Central Nervous System T-Cell Lymphoma Secondary to Mycosis Fungoides

Diagnostic and Management Challenges—A Case Report

AUTHORS

Mutasim Idriss[1], Mohamed Zeinab[2], Modather Grain[1], WaailRozi[1], Reed Robin[3], Saad Jamshed[3]

AFFILIATIONS

[1] Rochester Regional Health / Unity Hospital
[2] Sudan Medical Specialization Board
[3] Rochester Regional Health / Lipson Cancer Institute

INTRODUCTION

Mycosis fungoides (MF), the most common cutaneous T-cell lymphoma, rarely involves the central nervous system(CNS). (1) When it does, CNS infiltration signifies an aggressive course and poses major diagnostic and therapeutic challenges(2).

ROCHESTER REGIONAL HEALTH
Unity Hospital
LIPSON CANCER INSTITUTE

CASE PRESENTATION

A 75-year-old male with stage 1a MF affecting his face and neck, who had been treated with localized radiotherapy in 2021, presented in late 2024 with progressive symptoms, including altered mental status, ataxia, slurred speech, behavioral changes, and significant weight loss. Laboratory tests showed persistent peripheral eosinophilia (absolute eosinophil count 1,800/µL). MRI of the brain revealed T2 hyperintensities in the pons and the right middle cerebellar peduncle, as well as diffuse abnormal signals in the skull marrow. (see Figure 1) A lumbar puncture demonstrated eosinophilia and the presence of atypical lymphocytes in the cerebrospinal fluid (CSF). Flow cytometry identified a clonal T-cell population that was CD2+/CD4+ but lacked expression of CD3, CD5, CD7, CD8, and CD57. T-cell receptor gene rearrangement confirmed clonality. Extensive evaluations for infectious, autoimmune, and hematologic causes yielded no findings. Serologies for HIV and human T-lymphotropic virus (HTLV)-1/2 were negative.

Liver and bone marrow biopsies were negative for systemic lymphoma. Eosinophilia and mental status improved with high-dose corticosteroids, but systemic methotrexate was contraindicated due to transaminitis and upper-limb DVTs.An Ommaya reservoir enabled intrathecal methotrexate and cytarabine, clearing malignant CSF cells. Despite this, the patient developed aspiration pneumonia, COVID-19, atrial flutter, and imaging evidence of a left corona radiata infarct with pituitary enlargement. With progressive neurological decline and recurrent aspiration, he was transitioned to hospice care.

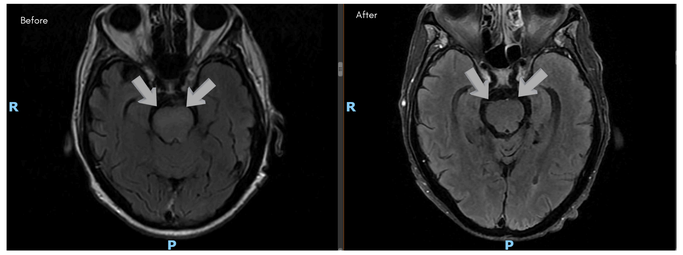


Figure 1: MRI of the brain shows hyperintense signal changes in the pons before treatment, with significant improvement noted after therapy.

DISCUSSION AND CONCLUSION

Our patient's early-stage (IA) MF with secondary CNS lymphoma within three years is highly unusual. Diagnosis is often delayed due to nonspecific neurological symptoms; Eosinophilic CSF may serve as an important early diagnostic clue. (3) Confirmation requires CSF immunophenotyping and T-cell receptor gene rearrangement. High-dose methotrexate is the primary treatment for CNS lymphoma, but its efficacy in CNS-MF is limited. When systemic therapy is not feasible, intrathecal chemotherapy via Ommaya reservoir can provide temporary CSF clearance without altering overall prognosis. CNS-MF remains highly aggressive, highlighting the need for early recognition and exploration of novel targeted therapies.

REFERENCES

1-Preston JD, Jansen CS, Kosaraju S, Niyogusaba T, Zhuang TZ, Iwamoto SW, Hutto SK, Lechowicz MJ, Allen PB. Cutaneous T-cell lymphoma with CNS involvement: a case series and review of the literature. CNS Oncol. 2023 Dec 1;12(4):CNS105. doi: 10.2217/cns-2023-0014. Epub 2023 Oct 25. PMID: 37877303; PMCID: PMC10701703.

2-Yang Y, Wickless H. Thinking about CNS metastasis in cutaneous lymphoma: Analysis of existing data. Leuk Res Rep. 2017 Oct 16;8:14-18. doi: 10.1016/j.lrr.2017.10.001. PMID: 29085772; PMCID: PMC5655392.

3-Jeffrey Schowinsky, Michelle Leppert, Douglas Ney, B.K. Kleinschmidt-DeMasters, Prominent Vascular and Perivascular Eosinophilic Infiltrates Heralding CNS Mycosis Fungoides, Journal of Neuropathology & Experimental Neurology, Volume 74, Issue 10, October 2015, Pages 948–951, https://doi.org/10.1097/NEN.0000000000000245