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Case Report

# Unprecedented Brain Imaging Findings in a Case of Acute Disseminated Encephalomyelitis

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## Abstract

Acute Disseminated Encephalomyelitis (ADEM) is an acute widespread autoimmune demyelinating disease affecting central nervous system. It is characterized by multifocal white matter lesions on neuroimaging. Grey matter can also be affected, particularly basal ganglia, thalami, and brainstem. Variant Creutzfeldt - Jakob disease (vCJD) is a rare and fatal human neurodegenerative condition affecting younger patients. Pulvinar sign on Magnetic Resonance Imaging (MRI) is considered to be a strong indicator of variant CJD. We report a young patient who presented with febrile illness and altered mental state. Brain imaging revealed hyperintensity in bilateral medial and posterior thalamus. These imaging abnormalities are similar to those seen in variant CJD.

## Introduction

Acute Disseminated Encephalomyelitis (ADEM) is an acute widespread autoimmune demyelinating disease, which principally affects brain and spinal cord that typically follows a febrile infection or a vaccination [1]. It is characterized by multifocal white matter lesions on neuroimaging. Demyelinating lesions of ADEM usually exhibit no mass effect and are scattered throughout the white matter of the cerebral hemispheres [2]. Grey matter can also be affected, particularly basal ganglia, thalami, and brainstem [3]. Variant Creutzfeldt - Jakob disease (vCJD) is a rare and fatal human neurodegenerative condition affecting younger patients (median age at death of 28 years) and has a relatively longer duration of illness (median of 14 months). Pulvinar sign on Magnetic Resonance Imaging (MRI) is considered to be a strong indicator of variant CJD [4]. Herein, we report a 28 year old patient who presented with febrile illness followed by altered mental state. MRI brain revealed bilaterally symmetrical, hyperintense signal changes predominantly in medial and posterior thalami along with caudate, putamen and insular cortex on T2-weighted and Fluid-Attenuated Inversion Recovery (FLAIR) sequences. These MRI abnormalities are commonly seen in vCJD.

## Case Report

A 28-year-old man presented with fever of 7 days duration followed by progressive decrease in mental state of 3 day's duration. There was no history of seizures, vomiting or loose stools. No cranial nerve symptoms, weakness of limbs, sensory symptoms or limb/ gait incoordination. He had retention of urine requiring catherization. No history of upper respiratory tract infection, dog bite, recent vaccination. On examination, he was stuporous, opening eyes briefly to painful stimulus. No paucity of limb movements. Pupils were equal but sluggishly reacting to light. Doll's eye response was intact. Tone was decreased in all four limbs. Deep tendon reflexes were sluggish and plantar response was mute. He was evaluated with Magnetic Resonance Imaging (MRI) of the brain and spine which showed bilaterally symmetrical hyperintense signal changes on T2-weighted (Figure 1) and FLAIR (Figure 2) sequences in medial and posterior thalami, caudate, putamen and insular cortex without any enhancement on contrast or white matter involvement or diffusion restriction. There was no imaging abnormality in the spinal cord. Cerebrospinal Fluid analysis (CSF) showed 10 cells per $mm^3$ predominantly lymphocytes, protein 86 mg/dl, sugar 64 mg/dl. Testing for tubercular bacilli on CSF by acid fast staining and Polymerase Chain Reaction (PCR) was negative. CSF Herpes Simplex Virus (HSV) Deoxyribonucleic Acid (DNA) PCR and Japanese Encephalitis (JE) Immunoglobulin (Ig) M and G were negative. Serological testing for Human Immunodeficiency Virus (HIV) and Hepatitis B Virus (HBsAg) was negative. Complete blood counts, renal, hepatic, thyroid function tests and ammonia were within normal limits. The chest X-ray and ultrasonography of the abdomen and pelvis were normal. Electroencephalography (EEG) showed diffuse slowing of background rhythm. A final diagnosis of ADEM was made based on the clinical and imaging findings and ruling out viral encephalitis like JE and HSV. He was given injection methylprednisolone 1g daily for 5 days and showed good response to that in terms of gradual improvement in sensorium. He was put on tapering course of oral prednisolone and was maintaining improvement at 2 week follow-up.

