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CASE REPORT: CLINICAL PRESENTATION, DIAGNOSIS AND MANAGEMENT OF ACUTE DISSEMINATED ENCEPHALOMYELITIS

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ABSTRACT:

Acute disseminated encephalomyelitis (ADEM) is a rare inflammatory condition of the central nervous system characterized by extensive demyelination and inflammation. Typically triggered by viral infections or vaccines, ADEM can manifest with various neurological symptoms including encephalopathy, motor impairments, and cranial nerve abnormalities. Diagnosis relies on clinical examination, neuroimaging findings, and the exclusion of similar conditions. Treatment often involves corticosteroids to reduce inflammation, and in some cases, immunosuppressive medications may be necessary. Prognosis varies widely, with many individuals experiencing partial or complete recovery, while others may continue to have neurological deficits or experience disease relapses.

Keywords: Demyelination, inflammation, ADEM, encephalopathy, Corticosteroids, immunosuppressive,

Introduction:

Acute disseminated encephalomyelitis (ADEM) is a rare inflammatory disease of the central nervous system (CNS) predominantly affecting the white matter of the brain and spinal cord. It often follows an infection, frequently triggered by a viral infection or vaccination [1]. ADEM is characterized by demyelination in these areas, occasionally involving the spinal cord, due to inflammatory responses following previous infection or immunization [2]. The onset of neurological symptoms closely following an infectious disease or vaccination strongly suggests the clinical diagnosis of ADEM [3].

ADEM can be classified into three types as described by Brain, Hunter, and Turnbull based on distinct pathological features: acute hemorrhagic encephalitis with multiple gross hemorrhages, acute cerebral cell degeneration accompanied by hyperemia and often edema but without gross hemorrhage, and perivascular demyelination [4].

Certain vaccines and pathogens have been associated with ADEM. While the specific causative pathogen is often unknown, common associations include cytomegalovirus, Epstein-Barr virus, herpes simplex virus, human herpesvirus-6, influenza virus, hepatitis A virus, human immunodeficiency virus (HIV), and Mycoplasma pneumoniae. Bacterial

infections such as Borrelia burgdorferi, beta-hemolytic streptococci, and Leptospira have also been linked to this condition[5].

The epidemiology of ADEM indicates an annual incidence ranging from 0.4 to 0.8 per 100,000 people, with children and young adults more frequently affected during winter and spring. Most cases are associated with vaccinations or post-exanthematous infections, and there is no apparent gender bias. The average age of presentation is around 7-8 years old, typically appearing between six days to six weeks after exposure to an antigen [6].

ADEM is often characterized initially by acute hemiparesis with bilateral pyramidal tract symptoms, followed by ataxia and altered consciousness, often accompanied by other neurological manifestations [7].

The pathophysiology of ADEM involves lymphocyte and macrophage infiltration, perivenous demyelination, hyperemia, endothelial enlargement, inflammatory cell invasion of artery walls, perivascular edema, and hemorrhage. These changes affect both small blood vessels in the white and grey matter [8]. Lesions primarily manifest as extensive, symmetric perivenular demyelination and inflammation in the Virchow-Robin spaces, affecting deeper cortical layers, thalami, hypothalamus, and other grey matter structures [9].

Diagnosis of ADEM relies on neurological examination, physical assessment, and diagnostic tests including blood tests, MRI scans revealing characteristic lesions, and analysis of cerebrospinal fluid which typically shows increased white blood cells [10].

Treatment for ADEM commonly involves corticosteroids as first-line therapy, with variations in steroid type, dosage, administration routes, and tapering schedules. In cases where corticosteroids are contraindicated, intravenous immunoglobulin (IVIG) serves as a viable alternative and has demonstrated efficacy in case series and reports [11].

Case Report:

A 22-year-old female patient presented to the emergency department with a chief complaint of weakness in both lower limbs over the past 4 days. She was asymptomatic until 4 days ago when she suddenly developed progressive weakness in both lower limbs. She experienced difficulty wearing slippers, getting up and sitting down, and walking. The patient denied any similar complaints in the past and had no other comorbid conditions.

Medications Prescribed:

- **Inj. Methylprednisolone 40mg/day**: A corticosteroid used to treat inflammation or immune reactions affecting various organ systems, endocrine conditions, and neoplastic diseases.
- **Inj. Piperacillin-Tazobactam 4.5gm/TID**: A combination of penicillin and beta-lactamase inhibitor used to treat bacterial infections.

- **Inj. Acyclovir 500mg/IV/TID**: A guanosine analog used to treat infections caused by herpes simplex virus, varicella zoster virus, and herpes zoster.
- **Inj. Thiamine 2 amp in 100ml DNS/BD**: A vitamin supplement used to correct vitamin B1 deficiency.
- **Inj. Sodium Valproate 500mg/IV/BD**: An anticonvulsant used to control complex partial seizures, simple and complex absence seizures.
- **Inj. Vitamin B12 1 amp in 100ml NS/IV/BD**: A vitamin supplement used to correct vitamin B12 deficiency.
- **IVIG 5 vials/day**: Intravenous immunoglobulin therapy used to treat immune deficiency states, autoimmune infectious diseases, and inflammatory conditions.
- **Tab. Chlorpheniramine 4mg/OD/HS**: A histamine-H1 receptor antagonist used for allergic reactions, hay fever, and rhinitis.
- **Inj. Ranitidine 1 amp/IV/BD**: A histamine H2 antagonist used to treat duodenal ulcers, Zollinger-Ellison syndrome, gastric ulcers, GERD (gastroesophageal reflux disease), and erosive esophagitis.
- **Inj. Ondansetron 4mg/IV/BD**: A serotonin 5-HT3 receptor antagonist used to prevent nausea and vomiting associated with cancer chemotherapy and postoperative recovery.

Discussion:

Many aspects of acute disseminated encephalomyelitis (ADEM) remain poorly understood. While considerable attention has been given to antecedent infections or vaccinations associated with ADEM, little is known about host factors such as genetics that may predispose individuals to develop this condition or influence its clinical course. ADEM can present with a multiphasic course, and there are currently no reliable biomarkers to predict the likelihood of subsequent development of multiple sclerosis (MS) [12].

Diagnosing ADEM relies on several clinical considerations, including initial symptoms, findings from both conventional and advanced MRI scans, long-term follow-up (up to 10 years), exclusion of alternative diagnoses, and, when available, brain histopathology. It remains unclear why the majority of ADEM cases are monophasic, with only a minority progressing to a multiphasic course[13].

Conclusion:

Acute disseminated encephalomyelitis (ADEM) is a rare yet serious condition marked by inflammation of the brain and spinal cord. While frequently triggered by viral infections or vaccinations, the exact cause remains elusive. Timely diagnosis and appropriate treatment are crucial for symptom management and to prevent complications. The long-term outlook varies, with some individuals achieving full recovery, while others may experience persistent neurological deficits. Continued research is essential for advancing our understanding and enhancing the management of this disease.

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