

## Discovery of A Myeloid Sarcoma in A Hemodialysis Patient: Don't Miss the Prior Hematological History!

Anibar S<sup>1\*</sup>, Racu L-M<sup>2</sup>, Trepant A-L<sup>2</sup>, Simon I<sup>1</sup>, Baudoux T<sup>1</sup>, Buntinx S<sup>3</sup>, Nortier J<sup>1</sup>

Depts of <sup>1</sup>Nephrology, <sup>2</sup>Pathology and <sup>3</sup>Hematology, Erasme Hospital, Université libre de Bruxelles, route de Lennik 808, 1070 Brussels, Belgium

DOI: [10.36347/sjams.2020.v08i02.059](https://doi.org/10.36347/sjams.2020.v08i02.059)

| Received: 13.02.2020 | Accepted: 20.02.2020 | Published: 27.02.2020

\*Corresponding author: Anibar Sara

### Abstract

### Case Report

Myeloid sarcoma (MS) is a rare tumor consisting of immature cells from granulocytic lineage and usually affecting bones. Other localizations are exceptional. We report the case of a 63 years old woman treated by chronic hemodialysis (HD) for end-stage kidney disease of multifactorial origin (diabetes and hypertension). She has been successfully treated for acute myeloblastic leukemia by intensive chemotherapy (induction with daunorubicin and aracytin, followed by FLT3 inhibitors as consolidation), for 11 months. After 9 months of complete remission, the patient presented with bilateral lymph nodes attributed to dental extraction. Further exams highlighted a solid submandibular mass, which was further biopsied. Histological analyzes detected the presence of myeloid cells (optical microscopy) with a positive immunostaining for myeloperoxidase, leading to the diagnosis of MS. A craniofacial CT scan did not exhibit local extension of the tumor. However, a hematological control revealed medullar blastic invasion (52% of blastic cells), confirming the diagnosis of leukemia relapse. The evolution was pejorative despite local radiotherapy and the patient deceased two months later. MS is an uncommon proliferation of immature myeloid cells occurring in any extramedullary organ. According to WHO 2016, it is considered as a form of myelodysplastic syndrome. Its incidence is similar in both genders and frequently seen in young patients under the age of 15 years. MS may precede or be concomitant to myeloproliferative/myelodysplastic syndromes, or even reveal these hematological disorders. Diagnosis is based on histological data, in particular myeloperoxidase immunofluorescence. To our knowledge, this is the first case reported in a HD patient. The diagnosis of MS must be considered by the clinician faced to any clinical or radiological lesion in a patient with a prior history of myeloproliferative syndrome.

**Keywords :** Chronic renal failure, hemodialysis, myeloid sarcoma, myelodysplastic leukemia, myeloproliferative syndrome.

**Copyright @ 2020:** This is an open-access article distributed under the terms of the Creative Commons Attribution license which permits unrestricted use, distribution, and reproduction in any medium for non-commercial use (NonCommercial, or CC-BY-NC) provided the original author and source are credited.

## INTRODUCTION

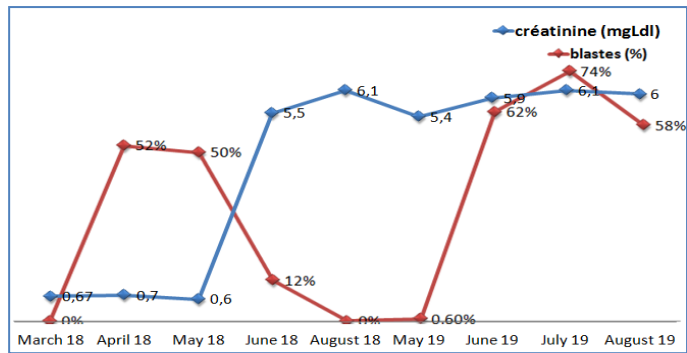
Myeloid sarcoma (MS), also called granulocytic sarcoma, is a rare tumor consisting of immature cells from granulocytic lineage. It often affects bones but other localizations are exceptional, making its diagnosis difficult in the absence of ongoing hematological disorders [1, 2].

