

# Acute disseminated encephalomyelitis as a complication of Systemic lupus erythematosus: A rare case report

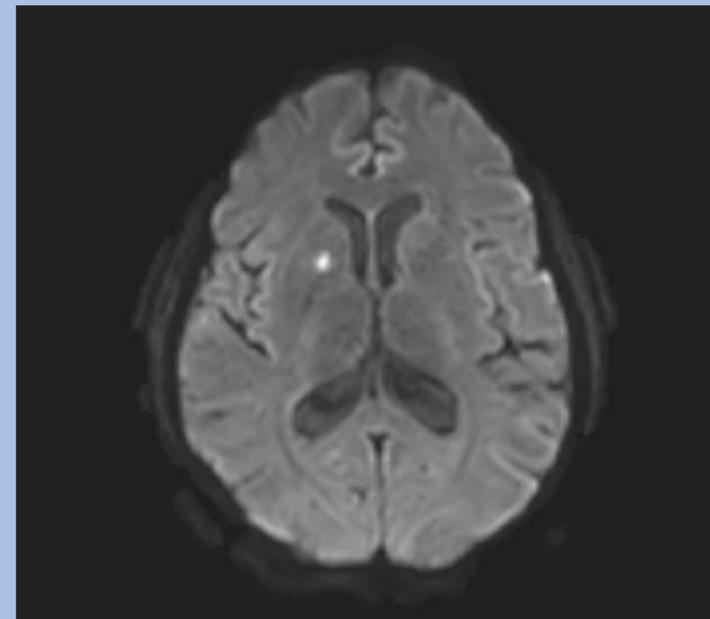
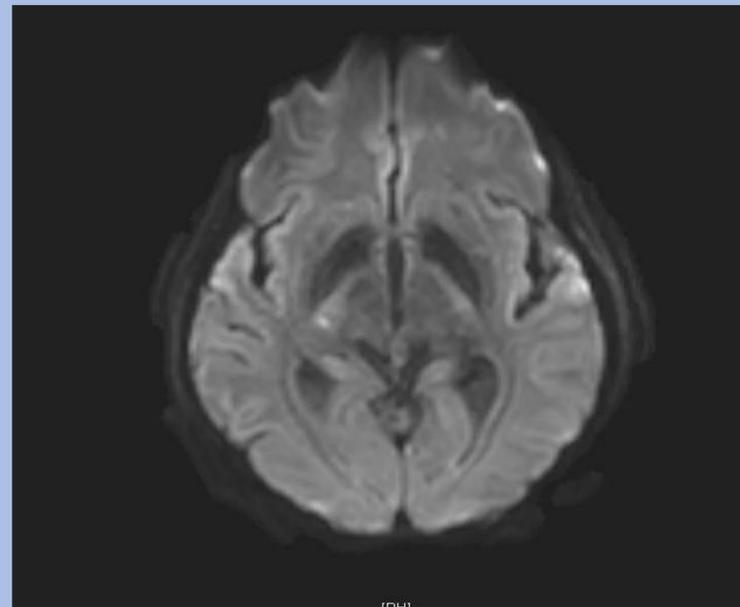
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## Learning Objectives

- ADEM can present as a complication of SLE and should be evaluated for when establishing the diagnosis.
- Early identification of disease, and timely initiation of treatment can result in good outcomes.

## Introduction:

Acute disseminated encephalomyelitis (ADEM) is a rare autoimmune disease, characterized by demyelination of the central nervous system (CNS). It may involve brain and spinal cord.[1] There is usually a preceding viral infection or antecedent immunization associated with the condition[2, 3]. SLE, also an autoimmune mediated disease, can present with diverse systemic manifestations, including involving the CNS. In this report, we describe the case of a patient presenting with acute neurological symptoms. Investigations that proceeded, lead to a diagnosis of concomitant ADEM and SLE, an association scarcely reported in medical literature.



## Case Description:

A healthy 41-year-old female presented with acute left lower extremity pain and weakness, with progressive cognitive decline over 2 weeks. She had been prescribed topical steroids for a rash on her scalp a few weeks ago. No fevers, insect, or tick bites were reported.

Vitals: Temperature 100.4 C, Blood pressure: 99/63 mm Hg, Pulse rate: 83 bpm. Oxygen saturations 93% on room air.

Physical examination findings : Weakness with left hip flexion power 4/5, loss of sensation on the plantar surface of her left foot, with preserved reflexes.

## Hospital Course:

Investigations during the hospitalization revealed mild normocytic anemia and mild transaminitis. Infectious workup, including urinalysis and chest x ray, were normal. A CT head was unremarkable. MRI cervical, thoracic, and lumbar spine findings were unrevealing. MRI brain done was suggestive of small foci of restricted diffusion and enhancement within the right basal ganglia, internal capsule, and pons. Symmetric restriction, T2 hyperintensity, and enhancement within the anterior bilateral temporal lobes and insulae were seen. Lumbar puncture was pursued with cerebrospinal fluid analysis significant for a WBC count of 22k and protein of 227mg/dL. CSF analysis showed 2 identical gamma restriction bands. Aquaporin antibody 4 Aquaporin-4 antibodies (NMO-IgG) were negative. CSF cultures, Cryptococcal antigen, Herpes Simplex Virus 1/2 PCR, and Varicella Zoster PCR testing were unremarkable. Syphilis screen and Lyme's testing done were also negative.

The development of a discoid rash on her forehead during the hospitalization prompted a rheumatologic workup. Serological studies were suggestive of SLE. Antinuclear antibodies titers were 1:1280, anti-double stranded DNA antibodies were 16 (<4 IU/ml). Hypocomplementemia was noted with complement 3 levels of 54 (81-157 mg/dL) and complement 4 levels of 6 (13-39 mg/dL).

She was treated with a 5-day course of pulse dose methylprednisolone, followed by steroids taper and azathioprine initiation. She followed up after discharge with resolution of the lesions on MRI, and complete neurological recovery.

## Discussion:

The initial presentation of subacute cognitive decline was concerning for central nervous system infection. Workup for infectious etiology was negative. The pattern of demyelination seen on MRI, central and peripheral nerve involvement, in addition to cerebral spinal serology, established a diagnosis of ADEM. The evolving rash and the preceding rheumatological workup led to diagnosis of SLE. The pathophysiology of both these entities is immune mediated. A review of prior literature revealed that this unique association has rarely been described[4,5]. ADEM can present as a complication of SLE and should warrant a high index of suspicion, as early identification and treatment can improve outcomes.

## References:

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