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Susac Syndrome as a Paraneoplastic Manifestation of Diffuse Large B-Cell Lymphoma: A Case Report

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Introduction

Paraneoplastic neurological syndromes are immune-mediated disorders that may precede the detection of an underlying malignancy. Susac's syndrome is a rare autoimmune microangiopathy affecting the brain, retina, and inner ear. We report a patient initially diagnosed with Susac syndrome whose initial malignancy work-up was negative, but who was later found to have diffuse large B-cell lymphoma (DLBCL) during rehabilitation follow-up, raising suspicion of a paraneoplastic process.

Case Report

A 52-year-old woman presented in April 2023 with right arm weakness (G4). Brain MRI revealed multifocal patchy T2 high signal intensities in bilateral white matter suggestive of CNS vasculitis (Fig 1.). Autoimmune and vasculitis studies were negative, and cerebrospinal fluid analysis showed mildly elevated protein without pleocytosis. Her symptoms improved after intravenous steroid.

Three weeks later, she developed psychomotor slowing and disequilibrium, which improved after intravenous immunoglobulin. In June 2023, she experienced photopsia and was diagnosed with bilateral retinal vasculitis (Fig 2.). One month later, she developed confusion and left arm weakness. Follow-up brain MRI demonstrated new multifocal lesions as well as cavitary chagne in corpus callosum (Fig 3.), and brain biopsy revealed perivascular lymphocytic infiltration with arterial wall damage without demyelination. Given the characteristic involvement of the brain and retina, she was diagnosed with Susac syndrome and initiated on intravenous cyclophosphamide for six months.

Despite immunosuppressive treatment, she continued to experience recurrent episodes, including sequential bilateral sensorineural hearing loss and left-sided weakness, leading to rituximab therapy. Spine MRI performed during this period showed focal T2 high signal intensities in the left anterolateral spinal cord at T4 level (Fig 4.) An extensive systemic evaluation, including tumor markers, paraneoplastic autoantibodies, and chest and abdominal CT scans, showed no evidence of malignancy at that time.

In May 2024, more than one year after her initial presentation, she was transferred to the Department of Rehabilitation Medicine for management with left-sided motor strength ranging from G2 to G4 and ambulating with a walker requiring assistance. After six weeks of inpatient rehabilitation, a hard anterior neck mass was incidentally detected. Neck CT (Fig 5.) and needle aspiration biopsy confirmed DLBCL. She completed six cycles of R-CHOP chemotherapy and achieved complete remission.

Since then, no further neurological deterioration has occurred. She continues outpatient rehabilitation; although, functional recovery remains limited.

Initial brain MRI

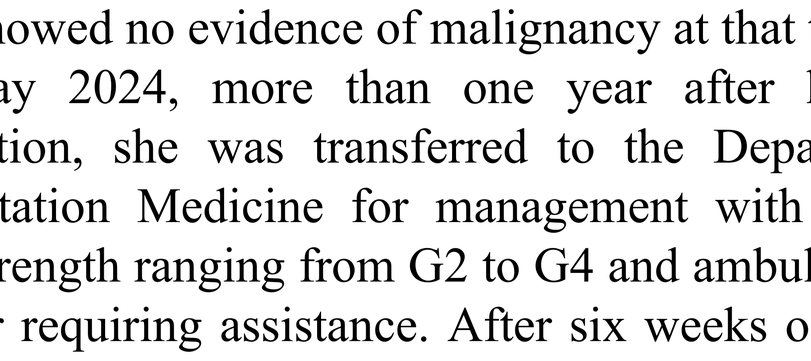

Figure 1. Axial T2-weighted image demonstrating multifocal hyperintense lesions involving the periventricular white matter.

Fundus photography

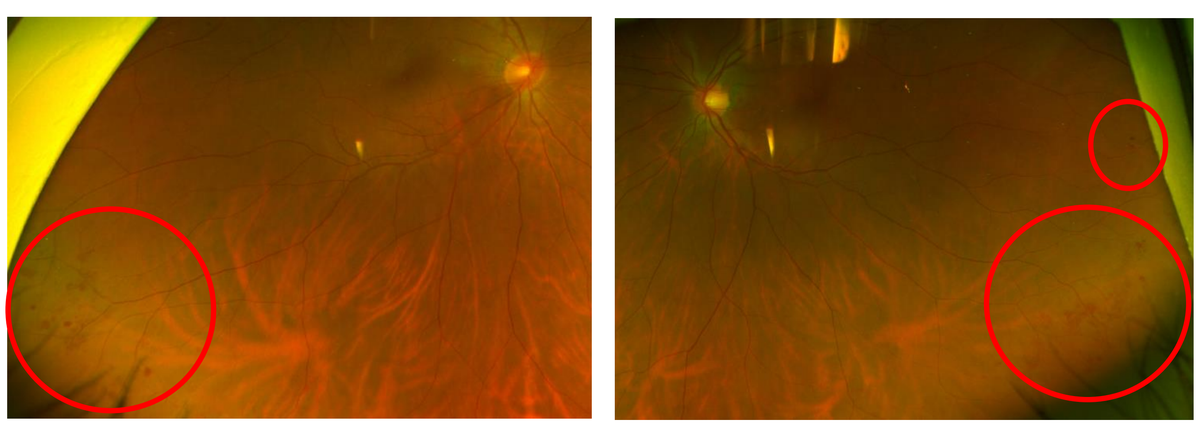

Figure 2. Image demonstrating focal hemorrhage in the peripheral retina, consistent with retinal vasculitis.

