

# Myeloid Sarcoma (Chloroma) An Uncommon Entity



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**Submission:** August 11, 2020; **Published:** August 19, 2020

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## Abstract

Granulocytic Sarcoma or Myeloid Sarcoma (MS) is a solid tumor of myeloid origin and extramedullary location. Its incidence is estimated between 2.5 - 9.11% of patients with Acute Myeloid Leukemia, 60% of MS cases occur in children under 15 years of age. The clinical characteristics depend on its location and range from an asymptomatic lesion to organ dysfunction due to infiltration. It appears as a circumscribed or diffuse soft tissue mass infiltrating the bone, skin and/or peritoneum. Definitive diagnosis requires biopsy and immunohistochemical study, the most sensitive markers are CD43 and lysozyme, with expression in up to 100% of cases, myeloperoxidase, is expressed in 66-96% of cases. Treatment includes radiation therapy, surgery, or both, combined with a specific systemic chemotherapy regimen or allogeneic bone marrow transplantation. Recognition of this entity, a timely diagnosis and early treatment is a diagnostic challenge when it occurs in isolation.

**Keywords:** Granulocytic Sarcoma; Myeloid Sarcoma; Chloroma; Leukemia Mieloid Acute; Tumor

**Abbreviations:** GS: Granulocytic Sarcoma; MS: Myeloid Sarcoma; LMA: Leukemia Mieloid Acute; MPO: Myeloperoxidase

## Introduction

Granulocytic Sarcoma (GS), Myeloid Sarcoma (MS) or also known as Chloroma is a solid tumor of myeloid origin and extramedullary location [1,2]. The term Chloroma (green tumor) is derived from the greenish staining of the lesion attributable to myeloperoxidase in cells of the granulocytic lineage first described by Burns in 1811. In 2002, the World Health Organization abandoned its original name and gave it its current nomenclature (Myeloid Sarcoma) [3]. Its estimated incidence is 2.5 - 9.11% of patients with Acute Myeloid Leukemia (AML), especially the M2 subtype, 60% of cases occur in children under 15 years of age. [4-7] Common sites of appearance include but are not limited to the skin, soft tissues, bone, peritoneum, lymph nodes, and digestive tract. Additionally, one can find lesions around the orbit, skull, and epidural spaces [2,7,8]. Its formation is believed to be due to extramedullary hematopoiesis secondary to proliferation of hematopoietic progenitor cells and an increase in the expression of the adhesion molecule CD56 in malignant myeloid cells, associated with the genetic translocation t (8; 21) [9].

## Diagnosis

### Clinical characteristics

Asymptomatic presentations are reported in up to 50% of patients. The clinical characteristics of MS depend on its location, where the tumor can create a displacing effect or organ dysfunction due to infiltration and tumor burden [10].

### Radiographic characteristics

MS usually appears as a diffuse or circumscribed soft tissue mass. Computed Tomography is the initial study of choice; however, Magnetic Resonance Imaging is preferred when central nervous system involvement is suspected [11,12].

### Histopathological characteristics

Definitive diagnosis of MS requires biopsy and immunohistochemical studies. Morphologically, infiltration of immature cells of myeloid lineage is observed, receiving their name according to the myeloid cell involved. The diagnosis is

confirmed with an immunohistochemical study by observing surface markers such as CD68-KP1, myeloperoxidase (MPO), CD117, CD99, CD68 / PG-M1, lysozyme, CD34, CD56, CD61, CD30, CD4, CD43, CD45. In 2008, the WHO established the stains required for the diagnostic study for MS and these should include chloroacetate esterase, myeloperoxidase, and non-specific esterase [13-15].

It has been reported that the most sensitive markers for MS are CD43 and lysozyme, with expression in up to 100% of cases, however, they are not specific markers. MPO, which is expressed in 66-96% of MS cases, is used to differentiate MS from lymphoma. CD68 and CD117 are also part of the panel most used for diagnosis [13,16]. Cytogenetic alterations are found in approximately 50% of patients. The most common mutation described in MS is mutated NPM1. The most common translocation is t (8; 21). Inv (16) is also a common abnormality associated with extramedullary disease in AML [17-19].

### Treatment

Treatment depends on the time of presentation of the disease, that is, if it is an isolated tumor, concomitant with AML or if it presents as a relapse of AML. Treatment includes radiation therapy, surgery, or both, combined with systemic chemotherapy specific to AML. Allogeneic bone marrow transplantation is also a therapeutic option in achieving remission of the disease or in case of relapse [19-23].

### Conclusion

MS is a rare soft tissue tumor that should be considered in a patient with AML for timely diagnosis and early treatment; it is a diagnostic challenge when it occurs in isolation.

### Conflict of interest

The authors declare that there is no conflict of interest to disclose.

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DOI: [10.19080/JOJOOS.2020.02.555597](https://doi.org/10.19080/JOJOOS.2020.02.555597)

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