

# Acute Disseminated Encephalomyelitis (ADEM): A Diagnosis of Exclusion with Atypical Neuroimaging

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## Abstract

Acute disseminated encephalomyelitis (ADEM) is characterized by a brief but widespread attack of inflammation in the brain and spinal cord that damages myelin-the protective covering of nerve fibers. Although most often observed as a single episode, relapsing or recurrent forms are also present. The true incidence in Pakistan is still undetermined. ADEM is a diagnosis of exclusion in many cases, and relies on neuroimaging. We present a case of young female having no history of immunization recently with nonspecific symptoms (lower limbs weakness, fever that progress to all four limbs weakness with urine incontinence and aphasia). An atypical MRI finding of extensive abnormal areas in white matter involving frontoparietal and occipital lobes on T2 and FLAIR. ESR, C.T brain and L.P came normal which subsided young stroke and multiple sclerosis hence diagnosis of ADEM was made. She was subsequently treated with high-dose steroids (methylprednisolone) and plasmapheresis with good outcomes.

**Keywords:** Acute disseminated encephalomyelitis; ADEM; Atypical MRI findings

## Introduction

Acute disseminated encephalomyelitis (ADEM) is an immune mediated central nervous system disorder [1]. It is characterized by an inflammation response and demyelination of the Central nervous system (CNS), which influences the white matter of brain and spinal cord [2]. ADEM can appear at any age but is more common in children with good prognosis [3] and rare in middle and elderly age group and due to non-reported cases in literature their outcomes are unknown [4]. ADEM seems more in winter and spring season, with male to female proportion of 3:1, in all parts of world in all ethnic gatherings [5]. ADEM is monophasic disease with beneficial long time prognosis in children [6]. The condition generally occur and accelerated by a viral disease mainly exanthemata's disease including measles, influenza, Epstein-Barr virus, rubella, hepatitis, varicella or inoculation e.g., rabies, small pox, polio or measles vaccines [7]. The hallmark of pathogenesis of post infectious encephalomyelitis is zones of perivenous demyelination and penetration of lymphocytes and macrophages. The current confirmation proposes that ADEM comes about because of a transient immune system reaction against myelin or different auto antigens, potentially, by means of molecular mimicry or by non-specific actuation of an auto reactive T cell clone. Peptides from microbial proteins that have adequate basic similitude with the host's self-peptides can enact auto reactive T cells; this mechanism is called molecular mimicry [8]. Initially symptoms of ADEM may start within 4-21 days after the prompting event. However in clinically hone if the impelling contamination is subclinical then it might end up plainly troublesome to separate the clinical disorder from intense viral encephalitis especially in nation like Pakistan where viral encephalitis is common [9]. The clinical signs and symptoms are identified with the place and seriousness of cerebrum injuries. Encephalopathy is the fundamental characteristic for infection that can quickly advance due to the multifocal neural lacks. That can quickly advance due to the multifocal neural lacks. Despite this, other neural reactions and signs may exist, for instance, Unilateral or two-sided pyramidal signs (60-95%), serious hemiplegia (76%), loss of visual power as a result of optic neuritis (7-23%), seizure (13-35%), spinal cord affliction (24%), cranial nerve loss of motion (22-45%), discourse impairment (5-21%), ataxia (18-65%), hemiparesthesia (2-3%) ultimately changes in levels of mindfulness from lethargy to unconsciousness [10]. The finding ADEM ought to be promptly considered at whatever point clinically there is a nearly connection between a contamination or an inoculation

and polysymptomatic beginning of neurological shortfalls owing to the CNS. Along with the diagnostic tool is brain magnetic resonance imaging (MRI) which detect widespread, multifocal or extensive white matter lesions (lesion load >50% of total white matter volume). The cerebrospinal liquid (CSF) may demonstrate a nonspecific lymphocytic pleocytosis and height of albumin levels. Oligoclonal banding might be available just briefly. In some cases analysis may not be clear neither the clinical introduction nor para clinical tests permit particular and unequivocal finding of ADEM. If all else fails, the analysis must be made by exclusion. We also present our case of young female in which neither clinical presentation nor CSF (Cerebral spinal fluid) analysis were consistent of ADEM also MRI findings were off atypical pattern [11].

