

# Leukaemic Ascites and Peritoneal Myeloid Sarcoma: Rare but Not Impossible

Luís Landeiro<sup>1</sup>, Tiago Neto Gonçalves<sup>1</sup>, Margarida Proença<sup>1</sup>, Albertina Nunes<sup>2</sup>, Alexandra Bayão Horta<sup>1</sup>

<sup>1</sup>Internal Medicine Department, Hospital da Luz Lisboa, Lisbon, Portugal

<sup>2</sup>Hematology Department, Instituto Português de Oncologia Francisco Gentil, Lisbon, Portugal

Doi: 10.12890/2022\_003184 - European Journal of Case Reports in Internal Medicine - © EFIM 2022

Received: 11/01/2022

Accepted: 14/01/2022

Published: 16/02/2022

**How to cite this article:** Landeiro L, Neto Gonçalves T, Proença M, Nunes A, Bayão Horta A. Leukaemic ascites and peritoneal myeloid sarcoma: rare but not impossible. *EJCRIM* 2022;9: doi:10.12890/2022\_003184.

**Conflicts of Interests:** The authors declare there are no competing interests.

This article is licensed under a [Commons Attribution Non-Commercial 4.0 License](https://creativecommons.org/licenses/by-nc/4.0/)

## ABSTRACT

A myeloid sarcoma is an extramedullary tumour arising from infiltration by leukemic cells at an anatomic site other than the bone marrow. Most commonly it precedes acute myeloid leukaemia but occasionally occurs simultaneously. It may also be associated with myeloproliferative neoplasms, myelodysplastic syndrome and the blast phase of chronic myeloid leukaemia.

The most common sites for extramedullary tumours are bone, periosteum, soft tissue, lymph node and skin. Although this disease can affect a wide range of body sites, there are very few reports of peritoneal myeloid sarcoma or cavity effusion.

The authors present the case of a 68-year-old man with myelodysplasia-related acute myeloid leukaemia and peritoneal myeloid sarcoma with myeloid ascites. The definitive diagnosis is challenging, requires a high level of suspicion, and relies on the exclusion of all alternative diagnoses and especially on complementary tests such as flow cytometry and immunohistochemistry analysis of ascitic fluid in order to detect the immature myeloid cells.

## LEARNING POINTS

- Myeloid sarcomas are extramedullary leukemic tumours that occur before or simultaneously with acute myeloid leukaemia, other myeloproliferative neoplasms or myelodysplastic syndrome.
- Myeloid sarcomas are most often seen in bone, soft tissue, lymph node and skin, but can present in most locations.
- Peritoneal myeloid sarcoma and leukemic ascites, although very rare, must be searched for when a patient with acute leukaemia presents with newly diagnosed ascites, through ascitic fluid flow cytometry and immunophenotypic analysis.

## KEYWORDS

Peritoneal myeloid sarcoma, leukemic ascites

## INTRODUCTION

Acute myeloid leukaemia (AML) is a malignant neoplasm characterized by clonal proliferation of myeloid precursors with a reduced capacity to differentiate into more mature cellular elements. It most often involves the bone marrow (BM) and peripheral blood and rarely extramedullary tissue<sup>[1,2]</sup>.

A myeloid sarcoma (MS), also called granulocytic sarcoma or chloroma, is an extramedullary proliferation of the blasts of one or more of the myeloid lineages that disrupt the normal architecture of the tissue in which it is found. It most frequently precedes, but may occur simultaneously with, bone marrow AML. It is also associated with myeloproliferative neoplasms, myelodysplastic syndrome and the blast phase of chronic myeloid leukaemia<sup>[1,3]</sup>.

The most common sites are bone, periosteum, soft tissue, lymph node and skin. Although this disease can affect most body locations, there are very few reports of presentation as peritoneal myeloid sarcoma or body cavity effusion [4–10].

We report the case of a 68-year-old man with a previous diagnosis of myelodysplasia-related acute myeloid leukaemia, now presenting with peritoneal myeloid sarcoma with ascites.

#### CASE DESCRIPTION

A 68-year-old man presented to the Emergency Department with a 4-week clinical picture of nausea, increasing abdominal volume, persistent abdominal discomfort, and fatigue with little exertion. The patient had a diagnosis of acute myeloid leukaemia, with myelodysplasia-related changes (karyotype 48XY, chromosome 8 trisomy, without variant mutations of NucleoPhosMin 1 (NPM1) and Fms-Like Tyrosine kinase 3 (FLT3)). He was treated initially with four cycles of azacitidine without achieving remission. Second-line treatment was instituted with fludarabine+high-dose cytarabine+granulocyte colony-stimulating factor (FLAG), remission was achieved and a consolidation course with high dose cytarabine was completed.

In addition to haematological disease, the patient had a medical history of cutaneous sarcoidosis, hypertension and diabetes.

On admission, he was haemodynamically stable, tachycardic at 106 bpm and afebrile. He was pale, dehydrated, and had tense ascites with prominent shifting dullness, as well as mild feet oedema. There were no features suggestive of chronic liver disease.

