Case Report

Isolated Intracranial Myeloid Sarcoma Occurring as Relapse in Acute Myeloid Leukemia

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Abstract

Myeloid sarcoma (MS) or chloroma is a rare extramedullary tumor composed of extramedullary proliferation of blasts of granulocytic, monocytic, erythroid, or megakaryocytic lineage occurring at sites outside the bone marrow. MS occurs in 2%–8% of patients with acute myeloid leukemia (AML), sometimes it occurs as the presenting manifestation of relapse in a patient in remission. We describe the case of a young male with AML in remission for 6 years presenting with central nervous system symptoms. Magnetic resonance imaging showed an extra-axial altered intensity lesion in the parasagittal parietal region, infiltrating anterosuperiorly into anterior falx, and posterosuperior aspect of the superior sagittal sinus. A biopsy from the lesion was diagnostic of MS which was positive for myeloperoxidase. He did not have any other sites of disease. He has received chemotherapy with FLAG (Fludarabine, Cytosine arabinoside) followed by cranial irradiation and is in complete remission.

Keywords: *Intracranial,isolated, myeloid sarcoma*

Introduction

Myeloid sarcoma (MS) or chloroma is a rare extramedullary tumor composed of an extramedullary proliferation of blasts of granulocytic, monocytic, erythroid, or megakaryocytic lineage occurring at sites outside the bone marrow. MS occurs in 2%-8% of patients with acute myeloid leukemia (AML). it can precede the diagnosis of AML, or it can occur concurrently with diagnosis of AML or it may occur during the course of AML. Sometimes, it occurs as the presenting manifestation of relapse in a patient in remission.[1-3] We describe the case of a young male with AML in remission for 6 years relapsing with an intracranial mass.

Case Report

A 20-year-old male patient was diagnosed as AML M5a with hypodiploidy and treated with induction chemotherapy and three cycles of high-dose cytosine arabinoside. He was asymptomatic, in complete remission, and on follow-up for 6 years when presented with a headache for 5 months. The headache was insidious in onset, bifrontal and was associated with vomiting. He also had two episodes of seizures. He did not have any visual symptoms, facial asymmetry, nasal regurgitation, or gait disturbance. On examination, the patient was conscious, higher motor functions were normal, pupils were equal and reactive, and cranial nerves and fundi were normal. A magnetic resonance imaging showed an extra-axial altered intensity lesion measuring 42 mm × 37 mm in the parasagittal parietal region. The lesion was seen infiltrating anterosuperiorly along the interhemispheric dura into anterior falx and posterosuperior aspect of the superior sagittal sinus. The lesion was isointense on T1 and T2 showing diffusion restriction and enhances homogenously on contrast administration. Flair hyperintense was noted in the posterior parietal and superior frontal lobes along the lesion suggestive of edema. The picture was suggestive of leukemic infiltration causing pachymeningitis [Figure 1a-d].

The patient underwent the right pericoronal parasagittal craniotomy and biopsy. The histopathological examination showed tissue infiltrated by atypical medium-sized cells

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with moderate cytoplasm and indented irregular nuclei. On immunohistochemistry (IHC), the abnormal cells were positive for CD68, CD34, CD117, CD11c, and myeloperoxidase (MPO) [Figure 2a and b]. The cells were negative for CD20, CD5, and CD3. A diagnosis of intracranial MS was made. His blood counts, bone marrow, and cerebrospinal fluid studies were normal. He received salvage chemotherapy with fludarabine and cytosine arabinoside (FLAG) for 3 cycles along with triple intrathecal chemotherapy followed by cranial irradiation (24Gy). Currently he is in complete remission and on follow up.

