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Neurosyphilis Presenting as Complex Partial Status Epilepticus

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A confused and unkempt 43-year-old man was brought to hospital by the police. He was found wandering along an electric underground railway line. On admission to the accident and emergency

Fig. 1. The electroencephalogram (EEG) demonstrates a system of scalp EEG electrode placement). Heart rate 1 sec

focal epileptiform discharge in the right posterior temporal region (international 10–20 on EEG=126/min.

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111 department, he was confused and had a very mild left hemiparesis. He was admitted to the Neurology unit. The patient denied any problem, or any drug or alcohol abuse. His family reported a change in personality over 3 years with increasingly strange behaviour and bouts of depression. He had developed a strange, often unintelligible way of speaking. His wife divorced him after he disclosed that he was bisexual. He then lived with a male partner for 10 years followed by 3 years of celibacy. Although agitated, he was no longer confused and was able to maintain a lucid conversation. The hemiparesis had resolved. The only signs were a right grasp reflex and brisk tendon reflexes. A CT scan demonstrated considerable cerebral atrophy only. In view of his sexual history, his HIV status was tested and found to be negative. A lumbar puncture revealed a raised total protein of 0.8 g/l with normal glucose of 3.6 mmol/l but no pleocytosis. An EEG revealed the reason for his fluctuating confusion and neurological signs. A continuous right temporal lobe epileptiform focus with spread of activity to the parietal and occipital regions was found (fig. 1). Intravenous phenytoin followed by carbamazepine stopped the complex partial status epilepticus, and the patient improved, but required assistance for self-care. Cerebral syphilis was diagnosed on the basis of a serum VDRL titre of 1:32, a TPHA titre of 1:10,240 and a cerebrospinal fluid TPHA titre of 1 in 2,560 (VDRL, FTA and CEPTIA all positive). He was treated for 3 weeks with intramuscular procaine penicillin with probenecid and sustained a mild Herxheimer reaction. Neurosyphilis presents as general paresis of the insane in approximately 40% of patients [1], involving men 4–7 times more frequently than women [2]. The brain shrinks, and lymphocytes infiltrate the leptomeninges. There is ganglion cell loss, demyelination of cortical and subcortical fibres and widespread gliosis mostly involving the frontal and temporal lobes. Spirochaetes are identified in the brain in 50% of cases [1]. Cerebrospinal fluid is invariably abnormal [2] for total protein content, IgG fraction and serology. The earliest clinical features are subtle and may not develop for 10–15 years. Impaired intellectual efficiency, poor memory and concentration precede changes in behaviour and affect. Problems with speech and cognitive function occur. Psychiatric features including confusion, disorientation, personality change, mania, irritability and paranoid delusions. Dementia leads to poor self-care and ultimately vegetation [3–5]. Focal (simple or complex) or generalized seizures can occur in approximately 50% of cases [1,

6, 7] but status epilepticus is rare. Mask-like facies, tremor of the tongue, Argyll Robertson pupils and optic atrophy may be present, but mental changes may occur in the absence of any physical signs. The presentation may be totally asymptomatic [8].

In our patient, mental changes dominated the picture for 3 years. The confusion and fluctuating motor signs were a consequence of complex partial status. The assumption that this man had an HIV-related problem was wrong. Syphilis remains an important sexually transmitted disorder that mimics many other conditions and may present with confusion. It is an important indolent and subtle infection that can devastate the brain over a period of many years if untreated.

Following a visit from his previous male partner, the patient learned that his partner had been treated for syphilis over 10 years ago during the early stages of their relationship, but had not told the patient for fear of rejection.

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