## Case Report

22 years young female patient recently married had history of positive treated pulmonary tuberculosis 3 years back with no other co-morbidity admitted on 11-10-17 in surgical ICU ( Intensive Care Unit) with complain of lower limbs weakness, fever for 7 days, urine incontinence 3 days aphasia and altered behavior since morning. At the start of illness she initially developed bilateral lower limbs weakness which initially was not much extent but after 3 days it progress to extent that she used to walk with support and also started incontinence of urine sometimes. Though history of fever remained of low grade fever subjective perception non documented. Patient visited local G.P only and no workup was done. Then on 11-10-17 in morning she suddenly developed aphasia with all four limbs weakness and altered behavior (starring pattern, left sided gaze mostly). On arrival (day 1) her vitals were normal (B.P=125/71, Pulse=74b/min, R/R=20/min, O<sub>2</sub>=96%, Temp=A/F, RBS=137) (Table 1). GCS was 8/15(E4V1M3) both upper limbs power was 1/5, lower limbs 3/5, tone of both lower limbs were increased, reflexes of all four limbs

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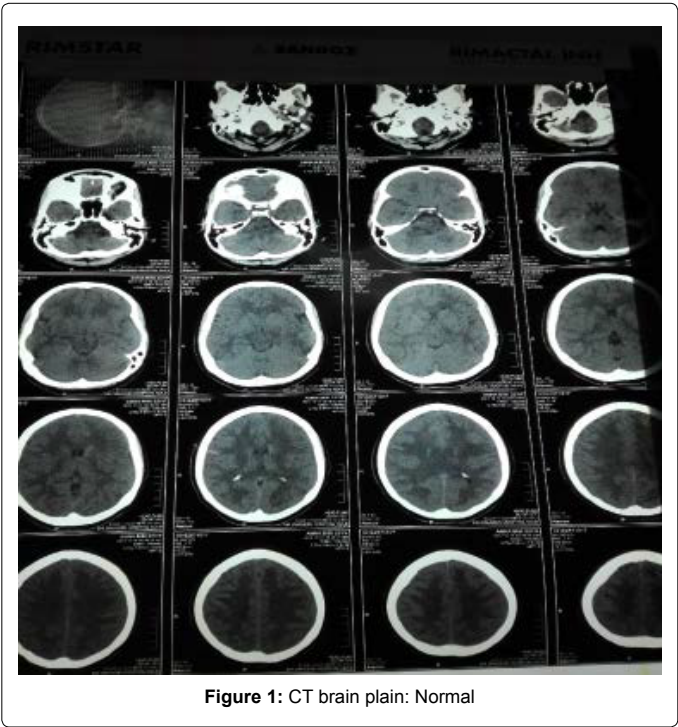
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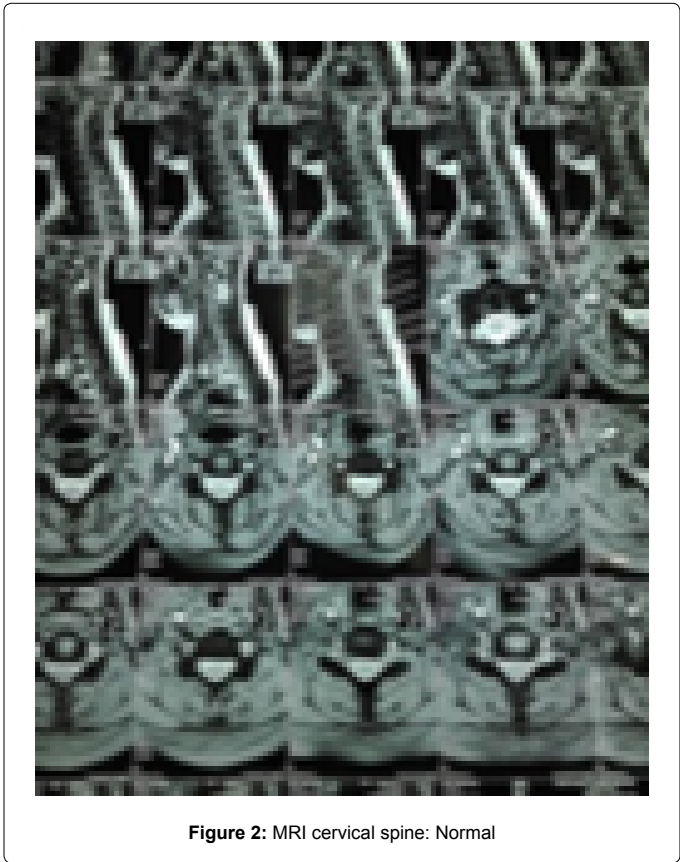
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Days	Vitals	GCS	Power	Therapy	Response
1 <sup>st</sup>	Remained stable	8/15	Upper limbs 1/5 lower limbs 3/5	Ceftriaxone, acyclovir started	No response
2 <sup>nd</sup>	Remained stable	8/15	All limbs 3/5	Ceftriaxone, acyclovir	No response
3 <sup>rd</sup>	Remained stable	8/15	All limbs 3/5	Ceftriaxone, acyclovir	No response
4 <sup>th</sup>	Remained stable	8/15	All limbs 3/5	Pulse therapy started along with ceftriaxone and acyclovir	No response
5 <sup>th</sup>	Remained stable	8/15	All limbs 3/5	Pulse therapy along with ceftriaxone and acyclovir	No response
6 <sup>th</sup>	Remained stable	8/15	All limbs 3/5	Pulse therapy along with ceftriaxone and acyclovir	No response
7 <sup>th</sup>	Remained stable	8/15	All limbs 3/5	Pulse therapy along with ceftriaxone and acyclovir	No response
8 <sup>th</sup>	Remained stable	11/15	All limbs 3/5	Pulse therapy along with ceftriaxone and acyclovir	Expressive response and sounds
9 <sup>th</sup>	Remained stable	11/15	All limbs 3/5	Oral dexamethasone ceftriaxone and acyclovir.	Expressive response and sounds

