

Encephalitis lethargica

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What is encephalitis lethargica?

Encephalitis lethargica (EL) is a serious and rare form of encephalitis, which can present with variable and serious symptoms. It is typically seen during discrete epidemics, but sporadic cases still occur.

History

Since the late 16th century, epidemic outbreaks of disease that appear to have been like EL have been reported in various European countries. The name encephalitis lethargica itself was given by the neurologist Constantin Von Economo in 1916 at an early stage of the epidemic that happened during and after the First World War.

Difficulty of diagnosis and the lack of statistical records in many countries mean that estimates of the number of cases have varied greatly. Most cases are at the younger end of the age spectrum (up to 30/40 years old). There was little confirmed data around race and ethnicity although some reports suggested mortality was lower among black people.

For many years EL was regarded as a phenomenon of the past. In recent years there has been a recurrence of interest in the disease, concerning its cause and links with post-encephalitic Parkinsonism. The neurologist Oliver Sacks described the 'awakenings' of past cases, but recent papers have described contemporary cases and defined the disease in modern terms.

Modern reports suggest that perhaps half a million to a million or more people were affected, of whom about one-third died. Patients who survived EL often developed a form of parkinsonism called post-encephalitic parkinsonism, which results in serious neurological disability.

Causes

So far, the cause is unknown. Several opinions argued that it started from the 1918 influenza epidemic as either an acute viral, or a post-viral syndrome. More recent work linking EL to a streptococcal infection may prove more compelling than the influenza theory. Although the last epidemic of EL occurred nearly a century ago, the cause was never scientifically established, and remains a matter of controversy today. This makes diagnosis difficult and is one of the reasons why we do not know how many cases of EL there have been in recent times. We still see sporadic cases of EL today. However, some may be forms of autoimmune encephalitis such as NMDA-receptor encephalitis and therefore all suspected cases of EL should be tested.

Symptoms and diagnosis

The term 'sleepy sickness', where people seem to fall asleep or freeze whilst eating or working, was first used to describe two cases in Vienna. However, the disease can present a wide and sometimes confusing range of symptoms, often with unusual and bizarre behaviour. There are indications that most cases were referred to psychiatrists before being admitted to hospital if the symptoms progressed. It is often mistaken for epilepsy, hysteria, intoxication, or a reaction to drugs.

Proposed diagnostic criteria for EL which have been widely accepted includes an acute or subacute encephalitic illness where all other known causes of encephalitis have been excluded. More recently it has been suggested that the diagnosis of EL may be considered if the patient's condition cannot be attributed to any other known neurological condition and that they show the following signs: influenza-like signs; hypersomnolence (hypersomnia), wakeability, ophthalmoplegia (paralysis of the muscles that control the movement of the eye), and psychiatric changes.

Treatment

There is no known cure and no clearly effective treatment. Historically, success has been claimed for steroids (anti-inflammatory drugs), anti-Parkinson's drugs and electroconvulsive therapy (ECT). During the initial stages of the illness, bodily functions need to be maintained, often involving intensive care therapy. As the condition settles it is a matter of maintaining and hopefully improving function by good physiotherapy, speech therapy and nutrition, as well as providing emotional support.

Outcome

The outcome can vary. Some people may make a full recovery, some, unfortunately, die, and for others a parkinsonian-type state may persist. Von Economo wrote "to look at these patients, one would suppose them to be in a state of profound secondary dementia. Emotions are scarcely noticeable in the face, but they are mentally intact".

*Some information contained in this factsheet is courtesy of The Sophie Cameron Trust (www.thesophiecamerontrust.org.uk)

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Thank you!

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