Acute Disseminated Encephalomyelitis (ADEM)

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This factsheet aims to provide people affected by Encephalitis, families, friends, carers and health care, social and educational professionals with a better understanding of Acute Disseminated Encephalomyelitis

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 (childhood rash) or after other viral infections or immunisations. There is usually a latent period of days to two to three weeks before symptoms emerge. 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 with white blood cells from the blood. Where these cells accumulate myelin (the protective fatty substance around nerves) 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 in which 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 (visual deterioration), clumsiness in arms and legs, 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 on develop multiple sclerosis, although this area remains controversial.

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, areas of abnormality in the white matter of the brain: these can be rather characteristic.

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.
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. Magnetic resonance imaging and CSF examination may help.

**Pathology**

The brain tissue may appear entirely normal or may show the signs of congestion. Histologically the ADEM lesions consist of infiltration of white blood cells, from the blood, which occur around small veins in the white matter. Demyelination (myeline loss) occurs. These are different from the lesions found in multiple sclerosis.

**Evidence for an Immunological aetiology**

There is general agreement that a causative organism cannot be isolated from the 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 brain antigens have shown that these patients indeed have mounted an allergic response against their own brain constituents.

**Treatment**

The ideal form of treatment is immunemodulation which should be instituted once the diagnosis is made. The diagnosis, however, may be difficult to make swiftly. 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 delay.
We try to ensure that the information is accurate and up-to-date as possible. 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.

The authors have used evidence, academic and professional experience in writing this factsheet. If you would like more information on the source material and references the author used to write this page please contact the Encephalitis Society.