**Table 1:** Patient's health condition during treatment.



were exaggerated, plantar were bilaterally up going ,meningeal signs were negative for irritation of meninges, pupils were of 3mm reactive to light. Other systemic examination was not remarkable. Laboratory investigation revealed increased TLC count=13.9, Hb=11.9, PLT=389, UCE, urine D.R was normal, MPICT negative, ESR=11, ECG, Chest X-ray normal. Computerized Tomography (CT) Brain done was normal (Figure 1) and young stroke was ruled out. MRI cervical and dorsal spine was done and also found normal (Figure 2). In view of altered sensorium Cerebrospinal fluid (CSF) analysis was done which came normal (Glucose=69, protein=22, RBC=00, WBC=01, RBS =108). CSF DLC was not possible due to low TLC count so as no pleocytosis possible. No oligoclonal bands were present, so also M.S was ruled out. Keeping in view in early viral encephalitis chances of normal CSF D.R treatment on line of viral encephalitis Injection Acyclovir 750mg BD and Injection Ceftriaxone started. On 2<sup>nd</sup> day her GCS (Glasgow Coma Scale) remained same but tone and reflexes become normal with little power regain being 3/5 in all four limbs. On Day 3<sup>rd</sup> still no improvement seen in GCS. Neuroimaging MRI brain contrast was done. MRI showed extensive atypical abnormal areas within white matter involving frontoparietal and occipital lobes following almost asymmetrical manner bilaterally appearing hyperintense on T2 and FLAIR images and showing faint diffusion restriction (Figure 3). ADEM was considered and PULSE Therapy (injection methylprednisolone 1G

