Cured case of relapsed acute myeloid leukemia with giant central nervous system chloroma

**Abstract**

Myeloid leukemias are a heterogeneous group of disease characterized by infiltration of blood, bone marrow and other tissues by neoplastic cells of hematopoietic origin. Rarely, patients may present with symptoms from a mass lesion located in soft tissues. The mass lesion represents a tumor of leukemic cells and is called granulocytic sarcoma or chloroma. This is more commonly seen in monocytic subtype with abnormalities of chromosome 11. In general, they are felt to augur a poorer prognosis, with a poorer response to treatment and worse survival. There have been few case reports of orbital and central nervous system (CNS) chloroma in pediatric age group who have done better than only a medullary disease. Here, we present a case of acute myelogenous leukemia M2 who had two relapses one medullary and one extramedullary (CNS relapse with giant CNS chloroma). Patient was reinduced with same drugs and was advised to go for allogenic stem cell transplantation, which he could not. Subsequently presented with features of raised Intracranial tension (ICT) and was diagnosed to have extramedullary relapse in the form of giant CNS chloroma. He was treated with surgical debulking of the tumor, fludarabine, AraC, idarubicin, and G-CSF chemotherapy protocol (FLAG-IDA) chemo, and local intraspinal triple therapy 5 years back and stands cured at present.

**Key words:** Chloroma, cured, giant central nervous system, relapsed acute myelogenous leukemia

**INTRODUCTION**

Acute myelogenous leukemia (AML) is a clonal, malignant disease of hematopoietic tissues that is characterized by accumulation of abnormal (leukemic) blast cells, principally in the marrow, and impaired production of normal blood cells. The diagnosis of AML specifically is confirmed by identification of myeloperoxidase activity in blast cells or by identifying characteristic cluster of differentiation (CD) antigens on the blast cells (e.g. CD13, CD33). Patients with AML most often present with nonspecific symptoms that begin gradually or abruptly and are a consequence of anemia, leukocytosis, leucocytopenia or leucocyte dysfunction or thrombocytopenia. Myeloid sarcoma (also known as granulocytic sarcoma, chloroma, myeloblastoma, monocytoma) is a tumor composed of myeloblasts, monoblasts, or megakaryocytes. The tumors may occur as extramedullary masses without evidence of leukemia in blood or marrow, so-called nonleukemic myeloid sarcomas, or in association with AML. When the tumors appear as isolated lesions, they initially may be misdiagnosed as extranodal lymphoma because they look like lymphoid cells on biopsy. They may be found in virtually any location, including the skin; orbit; paranasal sinuses; bone; chest wall; breast; heart; gastrointestinal, respiratory, or genitourinary tract; central or peripheral nervous system; or lymph nodes and spleen. The tumors originally were called chloromas because of the green color imparted by the high concentration of the enzyme myeloperoxidase present in myelogenous leukemic cells. Biopsy specimens are positive for chloracetate esterase, lysozyme, myeloperoxidase, and CD markers of myeloid cells. When myeloid sarcomas are the initial manifestation of AML, the appearance of the disease in the blood and marrow may follow weeks or months later. Abnormalities in chromosome 8 are the most frequent cytogenetic disturbance in nonleukemic sarcomas. Systemic chemotherapy, rather than local therapy, should be used for treatment, although the long-term outcome in such cases usually is poor. Patients having AML with t(8;21) have a propensity to develop extramedullary leukemia and such patients with myeloid sarcomas have a poorer outcome after treatment. Johnston et al reported that Patients with myeloid sarcomas involving orbital and central nervous system (CNS) sites had significantly better survival than patients...
with non-CNS myeloid sarcomas with cerebrospinal fluid leukemia, or with no extramedullary leukemia. Treatment of meningeal leukemia can include high-dose intravenous cytarabine (that penetrates the blood–brain barrier), intrathecal methotrexate, intrathecal cytarabine, cranial radiation, or chemotherapy and radiation in combination. Systemic relapse commonly follows relapse in the meninges, and concurrent systemic treatment usually is indicated.

**CASE REPORT**

Our patient was a 16-year-old boy who had been treated for AML 7 years back with standard AML chemotherapy protocol. Patient on diagnosis presented with fever and petechial rash of 2 weeks duration. On presentation patient had anaemia (Hb 6.3 g/dl), white blood cell count was 33,600/cubic mm and platelet count of 18,000/cubic mm. Patient had mild biochemical coagulopathy with prothrombin time of 15s and activated partial thromboplastin time of 28s. Peripheral blood film examination showed 60% blasts with occasional auer rods. Bone marrow examination was consistent with AML M2 that was further confirmed on flow cytometry. Patient had a normal baseline cytogenetics. Patient received AML 3:7 induction chemotherapy and post induction marrow was in remission. Patient was consolidated with 4 high doses of cytarabine. Patient remained in remission for a period of 8 months when had first the medullary relapse and was reinduced with same drugs 2 years back. The diagnosis was reconfirmed on the basis of marrow and flow cytometry findings. Patient was advised to go for allogenic stem cell transplantation as consolidation that was not possible because of lack of human leukocyte antigen matched donor. Patient defaulted for a period of 3 months and presented 5 years back with 1-week history of headache, vomiting and got admitted in casualty with generalized tonic clonic seizures and went into coma. Emergency computed tomography (CT) head was done, which revealed left sided parietal lobe intracranial space occupying lesion [Figure 1]. Patient was immediately dilantinized, decongestant treatment was started and magnetic resonance imaging (MRI) brain was planned. Subsequently MRI brain also showed same findings [Figures 2 and 3]. After stabilization of the patient, an open brain biopsy was taken which proved this lesion as a CNS chloroma. Patient was treated with an intensive induction chemotherapy.
regimen that is, FLAG-IDA chemotherapy, received intrathecal triple chemotherapy (cytarabine, dexamethasone and methotrexate) along with local radiation therapy. Postchemotherapy CT head [Figure 4] showed resolution of chloroma and presently patient is in remission and is doing well but continues to be on antiepileptic medication.

CONCLUSION

Cure rates with chemotherapy alone in extramedullary disease are very low, and one should not go by responses shown by isolated cases as in our case. These patients should go for allogenic transplant where ever available and short of which these patients can be managed the way we managed our case.

REFERENCES


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