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Leukemic Meningoencephalitis with Acute Quadriparesis: A Case Report of CNS Involvement in Chronic Myeloid Leukemia

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Abstract: ***Background**: Central nervous system (CNS) involvement in chronic myeloid leukemia (CML) is rare but represents a medical emergency, with presentation as leukemic meningoencephalitis. This case highlights the diagnostic and therapeutic challenges in managing CNS blast infiltration in a patient with CML. **Case Presentation**: A 26-year-old male farmer with a 5-year history of CML (on Nilotinib therapy) presented with sudden-onset acute quadriparesis, urinary retention, and autonomic dysfunction. Clinical examination revealed flaccid weakness (power 0–1/5 in lower limbs), areflexia, and elevated blood pressure with postural hypotension. Cerebrospinal fluid (CSF) analysis showed leukocytosis with hyperproteinemia; cytology revealed blast cells positive for BCR-ABL. The patient developed respiratory muscle weakness requiring mechanical ventilation. **Outcome**: Despite intravenous corticosteroid therapy and subsequent management, the patient developed critical illness with refractory autonomic dysfunction and respiratory failure. **Conclusion**: This case emphasizes the importance of recognizing CNS involvement in CML as a life-threatening complication and highlights the challenges of rapid diagnosis and management of leukemic meningoencephalitis.*

Keywords: Chronic Myeloid Leukemia, CNS involvement, Leukemic meningoencephalitis, Quadriparesis, BCR-ABL

1. Introduction

Chronic myeloid leukemia (CML) is a myeloproliferative disorder characterized by the translocation of chromosomes 9 and 22, resulting in the BCR-ABL fusion gene. With the advent of tyrosine kinase inhibitors (TKIs), the prognosis and survival of CML patients have improved significantly. However, central nervous system (CNS) involvement in CML remains a rare but serious complication, occurring in less than 1% of cases.

CNS involvement in CML typically manifests as leukemic meningoencephalitis, characterized by infiltration of blast cells into the meninges and brain parenchyma. This complication often presents with acute neurological deficits, including weakness, altered mental status, seizures, or coma. The diagnosis often requires rapid neuroimaging and cerebrospinal fluid analysis, followed by immediate therapeutic intervention.

This case report presents a challenging presentation of CNS leukemic involvement in a 26-year-old male with a history of CML, manifesting as acute quadriparesis and autonomic dysfunction.

2. Case Presentation

Patient Demographics

Parameter	Details
Name	Maridurai
Age/Sex	26 years / Male
Occupation	Farmer
Address	Sankarankoil, Tamil Nadu

Chief Complaints

Weakness of all four limbs for 1 day • Inability to pass urine for 1 day

History of Presenting Illness

The patient was in his usual state of health until one day prior to admission, when he experienced sudden-onset weakness of all four limbs while performing routine activities. The weakness was progressive, symmetric, and without diurnal variation. The patient initially required support to walk but progressed to complete bedridden status within 24 hours.

Motor Symptoms

The patient reported difficulty in holding slippers, with awareness of slippage. He had difficulty walking, sitting, and rising from a chair, with buckling of the knees. Additionally, he experienced difficulty in climbing stairs, rolling over in bed, raising his head from the pillow, raising arms above the shoulders, and performing fine motor tasks such as buttoning shirts or mixing food. No wasting, twitching, or stiffness was noted.

Autonomic Involvement

The patient reported inability to perceive bladder fullness, with difficulty in initiating micturition and urinary hesitancy. Catheterization yielded 1.3 liters of urine. Constipation for 2 days, abnormal sweating, and postural giddiness were also noted.

Sensory and Cranial Nerves

Sensory examination revealed intact perception of cloth, hot, and cold sensations. No numbness, paresthesias, or burning sensations were reported. Cranial nerve functions were intact, with no difficulty in smell, vision, chewing, hearing, swallowing, or head movements.

Past Medical History

The patient was diagnosed with Chronic Myeloid Leukemia (CML) 5 years prior to this presentation. He was initially treated with Imatinib for 2 years, followed by 2 years of irregular follow-up. Currently, he was receiving Nilotinib 400

mg twice daily for the past 1 year. The patient denied history of diabetes, hypertension, coronary artery disease, chronic kidney disease, tuberculosis, HIV infection, or bronchial asthma. No recent vaccinations were reported.

Physical Examination

General Examination
The patient was conscious and oriented, moderately built and nourished, afebrile, and not tachypneic. He was lying supine with all four limbs extended. General examination revealed no pallor, icterus, clubbing, generalized lymphadenopathy, neurocutaneous markers, or external markers of tuberculosis.

Vital Signs

Parameter	Value
Heart Rate	118/min, regular rhythm
Blood Pressure	200/110 mmHg (supine); 160/90 mmHg (standing)
Temperature	Normal, afebrile
Respiratory Rate	16/min, thoracoabdominal

Neurological Examination

Higher Mental Examination
Patient was conscious and oriented to time, place, and person. Attention and memory (immediate, recent, and remote) were normal. Speech comprehension, naming, and repetition were intact. Behavior was normal.

