

# Myeloid Sarcoma in Brain and Optic Nerve Presented as a Relapse of Acute Myeloid Leukemia: A Case Report

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### **Author’s contributions:**

Mais Musleh: design of the study, data collection, data interpretation and analysis, drafting, critical revision, and the approval of the final manuscript.

Samer Musleh: data interpretation and analysis, drafting, critical revision, and the approval of the final manuscript.

Manal Shakhee: data interpretation and analysis, drafting, critical revision, and the approval of the final manuscript.

### **Abstract:**

Myeloid sarcoma (MS) is a rare extramedullary infiltration of Acute Myeloid Leukemia (AML). We present a case of 19-year-old male with AML-M2 who relapse with AML sarcoma in brain and optic nerve. MS as AML extramedullary relapse had a poor prognosis.

### **Key Word:**

Acute Myeloid leukemia, Sarcoma, Brain, Relapse.

### **Abbreviations:**

AML: Acute Myeloid Leukemia

MS: Myeloid Sarcoma

CR: Complete remission.

CNS: central nervous system

### **Introduction:**

Myeloid sarcoma (MS), also known as chloroma or granulocytic sarcoma is a condition characterized by abnormal proliferation of blast cells, from one or more myeloid cell, and leads to a disruption in the structure of tissues (1). Its incidence ranges from 2.5% to 8%. It is commonly associated with subtypes of Acute Myeloid Leukemia (AML) such as M5a, M5b, M4 and M2 (2). Myeloid sarcoma can affects various body parts including lymph nodes, skin, soft tissues, testicles, bones, peritoneum and gastrointestinal tract(3,4). It can occur either on its own or as part of an AML relapse. The diagnosis is typically confirmed through immunohistochemistry analysis of a sample taken from the area. Unfortunately the prognosis of myeloid sarcoma is generally poor especially when it is associated with an AML relapse.(5)

### **Case report:**

A 19-year-old male was diagnosed with AML M2 with a normal karyotype in 2018, and underwent standard treatment with induction 7+3, re-induction 5+2, and consolidation therapy (Cytarabine 3 g/m<sup>2</sup>). And had achieved complete remission (CR) for three years. However, in 2022, the patient began experiencing neurological symptoms, including headaches, loss of vision, weakness in both lower limbs, and difficult to walk. A Brain MRI revealed a massive heterogeneous mass 9× 7.5×3cm in the left sphenoid wing region, This mass exerted pressure on critical structures such as the carotid artery, optic nerve, and cavernous sinus. Figure (1).

The patient underwent surgery using the frontotemporal orbit zygomatic approach (FTOZ) which successfully identified a tumor mass pressing against the carotid artery, median cerebrum, optic nerve, on the left side, optic chiasm and left cavernous sinus. The patient underwent neurosurgical surgery using the frontotemporal-orbit zygomatic approach (FTOZ), which detected a tumor mass compressing the carotid artery, median cerebrum, optic nerve on the left side, optic chiasm, and left cavernous sinus. The tumor mass was removed

until intact edges were reached with orbital scraping and optic nerve severing Figure (2). Pathological analysis of the mass confirmed the presence of a myeloid sarcoma involving both the brain and optic nerve, with orbital involvement. Immunohistochemistry further revealed positive staining for LCA, MPO, and CD34, while CD20 and CD3 tests returned negative results. Ki67 proliferative marker showed intermediate activity.

The patient received high-dose cytarabine (1.5 mg/m<sup>2</sup>) combined with intrathecal injections (cytarabine, dexamethasone, Methotrexate), and achieved clinical improvement and remission. Despite initial progress, the patient died four months after treatment began due to chemotherapy side effects and neutropenic fever.

## Discussion :

Myeloid Sarcoma (MS) is classified as a subtype of Acute Myeloid Leukemia (AML) and related neoplasms by the World Health Organization (WHO) (6). MS is similarly classified by the European Society for Hematology (ESH) into four separate categories: [1] MS in conjunction with AML, [2] extramedullary relapse of AML, including cases after bone marrow transplantation, [3] blast phase/transformation from myeloproliferative neoplasms or Chronic Myelomonocytic Leukemia (CMML), and [4] isolated MS, occurring without a history of myeloid dysplasia and with normal bone marrow aspirate findings.

The incidence of central nervous system (CNS) involvement in Myeloid Sarcoma (MS) is relatively rare,(7) accounting for only 0.4% of cases involving cranial bone marrow, vertebrae, or orbital bones. Its migration to the brain parenchyma is attributed to the disruption of the blood-brain barrier (8,9). While Orbital involvement as an initial manifestation of AML is uncommon, it is less than 3% of cases.(10) In our case, brain MRI revealed a heterogeneous mass measuring 9×7.5×3cm in the left temporal region, extending into the left sphenoid wing and causing destruction of the pilgrims. The mass compressed the left middle cerebral artery and bulged into the left cavernous sinus figure (1). MS can sometimes express B-cell antigens (CD19 and CD79a), which may lead to misdiagnosis as CNS lymphoma. However, in our case, immunostaining confirmed AML with myeloperoxidase (MPO) positivity.

Treatment approaches for MS lack consensus due to its rarity and limited randomized controlled trials (RCTs). Therapeutic decisions are influenced by factors such as tumor location, the timeline of MS occurrence (before AML onset or AML relapse), patient age, and performance status. Chemotherapy, surgery, radiotherapy, allogeneic hematopoietic stem cell transplantation (allo-SCT), targeted therapy, and immunotherapy are available therapeutic options [10]. Surgery plays a vital role in relieving mass effect symptoms, confirming diagnosis, and debulking large-sized MS before initiating systemic therapy [3]. In cases of isolated MS with inadequate response to chemotherapy or when rapid relief of vital function impairment is necessary, radiotherapy may be recommended [1,10]. In our case, we opted for neurosurgical surgery utilizing the frontotemporal-orbit zygomatic approach (FTOZ), followed by an induction chemotherapy protocol involving high-dose cytarabine (1.5 g/m<sup>2</sup>). Cytarabine has a good outcomes in MS.

The prognosis of Myeloid Sarcoma (MS) remains uncertain due to limited available data. However, it is generally acknowledged that MS occurring concomitantly with Acute Myeloid Leukemia (AML) or as a relapsed AML is associated with a poor prognosis (11). Patients who received chemotherapy showed better prognosis compared to those who did not (12). The life expectancy of individuals with MS varies based on several factors like age, performance status, and location of the disease, with a reported 5-years survival rate of approximately 24% (12,13). Disease relapse and infections are the most common causes of mortality in MS patients, in our case the patient dead with infection after 6 months from diagnosis MS.

In our case, the patient developed MS in multiple organs three years after achieving complete remission (CR) from AML, and unfortunately, his condition rapidly deteriorated within Six months of chemotherapy protocols. This highlights the challenges associated with MS and emphasizes the need for further research and advancements in treatment strategies to improve patient outcomes.

## Conclusion :

Myeloid sarcoma can occur in patients with AML who have been in complete remission, and can manifest in various organs. Awareness of MS in various organs in relapsed AML is essential, and this diagnosis demands

further individualized treatment due to the very high mortality risk.

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