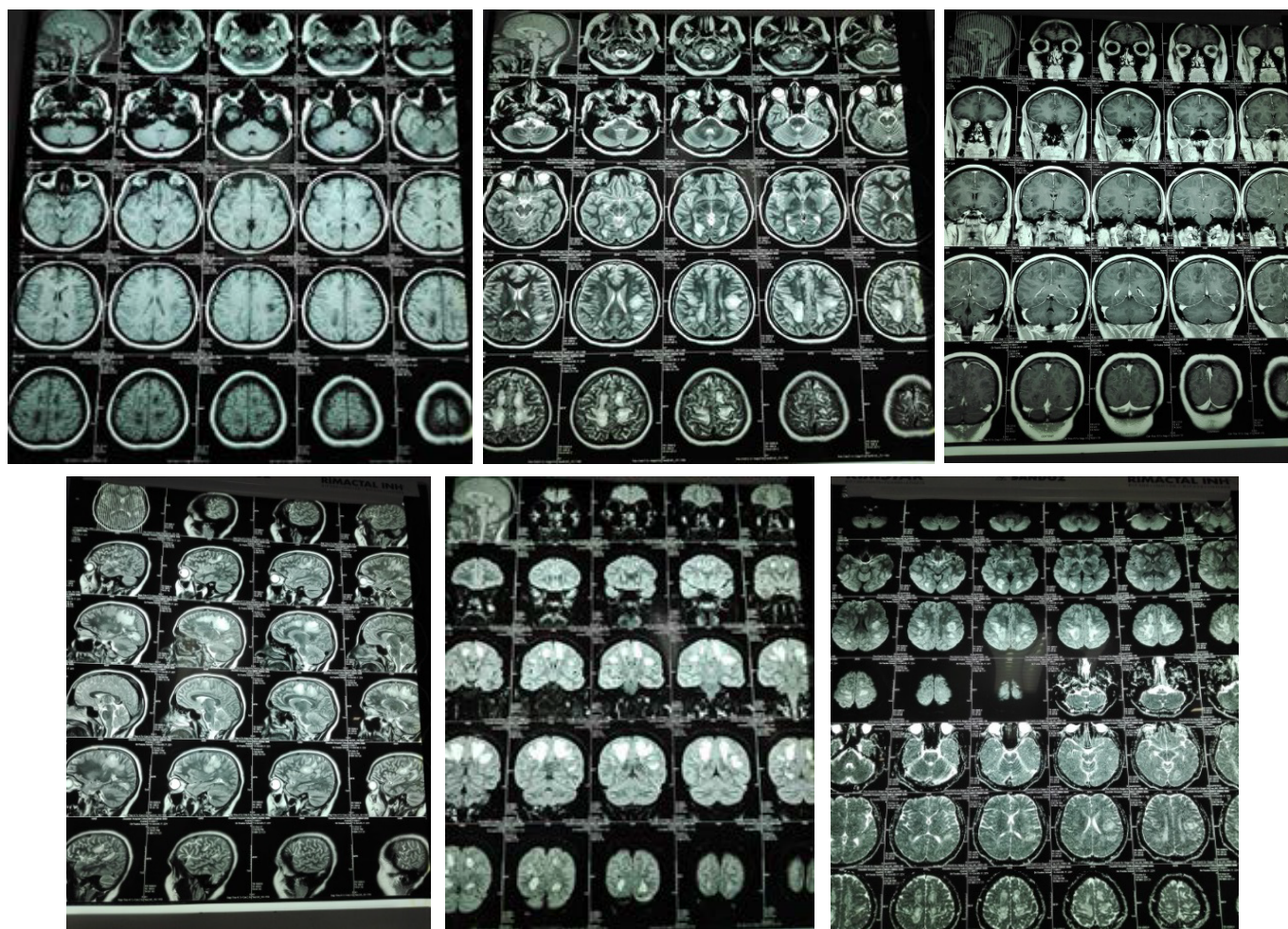


daily) started on day 4. Patient remained same on day 4<sup>th</sup>. On 5<sup>th</sup> day shifted to ward and very next day (day 6) she experienced 2 episodes of focal fits, was loaded with lerrace and then continued to maintenance 500mg twice daily. On 5<sup>th</sup> day of pulse therapy (day 8) patient started some expressive response and making meaning sounds to coherant. She was switched to oral dexamethasone on day 9<sup>th</sup> and shifted to Neurological setup for further plasmapheresis. She remained on follow up. Her five session on alternate days done and she showed marked improvement with regain of power 3/5 in all limbs and started giving response (Table 1).

**Discussion**

Many articles and case reports had been published on ADEM worldwide though it still remained a disease of dilemma. ADEM is conjectured to be an immunologically interceded demyelinating ailment activated by a febrile disease or later immunization, evoking a provocative reaction influencing the focal sensory system. Being more common in children and after post infectious or vaccination





**Figure 3:** Extensive atypical abnormal areas are seen within white matter involving the frontoparietal and occipital lobes following almost asymmetrical manner bilaterally appearing hyper intense on T2 and FLAIR images and showing faint diffusion restriction. These areas shows minimal enhancement on post contrast images predominantly in the parietal lobe. No signal dropout on SWI map. These imaging features are atypical.

and rare in adults. ADEM is diagnosis of exclusion criteria [12]. Introductory indications of ADEM may emerge 4-21 days after the actuating contamination [13]. An indicative problem may emerge in situations where the former disease is subclinical or when the inert period between the contamination and clinical manifestations is too short. In such cases, it is hard to clinically recognize ADEM and viral encephalomyelitis. We confronted a comparable issue in our case too [13]. MRI remained best modality to diagnose it. Radiologically, the T2/FLAIR (MRI) lesions of ADEM are diffuse, ill-defined, symmetric, often irregular, and occasionally patchy areas of homogeneous signal hyperintensities often involving both the gray and white matter of the brain with over half of cases involving infra-tentorial structures and greater than a third involving the spinal cord [7]. Involvement of the cerebellum and brainstem is more common in children. The blood-brain barrier disruption in ADEM is patchy and is mainly due to perivenular inflammation. Hence, the contrast enhancement pattern is variable, and may be described as nodular, diffuse, gyral, complete and incomplete ring. The precept strategy within management concerning ADEM includes controlling the immune response in opposition to nervous system with the aid of the use of immunosuppressant agents, as quickly as possible. High dose corticosteroids are at first line remedy

in ADEM. Salient features in our patient encompass a clinically and radiological and CSF D.R consequences indistinguishable condition from viral encephalitis additionally strange presentation of MRI findings as for ADEM findings and a superb reaction to intravenous methylprednisolone therapy and early plasmapheresis. This situation illustrates the importance of early diagnosis and immunosuppressive treatment in ADEM, as put off in treatment can cause irreversible neurological sequel.

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