infections, systemic diseases and neoplasia are the most common causes of fibrinous pericarditis. Histology revealed the pericarditis was the result of AML involving the pericardium and myocardium without forming a tumor mass, but with a rather subtle infiltrate composed by single cells, often within vascular spaces (Fig. 1, upper right), positive for myeloperoxidase (Fig. 1, lower right), with partial expression of CD68PGM1 (Fig. 2) and CD56. CD34, TdT, CD3, Cd79α, glycoforin and LAT were negative. Liver, spleen, brain, lungs and kidneys showed a similar picture of "intravascular" leukemia, consistent with the diagnosis of full-blown AML with systemic involvement.

AML represents the most common form of acute leukemia in adults ¹. The treatment involves chemotherapy to induce remission, followed by consolidation treatment with hematopoietic stem cell transplantation (HSCT) to reduce the risk of relapse ². Extramedullary relapse after allo-HSCT is considered a rather rare occurrence, ranging from 0.65% to over

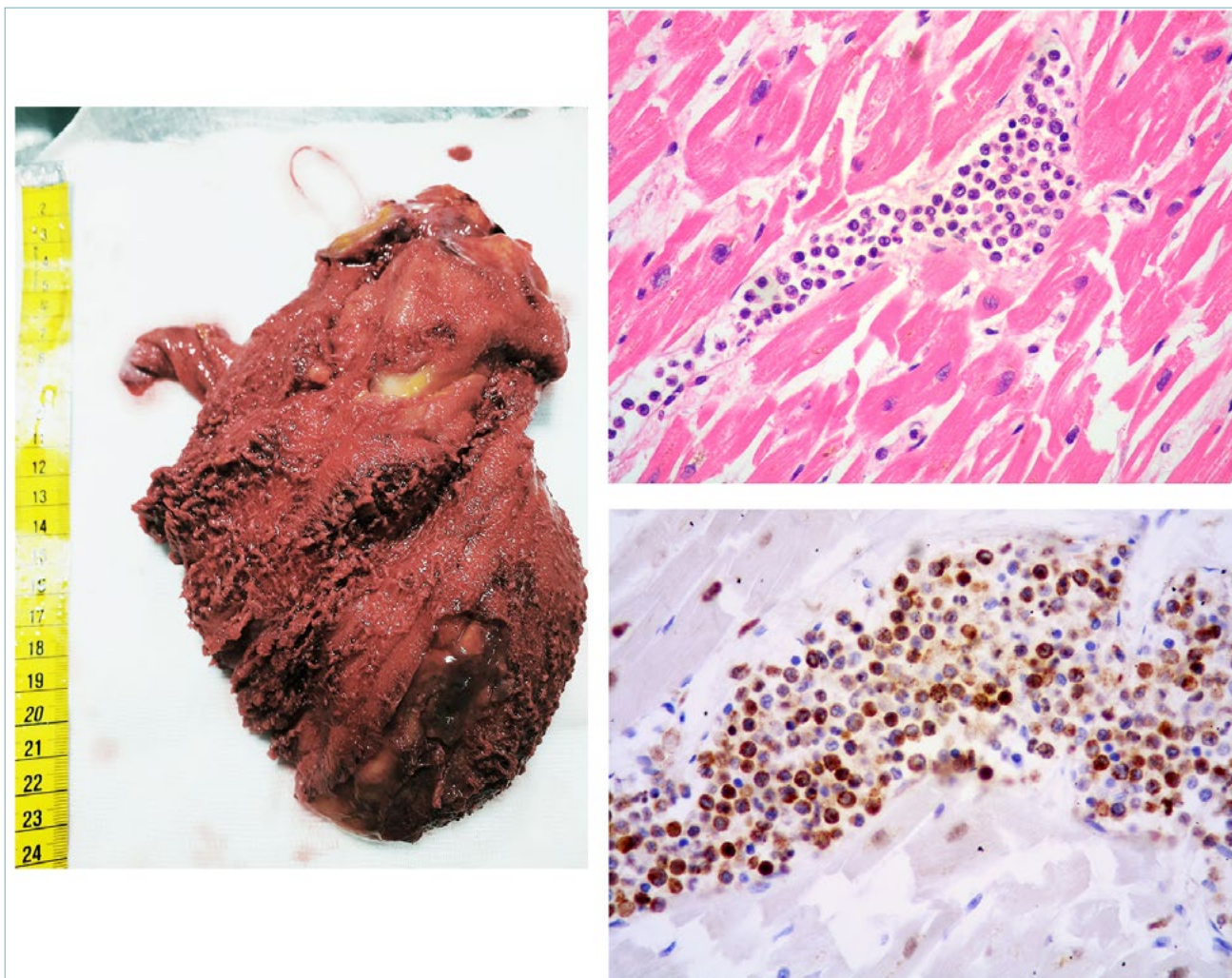


Figure 2. Figure 1. *Left* - Macroscopic view of the heart with the pericardium diffusely covered by villus-like projections; *upper right* - Single neoplastic cells identified within vascular spaces in the myocardium (HE 200x); *lower right* - Myeloperoxidase positivity of intravascular neoplastic cells (immunostain 200x).

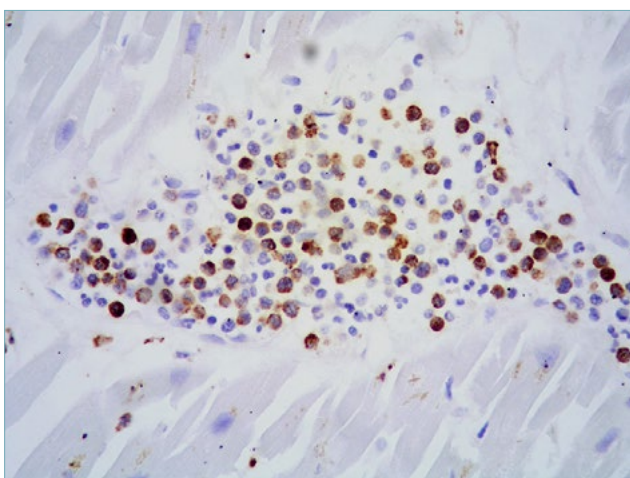


Figure 2. CD68PGM1 expression of intravascular neoplastic cells (immunostain 200x).