Discussion

Apart from AML, MS can occur in chronic myeloid leukemia and other chronic myeloproliferative disorders. The common sites for MS are bone, soft tissue, lymph node, and skin. Rarely, they can involve the central nervous system (CNS), gastrointestinal tract, genitourinary tract, breast, and testes.[1-4]

MS is commonly seen in children and young adults and has a male predilection.[2] MS of CNS is a rare presentation of AML and may involve the subperiosteum, dura mater, and occasionally the brain parenchyma. Leukemic cells invade the CNS through the bone marrow of the adjacent cranium, vertebrae, or orbital bones.[5,6] Clinically, it may present as a meningeal disease (carcinomatous meningitis) or intravascular tumor aggregates in the brain (carcinomatous encephalitis) or as local tumor mass of MS.[7] The prevalence of MS in AML was 9.7%, but MS of the CNS was only 0.4% in a population-based study.[8]

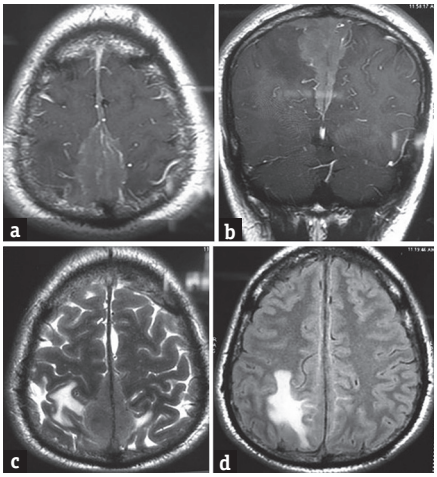

Figure 1: (a) Postcontrast T1-weighted magnetic resonance imaging brain showing homogenous enhancement. (b) Postcontrast T1-weighted magnetic resonance imaging brain coronal view. (c) T2-weighted magnetic resonance imaging showing isointense lesion with diffusion restriction and homogenous enhancement on contrast. (d) Magnetic resonance imaging showing flair hyperintensities suggestive of edema

In their review on the radiology of 24 intracranial MS, 48% presented as intraparenchymal tumor mass.[7] Radiologically, intracranial MS presents as an extra-axial hyperdense mass on noncontrast computed tomography scan and thus mimicks a meningioma, lymphoma, or metastasis.[7] Another pattern of the presentation was patchy leptomeningeal enhancement, mimicking leptomeningeal carcinomatosis, meningitis, neurosarcoidosis, dural sinus thrombosis, etc. Vasogenic edema was present in 45% of cases, and destructive bony changes are uncommon.[7] This patient presented with isolated MS of the brain as the first sign of relapse after 6 years of remission.

MS is classified into granulocytic, monoblastic, or myelomonocytic based on the predominant cell type. Microscopically, the characteristic pattern is Indian file pattern, and the MIB index is usually high.[2,4] IHC and immunophenotyping (IPT) are essential for a correct diagnosis. MS has IPT features identical to leukemic cells, the most common positive markers are CD68, MPO, CD117, CD99, CD34, Tdt, CD56, CD61, CD30, glycophorin, and CD4. MS is associated with chromosomal abnormalities such as AML and those with t(8:21) translocation have a predilection for MS of the CNS.[9] The present case was positive for MPO, CD68, CD117, and CD11c.

The currently recommended treatment for MS is combination chemotherapy similar to AML. There is no prognostic clinical or pathologic features, however, in patients who undergo allogeneic bone marrow transplantation, the survival is better.[2,10] The outcome of 23 patients with MS was compared with AML, and the event-free survival was found to be longer in patients with isolated MS.[1] Our patient received salvage chemotherapy with FLAG followed by cranial irradiation and is on follow up.

Conclusion

AML relapsing as an isolated intracranial MS is rare. A high index of suspicion for extramedullary disease

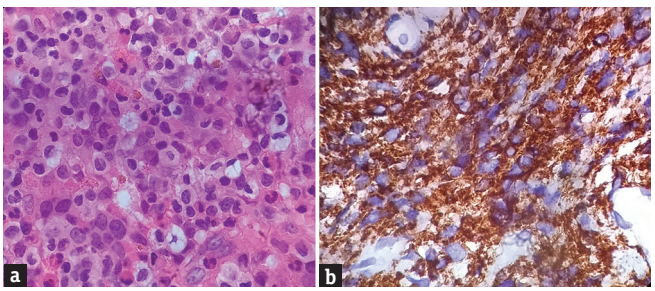

Figure 2: (a) Tissue infiltrated by abnormal cell (H and E, ×100). (b) Tumor cells positive for myeloperoxidase (IHC, ×100)

should be kept in patients with AML on follow-up who present with CNS symptoms. This is important for early diagnosis and salvage treatment.

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Conflicts of interest

There are no conflicts of interest.

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