Follow-up brain MRI

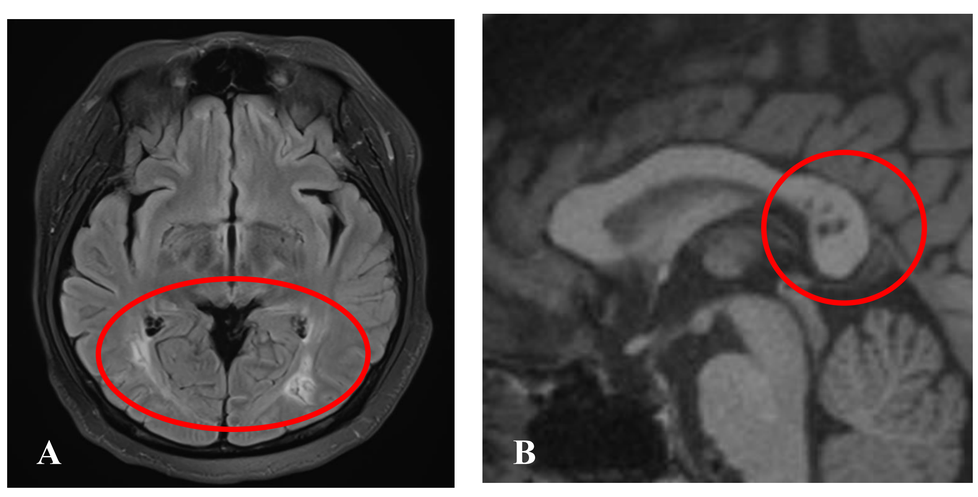

Figure 3. Brain MRI findings suggestive of Susac syndrome.
(A) Axial T2-weighted image demonstrating multifocal new hyperintense lesions. (B) Sagittal T1-weighted image demonstrating corpus callosum with cavitary change.

Spine MRI

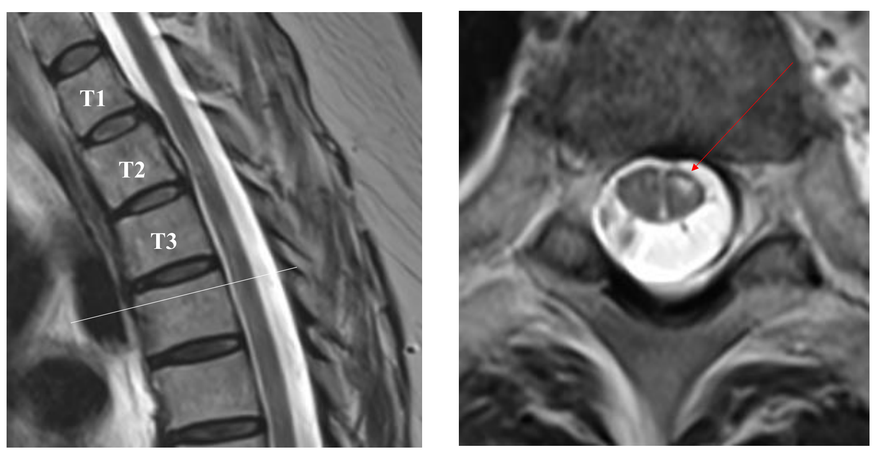

Figure 4. Sagittal and axial T2-weighted images demonstrating focal T2 hyperintensity in the left anterolateral spinal cord at T4 level.

Neck CT

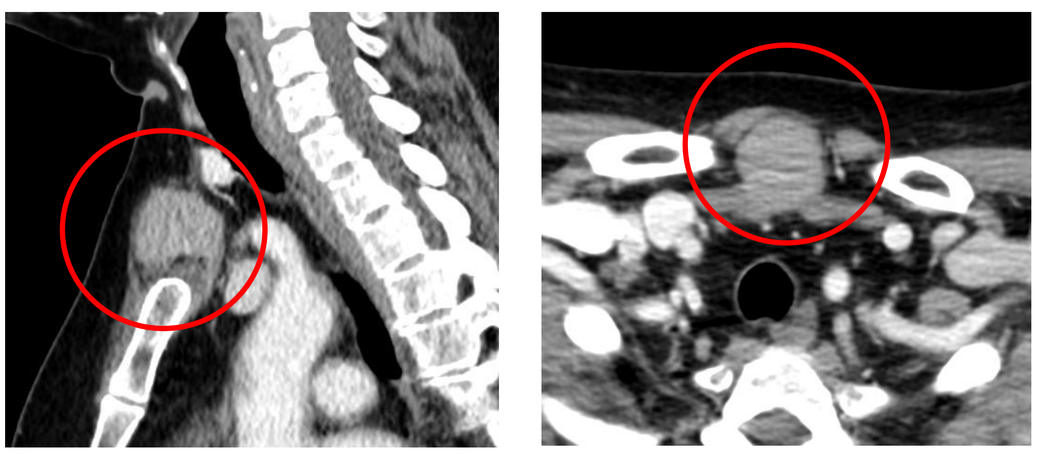

Figure 5. Sagittal and axial contrast-enhanced CT images demonstrating a well-defined enhancing mass at the suprasternal notch. Follow biopsy confirmed DLBCL.

Conclusions

To our knowledge, lymphoma-associated Susac syndrome has rarely been reported. Although the patient fulfilled clinical criteria for Susac syndrome and initial cancer screening was negative, DLBCL was later detected during rehabilitation follow-up. The temporal sequence and relapse pattern raise the possibility that the preceding neurological manifestations represented a paraneoplastic presentation of Susac syndrome.