20%; most cases of extramedullary relapse after allo-HSCT are concomitant with bone marrow relapse³. Extramedullary relapse is reported to develop later than isolated bone marrow relapse and, although not frequently, it can occur even 5-10 years after HSCT³. Some interesting features are noted in the present case. AML developed on full donor marrow (transformed MDS/MPN-U), 17 years after allo-HSCT for AML. Post mortem showed multi-organ leukemic involvement and an impressive hemorrhagic and fibrinous pericarditis due to cardiac leukemia infiltration. Although pericardial effusions in leukemia patients are not unusual, such a severe pericarditis represents an uncommon event⁴. Neoplasms most often causing cardiac involvement include lung and breast cancer, melanoma and lymphomas.⁵ Cardiac relapse after

HSCT for leukemia is seldom reported and apparently mainly in acute lymphoblastic leukemia and more rarely in AML⁶⁻⁹. Additionally, in our case multi-organ infiltration by myeloid blasts did not form any tumor mass, and therefore could not be classified as myeloid sarcoma¹. Lastly, the intravascular pattern of AML is rather infrequently described and the differential diagnosis with other intravascular malignancies such as metastatic carcinoma or intravascular lymphoma is mandatory.

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Ethical approval

Local ethics committee (Comitato Etico dell'Area Vasta Emilia Nord, Italy) ruled that no formal ethics approval was required in this particular case.

Informed consent

Written informed consent was obtained from patient's relatives for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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