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RESEARCH ARTICLE

FULMINANT SUBACUTE SCLEROSING PANENCEPHALITIS PRESENTING AS REFRACTORY FACIO-BRACHIAL MYOCLONUS IN A YOUNG ADULT WITH RETROVIRAL DISEASE: A CASE REPORT

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ABSTRACT

Subacute sclerosing panencephalitis (SSPE) is a rare, progressive, and fatal neurodegenerative disorder caused by persistent measles virus infection. It typically affects children and adolescents, with adult-onset presentations being uncommon and often atypical. We report a case of a 19-year-old female with retroviral disease on antiretroviral therapy who presented with progressive facio-brachial myoclonic jerks and preserved consciousness. Initial neuroimaging and routine investigations were unremarkable, posing a diagnostic challenge. Cerebrospinal fluid (CSF) analysis revealed positive measles IgM antibodies, confirming SSPE. Despite aggressive supportive care and antiepileptic therapy, the patient deteriorated rapidly, developed septic shock, and succumbed. This case highlights an atypical, rapidly progressive presentation of SSPE in an immunocompromised host.

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INTRODUCTION

SSPE is a chronic encephalitis caused by persistent defective measles virus infection, usually manifesting years after primary infection. Classical features include cognitive decline, behavioral changes, and myoclonic jerks. Adult-onset SSPE is rare and often presents atypically, leading to diagnostic delays. Immunocompromised states, including retroviral disease, may alter disease course and progression.

Case Presentation

A 19-year-old female with known retroviral disease on antiretroviral therapy presented to the emergency department with:

- Chief complaints:
- Involuntary jerky movements involving face and upper limbs for 1 week
- Progressive increase in frequency
- Preserved consciousness throughout
- Initial assessment:Patient was hemodynamically stable but had frequent facio-brachial myoclonic jerks.
- Investigations:
- MRI brain (plain): Normal
- EEG: No definitive epileptiform abnormalities
- Routine labs: CBC, RFT, LFT, electrolytes – within normal limits
- Initial management:

Patient was started on multiple antiepileptic drugs:

- Levetiracetam
- Lacosamide
- Additional antiepileptic (as per ICU protocol) and othert supportive medications

Despite therapy, jerky movements persisted.

Further Evaluation

Given persistent unexplained myoclonus:

- CSF analysis:
- Routine: Normal
- Culture: Negative
- India ink, Gram stain, AFB: Negative
- CSF measles IgM: Positive

This confirmed the diagnosis of Subacute Sclerosing Panencephalitis (SSPE).

Clinical Course

- Patient was shifted to ICU for close monitoring
- Continued to have refractory myoclonic jerks
- Gradual neurological deterioration
- Required intubation and mechanical ventilation
- Developed sepsis with septic shock
- Despite aggressive management, patient succumbed after 5 days of intubation

DISCUSSION

This case demonstrates several important atypical features:

Adult-onset SSPE

SSPE commonly affects children; adult presentation is rare and often atypical.

Atypical Presentation

- Isolated facio-brachial myoclonus
- Preserved consciousness initially
- Normal MRI and EEG early in disease

These findings can mimic

- Autoimmune encephalitis
- Epileptic syndromes
- Movement disorders

Diagnostic Challenge

Routine imaging and EEG may be normal early. Diagnosis hinges on:

- CSF measles antibody detection
- Clinical suspicion

Role of Immunocompromised State

Retroviral disease may

- Accelerate disease progression
- Alter classical presentation
- Lead to fulminant course

Prognosis

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SSPE carries a poor prognosis, especially in:

- Rapidly progressive forms
- Adult-onset cases
- Immunocompromised patients

CONCLUSION

SSPE should be considered in young patients presenting with unexplained progressive myoclonus, even with normal MRI and EEG. Early CSF evaluation for measles antibodies is crucial. This case highlights a rare, fulminant adult-onset SSPE in a patient with retroviral disease, emphasizing the need for high clinical suspicion and early diagnosis

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