

# Cardiac Valve Granulocytic Sarcoma Infiltration as an Complicating Acute Myelogenous Leukemia: a Case Report

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## Abstract

Granulocytic sarcomas (chloromas) are rare extramedullary tumors consisting of primitive granulocytic cells. We report here on a case of a 23-year-old man who presented with a generalized swelling. He is known case of acute myelomonocytic leukemia. Granulocytic sarcomas are rare, destructive, extramedullary tumor masses that consist of immature granulocytic cells. In this unusual patient's case, the location and invasive nature of the tumor be important that can die the patient if we cannot treat patient urgently.

Optimal therapy for these patients has not been well defined: standard AML chemotherapy is moderately effective and should be considered for all suitable cases. To reduce the risk of subsequent ANLL in patients with nonleukemic GS, it is important that accurate histologic diagnosis is established initially for GS and that all isolated cases of GS, even those that appear to be cured by resection or irradiation of the tumor, are treated with intensive chemotherapy similar to that used to treat ANLL during the nonleukemic period as soon as possible.

**Key Words:** AML, Cardiac valve, Chloroma

## Introduction

Granulocytic sarcomas (chloromas) are rare extramedullary tumors consisting of primitive granulocytic cells. They arise de novo, or are associated with other hematologic disorders such as acute myeloid leukemia, myelodysplastic syndrome, or myeloproliferative disorders.

## Material and method

We report here on a case of a 23-year-old man who presented with a generalized swelling. He is known case of acute myelomonocytic leukemia .He treated at the first presentation with classic plan 7+3 (Cytosar and Danurubicine).

After induction and access to complete remission in first follow up patient refer to BMT center because he has a full match sibling donor. Unfortunately patient deny and after 3 months come back with leukocytosis and dyspnea, in cardiopulmonary exam and in echocardiography a fragile mass find in the tricuspid valve, patient refer to cardiac

surgery. The mass was confirmed by excisional biopsy of tricuspid valve to be a granulocytic sarcoma that documented by MPO and IHC.

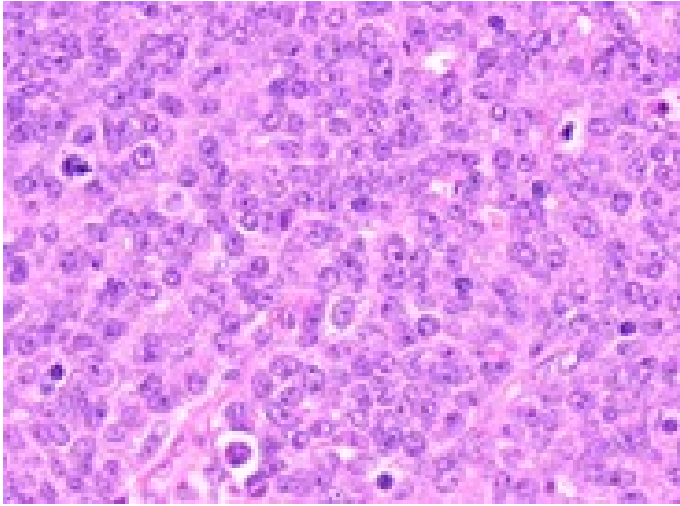
In Peripheral Blood Smear showed leukocytosis with immature cells and in Bone marrow examination hypercellularity with myeloblast and monoblast. However, cytogenetic examination of the marrow was normal.

At final patient retreated with salvage regimen High dose cytosar based regimen and refer to BMT center (Figure- 1). Immunohistochemistry was positive for CD33, 34, 43, 45, and 68. Ki67 was 80%.

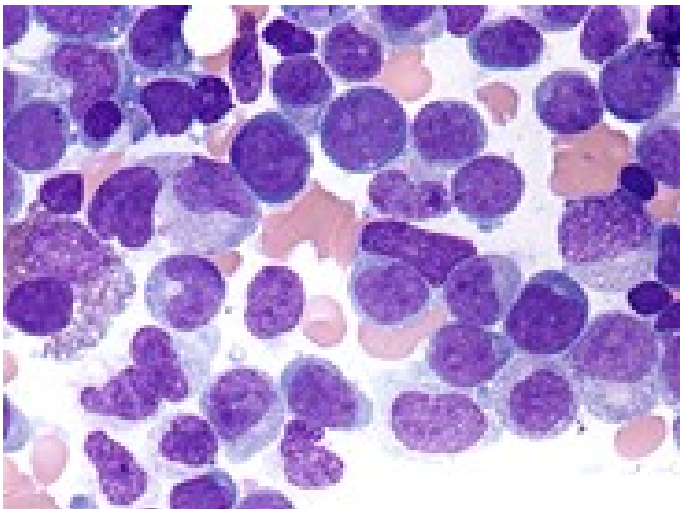
This was consistent with a myeloid malignancy- an extramedullary myeloid tumor.