Peripheral blood tests showed a white blood cell count of 11,480/mm<sup>3</sup> with 90% neutrophils and no circulating blasts, haemoglobin of 8.4 g/dl (similar to recent previous evaluations), a platelet count of 142,000/mm<sup>3</sup>, acute renal failure (serum creatinine 2.81 mg/dl, serum urea 144 mg/dl) without acid base or ionic abnormalities, serum lactate dehydrogenase (LDH) of 2482 IU/l, unremarkable liver tests, and C-reactive protein of 10.5 mg/dl.

Abdominal and pelvic computed tomography revealed massive ascites in association with mesenteric densification and heterogeneity, vascular congestion, and micronodularity suggestive of secondary deposits (Fig. 1).

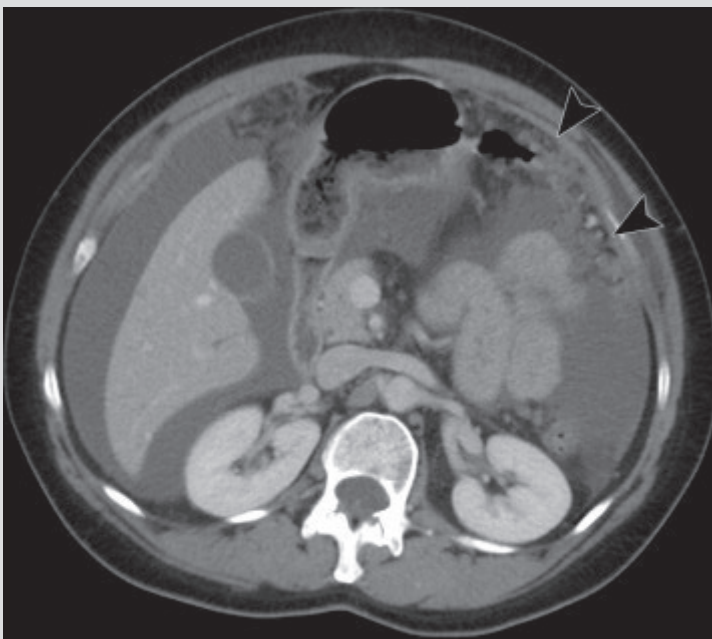


Figure 1. CT scan of the abdomen and pelvis. Black arrowheads indicate peritoneal secondary deposits

A diagnostic and therapeutic paracentesis was performed. Analysis of the ascitic fluid revealed a white blood cell count of 28,680 cells/ml, no cellular predominance but significant cellular destruction (LDH 12,364 IU/l), a serum ascitic albumin gradient (SAAG) below 1.1, protein concentration of 4.1 g/dl, and glucose of 110 mg/dl. Considering the highly inflammatory ascitic fluid, a very high risk of superimposed bacterial infection, and the presence of criteria for the diagnosis of spontaneous bacterial peritonitis, the patient was started on antibiotic treatment with cefotaxime while awaiting the results of bacteriological, mycobacteriological and cytological tests.

A bone marrow aspirate was performed and analysis showed hypercellularity with increased blasts (>50%), revealing acute leukaemia relapse. A bone marrow biopsy showed a hypercellular bone marrow (90%), predominantly occupied by myeloblasts and myeloid precursors (immunophenotypic analysis positive for myeloperoxidase (MPO) and CD117; CD34 negative).

The ascitic fluid cytological/immunophenotypic analysis showed a markedly elevated number of immature granulocyte precursors (MPO+/CD20-), interpreted as myeloblasts. Simultaneously, flow cytometry analysis of the ascitic fluid demonstrated that 60% of the cells were immature myeloid cells which expressed HLA-DR, CD117 and CD45 (CD34 negative), compatible with an extramedullary form of acute myeloid leukaemia. The mesenteric structural changes were therefore interpreted as myeloid sarcomas.

Work-up staging showed there was no clinical, laboratory or radiological evidence of another site of solid tissue extramedullary proliferation. Taking into account the AML relapse after two lines of chemotherapy and accounting for the patient's frailty, an extensive discussion with his attending haematologist took place and a decision was made to provide best supportive care through optimal palliative care. The patient died 12 days after admission.

## DISCUSSION

A myeloid sarcoma is an extramedullary proliferation of the blasts of one or more myeloid lineages that disrupt the normal architecture of the tissue in which it is found. It is an uncommon manifestation of AML that may occur before, during or after diagnosis, and may arise secondary to myeloproliferative neoplasms, myelodysplastic syndrome or a blast phase of chronic myeloid leukaemia (CML) [1,2]. It can also occur as a relapse, especially in recipients of allogeneic haematopoietic stem cell transplantation (HSCT) [6,11].

The most common sites of involvement are bone, periosteum, soft tissue, lymph node and skin. Presentation as peritoneal myeloid sarcoma or a body cavity effusion is rare [4-10] and mainly reported in AML with monocytic differentiation, including M4 and M5 AML (FAB classification) [12,13].

