Acute Myeloid Leukemia Presenting as Intracerebral Granulocytic Sarcoma

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Abstract
The CNS involvement of acute myeloid leukemia (AML) is more commonly manifest as meningeal involvement. Rarely it may present as intravascular tumor aggregates called granulocytic sarcoma which presents as intracranial hemorrhage. We are presenting a case of intracranial, intra-parenchymal granulocytic sarcoma (other names: chloroma, extramedullary myeloblastoma), presenting as acute hemiplegia without cerebral hemorrhage.

Introduction
More than one quarter of a million adults throughout the world are diagnosed annually with (AML).¹ Despite considerable progress during the past three decades in the therapy of AML, two-thirds of young adults and 90% of older adults still die of their disease.¹

Case Report
A 23 year postpartum female admitted with acute onset of right hemiplegia of one day duration. No history of undue bleeding manifestations.

On examination, she was conscious, aphasic, with minimal pitting pedal edema and oral thrush. Per abdomen, uterus was involuting and no organomegaly.

Neurological examination revealed, aphasia and right hemiparesis with power 2/5. There were no signs of meningeal irritation. Cranial nerves, sensory modalities, spine and cranium were normal. Fundoscopy showed papilledema.

Investigations showed, total leucocyte counts: 61,000 cells/ cumm; differential count: 50% Blasts; erythrocyte sedimentation rate: 13 mm/ hr; hemoglobin: 10.8 g/dl; hematocrit: 30%; platelets 1.24 lacs/cumm, serum creatinine 1.0 mg%.; sodium 135 meq/l, potassium 3.6 meq/l.

PT 20 sec, aPTT 34 sec, INR 1.5, serum LDH 300 IU/l, serum uric acid 4.0 mg/dl (the serum uric acid levels were unusually low since patient was not in tumor lysis syndromic phase), urine albumin trace, 10-12 pus cells/HPF, few RBCs. Peripheral smear showed >50% myeloblasts with occasional Auer rods, blast count of 58%, decreased platelets and marked micro and poikilocytosis.

Bone marrow revealed AML of M2 morphology (Figure 1). Blast count of 80% with few microblasts (micronormoblast: primitive erythrocyte). Cells were positive for myeloperoxidase.

Cytogenetic studies for AML could not be performed

NECT brain (non-contrast enhancement CT scan) showed diffuse cerebral edema. MRI brain showed multiple T1 hypo-intensities (Figure 2), T2 and FLAIR hyperintensities (Figure 3) noted in the left frontal hemisphere, right cerebellar hemisphere and corpus callosum with diffusion restriction (Figure 4),suggestive of acute infarcts³ in the left frontal lobe and right cerebellar hemisphere which was suggestive of granulocytic sarcoma and showed intense enhancement on gadolinium administration.

A diagnosis of AML-M2 with intracerebral intraparenchymal granulocytic sarcoma presenting as an acute cerebrovascular accident- right...
hemiplegia. Patient was treated with supportive measures, chemotherapy and anti-tumor lysis syndrome prophylaxis. She responded well to chemotherapy and other measures and after 7 days started conversing with others. However, during the course of the illness, she developed severe pancytopenia and despite the aggressive supportive care, patient died.

Discussion

Granulocytic sarcoma, also known as chloroma or extramedullary myeloblastoma, is a rare solid tumor composed of primitive precursors of the granulocytic series of white cells that include myeloblasts, promyelocytes and myelocytes. Rappaport named it granulocytic sarcoma in 1966. Granulocytic sarcomas have been observed in patients with acute myelogenous leukemia, chronic myelogenous leukemia, and other myeloproliferative disorders such as myelofibrosis with myeloid metaplasia, hypereosinophilic syndrome, or polycythemia vera. They occur in 2.5-9.1% of patients with acute myelogenous leukemia and five times less frequently in patients with chronic myelogenous leukemia.

Granulocytic sarcomas may develop during the course of, or as a presenting sign of, myelogenous leukemia. Less frequently (in up to 35% of patients), granulocytic sarcomas may precede the hematologic leukemia by months or years and can, therefore, be difficult to differentiate from lymphomas by clinical, radiologic, and even cytopathological methods. Special staining and histochemistry studies are required for accurate diagnosis.

Cerebral granulocytic sarcomas often appear as extra-axial masses. They are contiguous to meninges or ependyma and thought to arise from dural and subarachnoid veins and surrounding adventitia. Intracranial parenchymal masses, however, rarely have been reported. Multiplicity of intracranial lesions has been reported, as well as intraspinial and paraspinal involvement. Granulocytic sarcomas are isodense or hyperdense to brain or muscle on unenhanced CT, hypointense or isointense on T1-weighted MR images, heterogeneously isointense or hyperintense on T2-weighted MR images, and they typically enhance homogeneously after injection of contrast medium. They may be associated with edema and mass effect. In the setting of myeloid leukemia, these imaging features suggest granulocytic sarcoma, and, therefore, biopsy may be avoided.

Indeed, the signal intensity and homogeneous contrast-enhancement help in the differentiation of granulocytic sarcoma from hematoma and abscess.

References