Word finding difficulty: a rare presentation of CNS involvement with MF-LCT

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BACKGROUND

- Mycosis Fungoides (MF) is the most common cutaneous T cell lymphoma.
- It is usually indolent but can involve the lymph nodes, blood, and visceral organs.
- 10-20% of patients with MF develop transformation to large cell histology (LCT).
- MF-LCT is predictive of a more aggressive clinical course and inferior prognosis.
- Secondary CNS involvement by MF is a very rare occurrence but tends to be associated with MF-LCT when present.

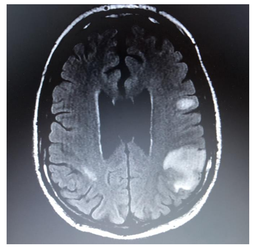
CASE PRESENTATION

- History:

A 58-year-old HIV-negative Caucasian male with extracutaneous MF-LCT presented with word finding difficult and right-sided weakness. The MF had been diagnosed three years prior with plaque-type MF which had since progressed to MF-LCT with Sezary cells by flow cytometry <250 cells/microL, B0 disease.

- PE:

Afebrile, hemodynamically stable.
Expressive aphasia, right-sided weakness.
Slit lamp and fundoscopic exam unremarkable.



- Labs:

CBC, CMP, LDH within normal limits.

- Imaging

MRI brain with an infiltrative pattern in left frontal lobe and left parietal lobe with associated leptomeningeal enhancement.

- Pathology

Stereotactic brain biopsy showed a diffuse infiltrate of highly atypical lymphoid cells with immunoperoxidase stains positive for CD3, CD7, CD43, and CD45 and negative for CD30 and TdT.

Bone marrow biopsy negative for involvement with lymphoma.

DIAGNOSIS AND TREATMENT

- The brain biopsy's phenotype was the same as that from a cervical lymph node biopsy when patient developed transformation to large cell histology 1 year prior.
- Patient was treated with high-dose methotrexate 3.5 g/m2 D1 and cytarabine 2 g/m2 BID D2, D3 for 21-day cycles x 4.
- Patient achieved an excellent response after 4 cycles of high-dose methotrexate and cytarabine
- Plan is to consolidate with thiotepa-based allogeneic HCT to offer best opportunity for durable remission.

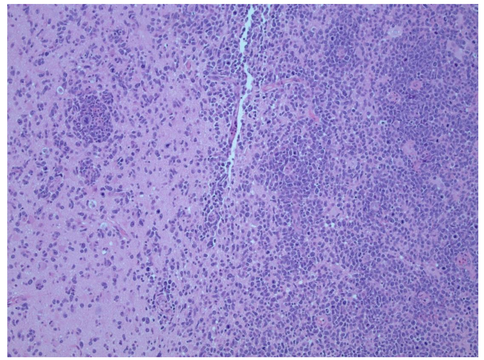


Figure 1: 10x magnification showing interface with background brain tissue.

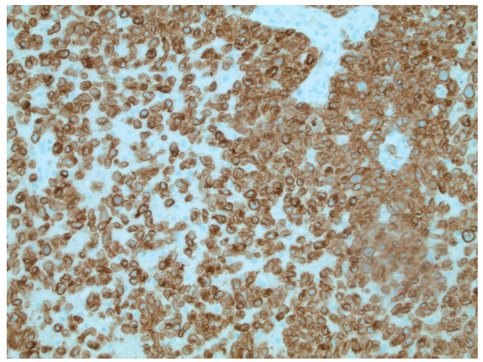


Figure 2:
CD3 IHC – atypical cells are CD3+

DISCUSSION

- MF is a rare malignant T-cell lymphoma, involving mainly the skin, but with potential involvement of the nodes, blood, and viscera.
- Skin lesions include patches or plaques, tumors, and erythroderma.
- MF-LCT has been reported in up to 10-20% of patients, most often arising from plaque-type or erythrodermic MF.
- Clinical findings of MF-LCT are variable but may include new solitary nodules within a long-standing patch or plaque, rapid development of multiple pink scattered nodules, or new or enlarging tumors.
- CNS involvement may be the only site of extracutaneous involvement in patients with MF-LCT.
- Focal neurologic deficits or mental status changes warrant CNS evaluation, particularly in those with known MF-LCT.
- Most cases of secondary CNS involvement from MF-LCT incorporate high-dose methotrexate and cytarabine followed by consolidative hematopoietic stem cell transplant in responding patients.

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