



Acute Disseminated Encephalomyelitis (ADEM)

Dr Clive Hawkins, Consultant Neurologist, University Hospital of North Staffordshire, Stoke-on-Trent, UK

Acute disseminated encephalomyelitis (ADEM) accounts for up to one third of all known cases of encephalitis. This illness usually follows in the wake of exanthema or after other viral infections or immunisations. There is usually a latent period of days to two to three weeks. This illness was first described 250 years ago by the distinguished English physician, Clifton who noted that it occurred occasionally in patients who had smallpox. The white matter of the brain is predominantly affected and under the microscope it can be seen that there is invasion around small veins by white blood cells from the blood. Where these cells accumulate myelin is destroyed. The illness has been poorly understood and a variety of terminologies used to describe it, these including post-viral, post-infectious or para-infectious

Clinical Presentation

The clinical presentation of ADEM is similar despite different causes. The illness usually begins with non specific symptoms such as fever, headache, stiff neck, vomiting and anorexia. These are rapidly followed by depression of consciousness when the patient may become confused, delirious and occasionally in coma. During this early period neurological examination usually shows focal neurological signs such as bilateral optic neuritis, ataxia of the limbs, clumsiness in walking, paralysis down one side and seizures may occur. The duration of these symptoms is variable, some cases lasting a few weeks to a month, and other fatal cases having a rapid progressive course over a number of days. The clinical sign 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.

Investigations

The cerebrospinal fluid is frequently abnormal showing an increase in white cells and protein. The electroencephalogram is abnormal in most cases showing diffuse slowing. Magnetic resonance imaging typically shows multiple, symmetrical areas of abnormality in the white matter of the brain..

Differential Diagnosis

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

Pathology

The brain at post mortem may appear entirely normal or may show the signs of congestion. Histologically the lesions consist of infiltration of mononuclear cells from the blood around small veins in the white matter. Demyelination occurs and is limited to the area of the perivenous cellular cuff. This is different from lesions found in multiple sclerosis.

Evidence for an Immunological aetiology

There is general agreement that a causative organism cannot be isolated from the

supporting people in the UK, the Republic of Ireland and worldwide

central nervous system of patients with ADEM. The association of the disease with an antecedent infection or immunisation suggests an immunological process and detailed laboratory studies involving measurement of anti-brain antibodies and of cellular immune responses to specific myelin antigens have shown that these patients indeed have mounted an allergic response against their own brain constituents.

Treatment

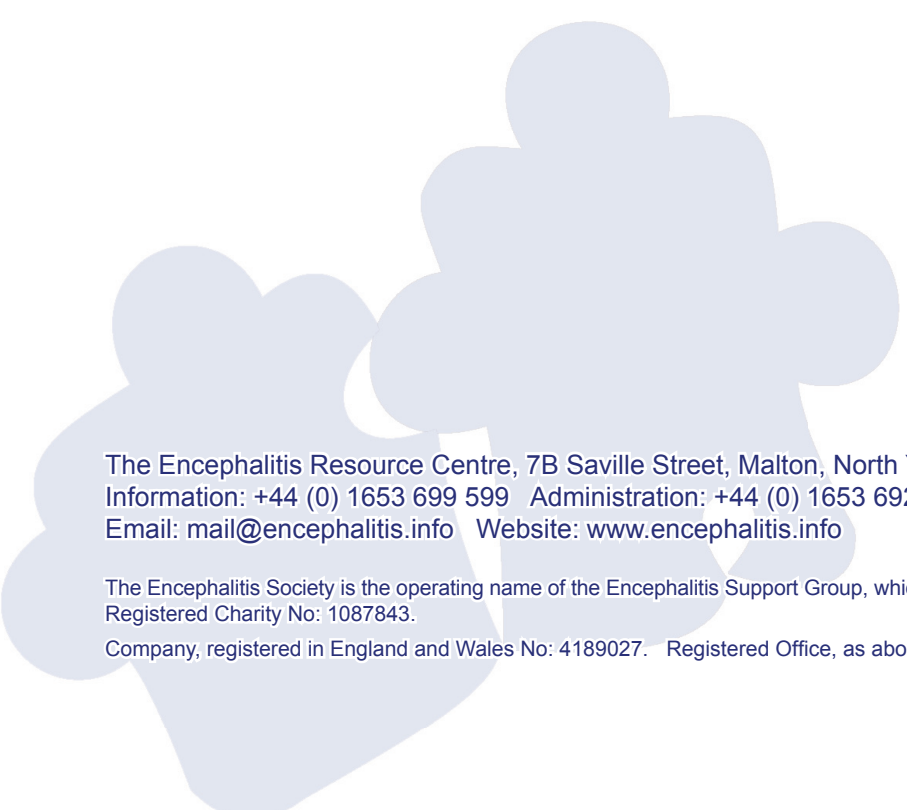
The ideal form of treatment is immune-modulation to be instituted once the diagnosis is made. High doses of steroids can often lead to a rapid resolution of symptoms with an excellent prognosis. Overall the prognosis is good where the diagnosis is made early and appropriate therapy instituted without undue delay.

© Copyright The Encephalitis Society All rights reserved.

FS 9 ADEM Created 11/2001

Last Update December 2008

The views expressed in any quoted resources represent those of the authors and are not the views or official policy of the Encephalitis Society and its Professional Panel.



The Encephalitis Resource Centre, 7B Saville Street, Malton, North Yorkshire YO17 7LL UK
Information: +44 (0) 1653 699 599 Administration: +44 (0) 1653 692 583 Fax: +44 (0) 1653 604 369
Email: mail@encephalitis.info Website: www.encephalitis.info

The Encephalitis Society is the operating name of the Encephalitis Support Group, which is a Charitable Company Limited by Guarantee. Registered Charity No: 1087843.

Company, registered in England and Wales No: 4189027. Registered Office, as above.