

Acute disseminated encephalomyelitis (ADEM)

By Professor Clive Hawkins, Professor of Clinical Neurology, Consultant Neurologist, University Hospital of North Staffordshire and reviewed by Dr Sarosh Irani, John Radcliffe Hospital, Oxford and Dr. Arun Venkatesan, Johns Hopkins Hospital, Baltimore, MD, USA

Introduction

Acute disseminated encephalomyelitis (ADEM) accounts for around 10% of all known cases of encephalitis. ADEM usually affects children and begins after a childhood rash (exanthema), other viral infections or immunisations. There is usually a latent period of days to two to three weeks before symptoms emerge. The illness has been poorly understood and a variety of terminologies are used to describe it, these including post-viral, post-infectious or para-infectious encephalitis.

The white matter of the brain (which contains nerve fibres and myelin) is predominantly affected in ADEM. Under the microscope it can be seen that there is invasion with immune cells from the blood. Where these cells accumulate, myelin (the protective fatty substance around nerves) is destroyed.

Symptoms

The symptoms can be similar despite different causes. The illness usually begins with less-specific symptoms such as fever, headache, stiff neck, vomiting and anorexia. These are rapidly followed by depression of consciousness in which the patient may become confused and occasionally comatose. Neurological features which may be detected include visual deterioration, clumsiness in arms and legs, paralysis down one side and seizures. The duration of these symptoms is variable. Some cases last a few weeks to a month, while other fatal cases have a rapid progressive course over several days. The symptom that correlates most closely with prognosis is the level of consciousness.

The illness usually has a monophasic course, i.e., once it is over, further attacks rarely develop. Recently long-term studies of patients with ADEM have shown that a small number later develop multiple sclerosis, although this area remains controversial.

Investigations

The cerebrospinal fluid (CSF) is frequently abnormal showing an increase in white cells and protein. The electroencephalogram (EEG) is abnormal in most cases showing diffuse slowing. Magnetic resonance imaging (MRI) typically shows multiple areas of abnormality in the white matter of the brain that can be rather characteristic.

Differential diagnosis

The differential diagnosis of ADEM includes acute meningitis, acute viral encephalitis, autoimmune encephalitis and acute multiple sclerosis. Differentiation of these diseases is not easy, certainly in the early stages. In viral encephalitis, the CSF is often abnormal and a rise in specific viral antibody may occur. To distinguish ADEM from multiple sclerosis in the initial phase may be more difficult. MRI and CSF examination may help.

Evidence for an immunological cause

There is a general agreement that a causative organism cannot be isolated from the brain of patients with ADEM. The association of the disease with a previous infection or immunisation suggests an immunological process.

Detailed laboratory studies involving measurement of anti-brain antibodies and of cellular immune responses to specific brain antigens suggest that these patients have developed an allergic immune response attack against their own brain constituents, and this is an 'autoimmune' response.

Treatment

The ideal form of treatment is immunomodulation (to dampen down the immune system) which should be started once the diagnosis is made as it has more benefit when given early. The diagnosis, however, may be difficult to make swiftly. High doses of steroids (drugs used to reduce inflammation) can often lead to a rapid resolution of symptoms with an excellent prognosis.

FS009V4 ADEM

Date created November 2001; Date updated: October 2021; Review date: October 2021

Disclaimer: We try to ensure that the information is easy to understand, accurate and up to date as possible. If you would like more information on the source material and references the author used to write this document, please contact the Encephalitis Society. None of the authors of the above document has declared any conflict of interest, which may arise from being named as an author of this document.

Support our information

With our support, no one has to face encephalitis alone. Our advice and information are available free of charge to everyone affected but we are truly grateful when supporters feel able to contribute a little to the cost of these resources. Please donate today by visiting www.encephalitis.info/donate or text the word DOCTOR to 70085 to donate £5.

Thank you!

The Encephalitis Society, 32 Castlegate, Malton, North Yorkshire, YO17 7DT, UK

Administration: +44 (0) 1653 692583 **Support:** +44 (0) 1653 699599

Email: mail@encephalitis.info **Website:** www.encephalitis.info

The Encephalitis Society is the operating name of the Encephalitis Support Group.

Registered Charity England and Wales No: 1087843; Registered Charity in Scotland No: SC048210

Charitable Company registered in England and Wales No: 04189027