The cytogenetic and molecular abnormalities associated with myeloid sarcoma are similar to those seen in leukemic AML. Abnormalities in chromosome 8, in particular t(8;21) and chromosome 8 trisomy, have been described as more frequent in myeloid sarcoma compared with leukemic AML [14]. Our patient had chromosome 8 trisomy.

As the diagnosis of this entity is challenging and requires a high level of suspicion, the true incidence of myeloid sarcoma involving peritoneal, pericardial and pleural fluids may be underestimated due to the underuse of flow cytometry and immunohistochemistry for its evaluation. These immunoassays are needed to identify immature myeloid cells, which is a requirement for definitive diagnosis. In this case, these methods were used to further support the hypothesis of myeloid sarcoma with myeloid ascites.

The diagnosis of peritoneal myeloid sarcoma can be also be difficult as it is often confused with other causes of peritoneal primary or secondary neoplasms, predominantly sarcomas, melanomas or carcinoid tumours, as well as with spontaneous bacterial peritonitis (something that initially occurred in our patient's case) [3,4,13,14]. All other relevant causes of ascites must be excluded.

Our case illustrates the diagnostic challenges associated with myeloid sarcoma, as the clinical, laboratory and radiological findings were initially perceived as due to an infectious or other malignant disease. The fact that our patient's ascites had cells with immature myeloid markers, in conjunction with evidence of disease relapse in the bone marrow, revealed the diagnosis, regardless of the alternative hypothesis initially presumed.

Unfortunately, the optimal treatment of myeloid sarcoma is unclear, but systemic treatment of the disease is currently the best option; there are no randomized controlled trials as this patient population is small [5,14]. Our patient was already on third-line systemic treatment for AML, and therefore no other viable therapeutic option was available.

This report is intended to highlight an uncommon manifestation of a common disease. As myeloid sarcoma may present in very unusual sites, it is essential to perform an extensive work-up when it is suspected, especially in patients with a history of AML.

---

## REFERENCES

1. Arber DA. The 2016 WHO classification of acute myeloid leukemia: what the practicing clinician needs to know. *Semin Hematol* 2019;**56**(2):90–95.
2. Narayanan D, Weinberg OK. How I investigate acute myeloid leukemia. *Int J Lab Hematol* 2020;**42**(1):3–15.
3. Almond LM, Charalampakis M, Ford SJ, Gourevitch D, Desai A. Myeloid sarcoma: presentation, diagnosis, and treatment. *Clin Lymphoma Myeloma Leuk* 2017;**17**(5):263–267.
4. Neiman RS, Barcos M, Berard C, Bonner H, Mann R, Rydell RE, et al. Granulocytic sarcoma: a clinicopathologic study of 61 biopsied cases. *Cancer* 1981;**48**(6):1426–1437.
5. Paydas S, Zorludemir S, Ergin M. Granulocytic sarcoma: 32 cases and review of the literature. *Leuk Lymphoma* 2006;**47**(12):2527–2541.
6. Frietsch JJ, Hunstig F, Wittke C, Junghans C, Franiel T, Scholl S, et al. Extra-medullary recurrence of myeloid leukemia as myeloid sarcoma after allogeneic stem cell transplantation: impact of conditioning intensity. *Bone Marrow Transplant* 2021;**56**(1):101–109.
7. Prolla JC, Kirsner JB. The gastrointestinal lesions and complications of the leukemias. *Ann Intern Med* 1964;**61**:1084–1103.
8. Khan MY, Hussein KK, Walter MG, Hasan MK, Kern W, Kharfan-Dabaja MA. Granulocytic sarcoma presenting with malignant anasarca in a patient with secondary acute myeloid leukemia. *Int J Hematol* 2004;**79**(3):250–252.
9. Shinagare AB, Krajewski KM, Hornick JL, Zukotynski K, Kurra V, Jagannathan JP, et al. MRI for evaluation of myeloid sarcoma in adults: a single-institution 10-year experience. *Am J Roentgenol* 2012;**199**(6):1193–1198.
10. Meyer HJ, Pönisch W, Schmidt SA, Wienbeck S, Braulke F, Schramm D, et al. Clinical and imaging features of myeloid sarcoma: a German multicenter study. *BMC Cancer* 2019;**19**(1):1–8.
11. Campidelli C, Agostinelli C, Stitson R, Pileri SA. Myeloid sarcoma: extramedullary manifestation of myeloid disorders. *Am J Clin Pathol* 2009;**132**(3):426–437.
12. Yonal I, Kayar Y, Nazligul E, Yenerel MN, Kalayoglu-Besisik S. Leukemic ascites as an initial presentation of acute myelomonocytic leukemia with inversion of chromosome 16. *Ann Saudi Med* 2013;**33**(2):197–198.
13. Pantanowitz L, Steingart R, Miller KB, Kruskal JB, Pihan G. Leukemic ascites. *Arch Pathol Lab Med* 2005;**129**(2):262–263.
14. Ravulapati S, Siegel C, Dara A, Lionberger JM. Myeloid sarcoma without circulating leukemia mimicking gastrointestinal malignancy and lymphoma. *Hematol Rep* 2018;**10**(2):53–55.