Extramedullary leukemia is common in pediatric acute myeloid leukemia (AML) and occurs as a solid tumor (myeloid sarcoma). We report a case of a child who presented with acute onset of paraparesis and found to have intracranial and paravertebral mass; subsequently, he was diagnosed as having AML on tissue biopsy. He was started on AML treatment protocol, and later he was in remission and myeloid sarcoma got cleared from intracranial and paravertebral region. Timely diagnosis and initiation of treatment are essential to improve survival in such cases.

**Keywords:** Central nervous mass, extramedullary leukemia, myeloid sarcoma

**INTRODUCTION**

Acute myeloid leukemia (AML), also termed as acute myelomonocytic leukemia, acute myelogenous leukemia, and acute granulocytic leukemia, is the second most common type of leukemia in children, which accounts for 15%–20% of leukemia in children. Central nervous system (CNS) myeloid sarcoma (MS) is a rare manifestation of AML, first described by Burns[1] in 1811 by the term chloroma. MS is a solid tumor consisting of abnormal myeloblasts that occur outside the bone marrow. Other names for this entity include granulocytic sarcoma, chloroma (because of their greenish color), myelosarcoma, myeloblastoma, or extramedullary myeloid tumor.[2] Intracranially, MSs are often continuous with the meninges or the ependyma. Simultaneous involvement of CNS and spinal cord compression is rare as a first manifestation of MS. The definitive diagnosis of MSs is important in the treatment of AML. Here we report a pediatric case of paravertebral and CNS granulocytic sarcoma presenting as paraparesis before making a diagnosis of AML.

**CASE REPORT**

A 7-year-old male child presented to us with fever and pain over bilateral lower limbs for the last 2.5 months. He also had decreased movements of bilateral lower limbs for almost 1 week. There was also a history of swelling of bilateral feet for 1 day. There was associated loss of sensation of bilateral lower limbs and loss of sensation of bladder fullness/micturition/defecation. There was no history of seizure. Family and past history was unremarkable. On examination, he had nonpitting edema over the lower limbs. There was a history of urinary and fecal incontinence. He also had pallor and hepatomegaly (4 cm below costal margin) but no splenomegaly. His meningeal signs were positive. Deep tendon reflexes were absent with 0/5 power in both the lower limbs. Initial possibility of compressive myelopathy was kept. Differential diagnosis of metastatic neuroblastoma or lymphoma was kept.

**INVESTIGATIONS**

His hemoglobin was 7.4 g/dL, total leukocyte count (TLC) was 11,500/mm³, platelet count was 71,000/mm³, and lactate dehydrogenase was 3,441 U/L. Chest X-ray and echocardiography were normal. His computerized tomography (CT) scan of the brain revealed enhancing masses having smooth outline in (a) atrium of left lateral ventricle, 3.8 × 3 cm; (b) anterior falx cerebri, 2 × 1.7 cm; (c) dura based in the left anterior middle cranial fossa, and (d) the lateral extraconal space of right orbit, 15 × 8 mm
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He underwent CT-guided biopsy of the left paravertebral mass, and it revealed mononuclear cells that were immunopositive for myeloperoxidase (MPO) [Figure 2]. The features were suggestive of infiltration by myeloid leukemia. Bone marrow examination suggested AML. Child was started on dexamethasone initially as the child presented with compressive myelopathy. Subsequently, he was started on MRC10-AML protocol when definitive diagnosis of AML was made. At the end of the induction chemotherapy, the bone marrow aspirate of our patient was in morphological remission, and the CT scan of the brain was repeated, which showed >90% reduction and minimal residual left atrial lesion and resolved middle cranial fossa and orbit lesions [Figure 3A and B]. Unfortunately, after completion of fourth cycle of chemotherapy, his bone marrow was not in morphological remission and his parents did not opt for further therapy.

Discussion

Patients with AML develop MS by different patterns: (a) MS may develop during the active phase of leukemia; (b) they may manifest as a relapse after months or years after clinical remission of AML, particularly after bone marrow transplantation; and (c) they may precede the AML diagnosis and may be detected in patients who have a normal peripheral blood cell count. Our patient had anemia and thrombocytopenia but normal TLC. Krause described that 0.6% of cases do not have blast infiltration in the bone marrow. Most of patients take an average of 10.5 months to develop myeloid leukemic blast infiltration after the diagnosis of an MS.

MS seen in patients with leukemia may involve any part of the body. The most common sites for MS deposits are skin (leukemia cutis), lymph nodes, the bones, gastrointestinal tract, soft tissues, and gingivae. The spinal column is much less commonly involved, with an incidence rate of 13%–19%. The most common site is thoracic spine (64%) followed by lumbar, sacral, and cervical spine with an incidence rate of 29%, 20%, and 5%, respectively. Rarely, they may manifest as single or multiple intracranial lesions, most commonly seen within the calvarias and orbits. Cervantes and Cayci reviewed from literature that of 24 intracranial MS lesions, 13 lesions (48%) have presented as focal tumor masses within the intraparenchymal compartment of the brain.

MS appears to be more common in the pediatric population, with an incidence of up to 30% in children.
with AML compared with 2%–5% in adults. MSs are more common in infants than in older children and adults. MS can occur at any age often after the diagnosis of AML. MSs appear before the initial diagnosis of AML by months or years in approximately 25% of patients. There are conflicting results of prognosis of MS. The index patient had a mass in CNS as well as at paravertebral location and ultimately he relapsed.

Intracranial MS in patients with AML most commonly presents as an extra-axial hyperdense mass on noncontrast CT scan. Cervantes and Cayci reviewed that except one, from 24 intracranial MS cases, all appeared as hyperdense masses on the noncontrast CT studies. The differential diagnosis of hyperdense intracranial masses may include metastatic neuroblastoma, meningioma, B-cell lymphoma, intracranial metastasis, and Ewing’s sarcoma.

**Conclusions**

MS of the CNS is a rare presentation of AML. There should be suspicion of MS if any child presents with a CNS mass with or without normal complete blood counts. It should always be a part of the differential diagnosis in the evaluation of unusual masses. This can be detected before the diagnosis of leukemia or any time throughout the course of the disease during routine neuroimaging studies.

**Consent**

Informed written consent was taken from father.

**Guarantor**

Corresponding author.

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**Conflicts of interest**

There are no conflicts of interest.

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