## CASE REPORT

We report the case of a 63 years old woman with a prior history of diabetes and hypertension for 4 years. She was initially admitted in our hospital for an acute respiratory symptom and a blood test revealing leucosis and a tumor lysis syndrome. A myelogram was rapidly performed and showed a medullar invasion by

52% blasts cells with marked dysplasia. The diagnosis of acute myeloblastic leukemia on chronic myelodysplastic syndrome was then retained. The karyotype was normal (NPM1, NRAS -, FLT3 +, CEBPA). The patient received an intensive chemotherapy (induction with daunorubicin and aracytin, followed by FLT3 inhibitors as consolidation), for 11 months. The evolution was favorable, as attested by the control myelogram free of any blast cells.

In parallel, renal function parameters rapidly deteriorated: the patient developed acute kidney injury with anuria as a consequence of the initial tumor lysis syndrome but also probably in relation with chronic lesions secondary to nephroangiosclerosis (Figure 1).

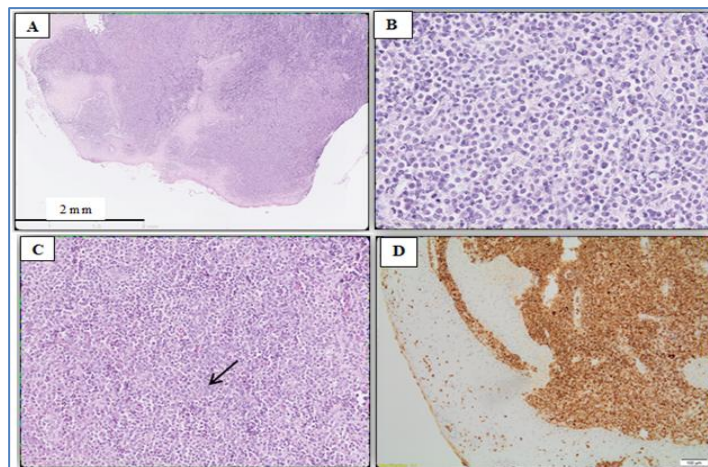


**Fig-1: Time course of serum creatinine and blast proliferation**

Kidney biopsy could not be done because of coagulation abnormalities (low platelet count). Continuous veno-venous hemofiltration was started for several days and the patient was shifted to conventional iterative hemodialysis (HD) (3 sessions per week).

After 9 months of clinical and hematological remission, the patient presented with bilateral lymph nodes attributed to dental extraction. Despite targeted

antibiotics, no clinical improvement was observed. Stomatological and otorhinolaryngological exams highlighted a solid submandibular mass, which was further biopsied. Histological analyzes detected the presence of myeloid cells (optical microscopy) with a positive immunostaining for myeloperoxidase and negative one for CD20, leading to the diagnosis of MS (Figure 2A-D).



**Fig-2: Biopsy of the submandibular mass: diffuse infiltration of the subepithelial tissue by a dense infiltration (A) composed of tumoral cells suggestive of myeloid cells (B) and immature blast cells (arrow) (C) at high magnification. Positive immunostaining for myeloperoxidase (D)**

A craniofacial CT scan did not exhibit local extension of the tumor (Figure 3).



**Fig-3: Craniofacial CT**

However, a control myelogram revealed a recurrent blast invasion (62% of blast cells), confirming the diagnosis of leukemia relapse (Figure 1). The evolution was pejorative despite local radiotherapy and the patient deceased two months later.

## DISCUSSION

Described for the first time in 1811 by Burns, MS was initially called chloroma, referring to the green color of the tumor caused by high levels of myeloperoxidase in the cells [1, 3]. However, considering the fact that this type of tumor is not always green, the term granulocytic sarcoma seems to be a more appropriate term [4].

It is a rare tumor, consisting of immature cells from granulocytic lineage, occurring in any extramedullary organ [1]. According to WHOM 2016,

it is considered as a form of myelodysplastic syndrome [2, 5]. MS may precede or be concomitant to myeloproliferative / myelodysplastic syndromes, or even reveal these hematological disorders. It may also reflect the relapse of such syndromes, which was the case of our patient [2, 5]. The incidence of MS ranges from 2 to 9% in adults; it is similar for both genders and frequently seen in young individuals (under the age of 15 years) [1, 5].

