Isolated CNS Blastic Plasmacytoid Dendritic Cell Neoplasm in a Patient with AML

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Objective:
Blastic plasmacytoid dendritic cell neoplasm (BPDCN) is a rare hematologic malignancy derived from type 2 dendritic cells. While the marrow involvement may present similarly to acute myeloid leukemia, its immunophenotype, histologic features, and systemic manifestations render it a distinct entity. Most cases present as cutaneous lesions with or without concomitant bone marrow involvement, though nervous system involvement has been rarely reported. We present a case of isolated CNS manifestations of BPDCN in a patient diagnosed with of acute myeloid leukemia (AML).
A 73 y/o woman with history of myelodysplastic syndrome s/p lenalidomide developed therapy-related AML treated with multiple lines of chemotherapy for relapsing disease. Three months after completing her most recent therapy, she presented with falls, confusion, and lethargy. MRI Brain revealed multiple contrast-enhancing intraparenchymal lesions. Brain biopsy was initially deferred due to thrombocytopenia. CSF was positive for blast cells, and she was treated with systemic fludarabine and systemic/intrathecal cytarabine, clearing CSF blasts. Empiric antimicrobial therapy for fungal, bacterial, and parasitic etiologies was also initiated. Despite these interventions, serial imaging demonstrated increased number of intracerebral lesions. With improved thrombocytopenia, she was amenable to surgery where a firm, dark rubbery specimen was extracted and reported as relapsed acute leukemia with mixed blastic plasmacytoid dendritic cell (80-90%) and myeloid (10-20%) immunophenotype. She received palliative whole brain radiation and was dispositioned to hospice care.
This unique case illustrates CNS involvement in the absence of cutaneous BPDCN manifestations. This highlights the importance of considering BPDCN in the differential of intraparenchymal brain lesions in patients with acute myeloid leukemia, in addition to other infectious, neoplastic, vascular and inflammatory etiologies of brain lesions that can arise in this population. Biopsy was needed to confirm the diagnosis and provide appropriate treatment.
Background: N/A

Design/Methods: N/A

Results: N/A

Conclusions: N/A