Bone marrow aspirate was performed to look for any evidence of an increased myeloblast population. Results showed only mild myeloid and eosinophilic hyperplasia (Figure- 2).

There was no evidence of acute myeloid leukemia. (BM immunophenotyping - CD34 blasts < 1%).



**Figure- 1.** Tru-cut biopsy histology of the tumor.



**Figure- 2.** Bone marrow aspirate morphology.

The final diagnosis was confirmed as bone marrow AML, with the presenting feature of the disease as unifocal extramedullary granulocytic sarcoma transformation.

### Discussion

Granulocytic sarcomas are rare, destructive, extramedullary tumor masses that consist of immature granulocytic cells. The term “chloroma” is derived from the Greek word chloros (for green). It describes the tumor's frequently greenish color, which is due to the presence and oxidation of the enzyme myeloperoxidase in the tumor. These tumors can arise de novo or can be associated with other myeloid disorders- acute myeloid leukemia (AML) or CML, myeloproliferative, or myelodysplastic conditions. Presentation can occur prior to, in association with the underlying myeloid disorder, or upon relapse. The location of the tumor varies- eg, subperiosteal bone- skull, pelvis, ribs, sternum; lymph node; skin; gums. In this case, the granulocytic sarcoma was a large tumor mass that caused symptoms due to cardiac valve obstruction.

The WHO(1) has classified granulocytic sarcomas into 3 main types, depending upon the degree of maturation:

- Blastic– composed mainly of myeloblasts
- Immature– myeloblasts and promyelocytes
- Differentiated– promyelocytes and more mature myeloid cells

Rarer types can consist of a monoblastic sarcoma, associated with monoblastic leukemia.

The diagnosis of granulocytic sarcoma can be difficult. As in this case, the disease is often not suspected on clinical grounds and a high index of suspicion is needed. Clinically and histologically, the diagnosis needs to be distinguished from the main differentials- ie, Hodgkin lymphoma, Burkitt's lymphoma, large cell lymphomas, and small round blue cell tumors, such as neuroectodermal tumors.

In this unusual patient's case, the location and invasive nature of the tumor be important that can die the patient if we cannot treat patient urgently. Full investigations into the morphology, immunohistochemistry, immunophenotyping, and cytogenetics are essential as the diagnosis may be inconclusive on the basis of morphology alone. The morphology of the cells can be variable. If well differentiated, a diffuse infiltrate of granulocytic cells can be seen (often containing all stages of myeloid cell maturation). If poorly differentiated, the majority of the cells are a large and monotonous population. The nucleoli may or may not be prominent. Cytoplasm is often scanty. A high mitotic index can be seen.(2) Dissection of collagen by strings of neoplastic cells is also a common feature. In lymph nodes, granulocytic sarcoma may show paracortical or sinus infiltration, or efface the architecture with a diffuse infiltrate. Bone marrow investigations need to be performed to look for any underlying myeloid conditions, as in this case. Tissue both from the tumor and the marrow needs to be examined as the cellularity and phenotype can differ between them.

Certain risk factors for developing granulocytic sarcomas associated with any underlying myeloid disorder have been recognized:

- FAB types M2, M4, and M5 of the underlying leukemia
- Expression of the surface markers CD 2, 7, and 56
- Cytogenetic abnormalities t(8;21) and inv(16),(3)

Optimal therapy for these patients has not been well defined: standard AML chemotherapy is moderately effective and should be considered for all suitable cases. The addition of radiotherapy can provide a survival benefit, especially in CNS-related tumors. Chromosome abnormalities are

associated with a worse prognosis. Intensive therapy should be considered early, especially for patients with chromosome 8 abnormalities.(4)

Patients in relapse after chemotherapy or stem cell transplant can also present with a granulocytic sarcoma.(6)

### Conclusion

To reduce the risk of subsequent ANLL in patients with nonleukemic GS, it is important that accurate histologic diagnosis is established initially for GS and that all isolated cases of GS, even those that appear to be cured by resection or irradiation of the tumor, are treated with intensive chemotherapy similar to that used to treat ANLL during the nonleukemic period as soon as possible.

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