The clinical presentation is variable, as an extra-medullary mass, preferentially in the bone, but also in other tissues [1, 5]. The diagnosis of MS is based on histological analyses: upon optical microscopy, a proliferation of myeloid cells at different stages of maturation is observed, with a positive myeloperoxidase immunofluorescence, which is pathognomonic [1, 3, 5-7]. Other immunostainings may be required, such as anti-CD 20 and anti-CD 117 in order to rule out the diagnosis of lymphoma, the main differential diagnosis [3, 6-8]. Therapeutic options are those of acute myeloid leukemia [1, 3, 9]. The prognosis of MS is reserved, depending on its response to chemotherapy, the severity of the clinical presentation, and then the frequency of relapses [1, 4, 9-11].

To our knowledge, this is the first case of MS reported in a HD patient. In fact, association of MS with end-stage kidney disease is most likely a coincidence. However, as uremia is usually considered as a factor of immune dysfunction, severe chronic kidney failure could have precipitated leukemia relapse in the present case.

## CONCLUSION

The diagnosis of MS must be considered by the clinician faced to any clinical or radiological lesion, most particularly in a patient with a prior history of myeloproliferative syndrome.

## REFERENCES

1. Kaplan J, Dardanelli E, Sierre S, Moguillansky S, Lipsich J. Granulocytic sarcoma of the urinary bladder in a pediatric patient. *Journal of Pediatric Urology*. 2013; 9:9-11.
2. Arber DA, Orazi A, Hasserjian R, Thiele J, Borowitz MJ, Le Beau MM. The 2016 revision to the World Health Organization classification of myeloid neoplasms and acute leukemia. *Blood*. 2016; 127:2391-2405.
3. Mottaz P, Becot-Bretez F, Jeanneau R, Vignon G, Bertin A, Carrere F, Augerau PF, Aucher P, Lellouche F. Localisation caecale d'un sarcome myéloïde a-leucémique : un diagnostic difficile. *Annals of Pathology*. 2004;24; 436-439.
4. Koudstaal MJ, Van der Wal KGH, Lam KH, Meeuwisc CA, Spelemanc L, Levind MD. Granulocytic sarcoma (chloroma) of the oral cavity: Report of a case and literature review. *Oral Oncology*. 2006; 42(2):70-77.
5. Mottaz P, Becot-Bretez F, Jeanneau R, Vignon G, Bertin A, Carrere F, Augerau PF, Aucher P, Lellouche F. Sarcome granulocyttaire: à propos de 2 cas et revue de la littérature. *Revue de médecine interne*. 2017. 38;774-777.
6. Klco JM, Welch JS, Nguyen TT, Hurley MY, Kreisel FH, Hassan A. State of art in myeloid sarcoma. *Int J Lab Hematol*. 2011; 33:555-565.
7. Quintanilla Martinaz L, Zukerberg LR, Ferry JA, Harris NL. Extramedullary tumors of lymphoid or myeloid blasts. *J Clin Pathol*. 1995; 104:431-41.
8. Menasce LP, Banejee SS, Beckett E, Harris M. Extramedullary myeloid tumor (granulocytic sarcoma) is often misdiagnosed : a study of 26 cases. *Histopathology*. 1999; 34:391-398.
9. Xie Z, Zhang F, Song E, Ge W, Zhu F, Hu J. Intraoral granulocytic sarcoma presenting as multiple maxillary and mandibular masses: A case report and literature review. *Oral Surg, Oral Med, Oral pathol, Oral Radiol, and Endod*. 2007; 103(6):44-48.
10. Suzer T, Colakoglu N, Cirak B, Keskin A, Coskun E, Tahta K. Intracerebellar granulocytic sarcoma complicating acute myelogenous leukemia: a case report and review of the literature. *J Clinical Neuroscience*. 2004; 11(8):914-917.
11. Smith ML, Hills RK, Grimwade D. Independent prognostic variables in acute myeloid leukemia. *Blood Rev*. 2011; 25(1):39-51.