Motor Examination
Muscle tone was reduced (flaccid) in all four limbs. Muscle bulk was normal with no wasting or fasciculations. Muscle power assessment revealed:

Joint/Movement	Right	Left
Shoulder	1/5	1/5
Elbow	1/5	1/5
Hip	0/5	0/5
Knee	0/5	0/5
Ankle	0/5	0/5

Neck muscle power was 1/5. Deep tendon reflexes were absent bilaterally (biceps, triceps, supinator, knee jerk, and ankle jerk). Plantar reflexes showed no response. Abdominal reflexes were absent.

Sensory Examination
Light touch, pain, temperature, proprioception, and vibration sense were intact bilaterally in the upper and lower limbs.

Investigations and Findings

Hematological Investigations

Parameter	16/5	18/5	Normal
WBC ($\times 10^3/\mu L$)	18.4	16.3	4.5–11
Hemoglobin (g/dL)	13.5	13.8	13.5–17.5
Platelets ($\times 10^3/\mu L$)	74	102	150–400

Key findings: Leukocytosis with thrombocytopenia consistent with CML blast crisis.

Cerebrospinal Fluid Analysis

Parameter	Findings
Glucose (mg/dL)	45.7
Protein (g/dL)	95.4 (elevated)
Cytology	BLAST CELLS present; BCR-ABL positive
Culture & ADA	No growth; ADA negative

Other Investigations
- **Blood Cultures:** No growth
- **Serology:** VDRL, HBsAg, HCV, HIV all negative
- **Inflammatory Markers:** ESR 130 mm/hr; CRP 1.6 mg/dL; LDH 700 U/L
- **Renal Function:** Creatinine 0.95–1.0 mg/dL (normal)
- **Liver Function:** Transaminases elevated (SGPT peak 268 U/L); bilirubin 1.8 mg/dL; improving on serial testing
- **Cardiac Echocardiography:** Concentric left ventricular hypertrophy with preserved systolic function (EF 60%); no regional wall motion abnormality

Course of Illness in the Hospital
The patient presented with sudden-onset ascending-type flaccid paralysis. Initially, he could walk with support but rapidly progressed to complete bedridden status within 24 hours. The patient was treated with intravenous corticosteroids, and initial improvement in muscle power was noted. However, 2 days into hospital management, he developed a seizure episode accompanied by respiratory muscle weakness, necessitating endotracheal intubation and mechanical ventilation support. His clinical course was complicated by severe autonomic dysfunction, refractory hypertension, and progressive neurological deterioration.

3. Discussion

This case presents a challenging and rare manifestation of chronic myeloid leukemia: leukemic meningoencephalitis with acute quadriparesis and autonomic dysfunction. CNS involvement in CML occurs in less than 1% of patients and typically occurs in the blast crisis phase, often despite TKI therapy.

Pathophysiology
In this patient, the development of CNS involvement despite Nilotinib therapy suggests inadequate CNS penetration of the TKI or development of resistant clones. Leukemic blast cells infiltrate the meninges and spinal cord parenchyma, causing direct pressure on neural structures and triggering inflammatory responses. The presented clinical manifestations- acute quadriparesis, areflexia, and autonomic dysfunction- suggest spinal cord involvement at multiple levels (anterior horn cells and autonomic nuclei).

Clinical Presentation and Differential Diagnosis
The acute ascending paralysis with areflexia and autonomic involvement in this case mimics Guillain-Barré syndrome (GBS). However, several features distinguish this as leukemic meningoencephalitis: (1) CSF with significantly elevated protein (95.4 g/dL) combined with hypoglycorrhachia (CSF glucose 45.7 mg/dL), (2) positive blast cells and BCR-ABL in CSF cytology, (3) recent history of CML, and (4) elevated inflammatory markers (ESR 130 mm/hr). GBS would typically show albumin-cytological dissociation, and CSF glucose would be normal. Acute transverse myelitis was also

considered but less likely given the presence of CSF blast cells.

Management Challenges

The management of CNS leukemic involvement is complex and requires aggressive multimodal therapy. Intravenous methylprednisolone was initiated for immunosuppression and to reduce inflammation. However, the patient's response was limited, and rapid progression to respiratory muscle paralysis necessitated mechanical ventilation. Standard chemotherapy penetration to the CNS is limited, and intrathecal chemotherapy would have been a consideration, though the patient's clinical deterioration precluded this intervention.

Autonomic Dysfunction

The pronounced autonomic involvement (hypertension, postural hypotension, urinary retention, and constipation) indicates hypothalamic and brainstem involvement or spinal autonomic nuclei infiltration. This aspect of the presentation was particularly concerning and difficult to manage, as the patient exhibited simultaneous hypertension and orthostatic hypotension, suggesting dysautonomia from multi-level neural involvement.

4. Conclusion

This case highlights the critical importance of maintaining a high index of suspicion for CNS involvement in CML patients presenting with acute neurological deficits, even when on tyrosine kinase inhibitor therapy. The diagnosis of leukemic meningoencephalitis requires rapid neuroimaging and cerebrospinal fluid analysis. Early recognition and aggressive intervention, including high-dose corticosteroids and consideration of intrathecal chemotherapy, may improve outcomes. Clinicians should counsel CML patients on the potential for serious CNS complications and maintain regular monitoring for neurological symptoms during follow-up care.

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