

A Case of Plasma Cell Leukemia Presented with Encephalopathy

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Plasma cell leukemia (PCL) is a rare plasma cell disorder characterized by peripheral blood plasmacytosis. We present a patient with altered mentality diagnosed with PCL. A 57-year-old woman visited emergency room presenting altered mentality after two days of vomiting. She was drowsy and could speak only 3 to 4 words. Vital signs were stable, and about 3cm-sized nontender swelling on Rt. frontal skull was noted, while there was no history of trauma. Neurologic examination was nonspecific without any other motor or sensory signs. Laboratory findings were as follows: WBC 50,000/uL, Hb 8.40 g/dL, PLT 134,000/uL, Na/K 137/4.2 mmol/L, ionized Ca 2.35 mEq/L, BUN/Cr 26/1.56 mg/dL, LDH 1662.0 U/L, Total/Direct Bilirubin 3.03/1.58 mg/dL, ALP/AST/ALT 92/186/182 U/L, Total Protein/Albumin 5.9/4.0 g/dL, PT 1.88 (INR), B2 microglobulin 12.90 mg/L. Immunoglobulin levels were in normal. There was no heavy chain increment, but serum kappa light chain was elevated as 200 times as upper normal limit. PB smear showed leukoerythroblastosis with many immature blasts up to 30%. Since the symptom rapidly worsened to severe coma and AKI with anuria developed in a day, CRRT was applied. EEG revealed continuous generalized delta slow activity with triphasic morphology. Abdomen CT scan was normal except a chronic liver disease. Brain MRI revealed no abnormal signal intensity in parenchyme, but there was a 35mm-sized osteolytic enhancing mass through scalp and skull. Bone marrow study showed hypercellularity with markedly increased (68%) immature cells with high N/C ratio and bizarre morphology. Flow cytometry showed blasts were positive for CD38 (99.4%) and CD138 (98.9%). CD19, CD79a were positive in 47.1%, 50.3% respectively, but CD3, CD5 and CD20 were negative. Karyotyping found complex abnormalities including 45,X,-X and t(11;14) (q13;q32). Scalp mass biopsy was positive for CD138. Given with the PB blast count, PCL was diagnosed. The patient was recovered with CRRT then received Velcade, adriamycin, and dexamethasone combination chemotherapy. We demonstrate this case as a rare manifestation of plasma cell disorder. Also, the physician should consider paraproteinemic neuropathy as a cause of atypical neuropathy in such conditions.

Blastic plasmacytoid dendritic cell neoplasm: Diagnosis and Therapeutic approaches of Ten cases.

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Background: Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare hematologic malignancy that typically presents in the form of skin manifestations with or without lymph node and bone marrow involvement. Given its rarity and recent recognition as a distinct pathological entity, no standard of treatment exists for this aggressive disease and its prognosis is particularly dismal. **Methods:** We retrospectively analyzed clinical features and treatment outcomes of patients who were diagnosed with BPDCN between 2000 and 2014. **Results:** Ten patients had a median age at diagnosis of 41 years (range, 18-79), and 7 patients were male. Sites of disease involvement were the skin (n=7), lymph node (n=5), bone marrow (n=2), liver (n=2), spleen (n=2), and soft tissue (n=1). Intensified chemotherapy regimens such as hyperCVAD regimen (cyclophosphamide, vincristine, doxorubicin, dexamethasone, methotrexate, cytarabine), and VPDL (vincristine, methylprednisolone, daunorubicin, L-asparaginase) were used as a first-line treatment. Although all patients treated with intensified chemotherapy showed an objective response (5 patients with complete response) with median progression-free survival of 11.2 months (range 6.2-19.4 months), complete remission was not sustained for more than two years in any case. The response was relatively long-lived compared with previously reported CHOP-like regimens, but the above regimens do not result in long-term remission. **Conclusions:** All patients treated with hyperCVAD or VPDL showed an objective response, but the duration of response was relatively short. Thus, the development of more effective induction as well as consolidation treatment strategy should be warranted to improve this rare disease entity.