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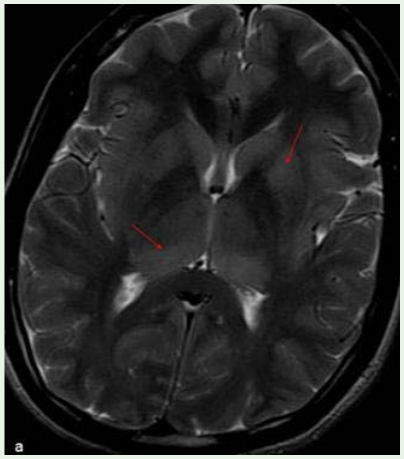

**Figure 1:** (A) Magnetic Resonance Imaging (MRI) T2-weighted imaging axial image of the brain showing symmetrical hyperintensity in bilateral medial and pulvinar nuclei of the thalamus, caudate, putamen and insular cortex.

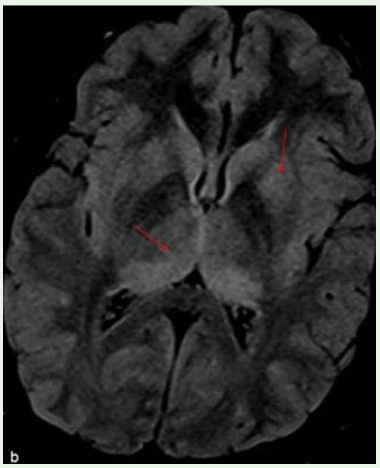

**Figure 2:** (b) Magnetic Resonance Imaging (MRI) Fluid-Attenuated Inversion Recovery (FLAIR) axial image of the brain showing symmetrical hyperintensity in bilateral medial and pulvinar nuclei of the thalamus, caudate, putamen and insular cortex.

## Discussion

Pulvinar nucleus is located at the caudal extremity of the thalamus. The presence of characteristic abnormalities on MR images in vCJD cases was first reported in 1997 and subsequent analysis of 36 cases and 57 controls suggested that, abnormal high signal intensity in the posterior thalamus (the pulvinar sign) and in dorsomedial thalamic nuclei (the "hockey-stick" sign) were sensitive and specific features of vCJD [5]. Pulvinar sign on MRI is considered to be a strong indicator of variant CJD. World Health Organization (WHO) has defined pulvinar sign as "a characteristic distribution of symmetrical hyperintensity (relative to the cortical and other deep grey matter nuclei signal intensity) of the pulvinar nucleus (posterior nucleus) of the thalamus seen on axial images" [6]. In addition to pulvinar sign, our patient had signal changes in caudate, putamen and insular cortex with sparing of white matter, brainstem. In view of bilateral thalamus involvement with CSF pleocytosis, Japanese encephalitis was considered but CSF JE serology was negative. Our patient had acute onset multifocal Central Nervous System (CNS) involvement with encephalopathy and MRI findings, he met the diagnostic criteria for ADEM as proposed by international pediatric multiple sclerosis study group [7]. In ADEM, lesions on MRI appear as patchy areas of increased signal intensity in the white matter on T2-weighted images and on FLAIR sequence. Usually, involvement of white matter predominates, but deep grey matter involvement in ADEM has also been reported. Pulvinar sign has rarely been reported as a radiological feature in ADEM [8]. Apart from vCJD and ADEM, it has been reported in Wernicke's encephalopathy [9], paraneoplastic limbic encephalitis [10] and benign intracranial hypertension. Hyperintensity in the basal ganglia is seen in a number of other conditions, the signal intensity of the pulvinar remains the most hyperintense as compared to basal ganglia, helping to categorize as probable vCJD. Abnormal EEG findings in ADEM vary greatly and ranges from mild to severe generalized slowing.

## Conclusion

The present patient had presented with clinical and imaging features suggestive of ADEM after ruling out viral encephalitis prevalent in this part of world. But the presence of increased hyperintensities of bilateral medial and posterior thalamus (pulvinar) as compared to caudate, putamen, sparing of white matter and EEG abnormality which are unusual in reported cases of ADEM. Variant CJD like imaging presentation of ADEM is rarely reported in literature.

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