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Neurosyphilis – The Extinct Imitator?

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CASE REPORT

The author describes a case of an organic mood disorder

Key words: Neurosyphilis, Organic mood disorder

A 37 year old Caucasian male, ZZ, was presented to casualty by his work colleagues, after he had turned up to work on his day off and been behaving unusually. On examination he was aroused, animated, and over-familiar, insisting upon using the doctor's first name, and later the Professor's. There was rapid, slurred speech, clang associations and tangentiality. He stated that there were voices in his head; that the TV and Radio were talking directly to him; and that his father had been a secret agent and his mother assassinated. He stated that he had saved a girl's life. He complained of a 4-month toothache and headache. He was too distractible for cognitive state examination and no further history was available from the patient. On physical examination there were no abnormalities elicited.

His family revealed that their father had been a diplomat and had possibly performed for the intelligence services. His mother had died in a car crash. He had not saved a girl's life as claimed. ZZ claimed to have had an accident after being 'sideswiped' on a motorway while driving a powerful motorbike at great speed; his family said it was a farm accident on a moped. ZZ had recently lost his flat after accumulating substantial debts for non-payment of his mortgage.

His supervisor reported that he had had difficulties over the last 2 years, and had required extra support in the past. ZZ had been moved repeatedly from positions of responsibility to lesser ones, he had been over-familiar with customers, via the loudspeaker system, and had had to be removed from cash related tasks as he had been less than careful in his duties.

The following morning the patient had 2 Tonic-Clonic seizures, was found to be pyrexial and had increased tone in the right side of his body. Haematology tests revealed a neutrophilia of 9.7 (2.2–7.5) and a lymphopaenia of 0.47 (1.2–4.0), overall the WCC was 11.1 (3.8–11.0). The RBC, Ht, MCV and platelet counts were all within normal parameters, as were the biochemistry results. A lumbar puncture showed an elevated

CSF protein of 0.85 (0.12–0.6) and a glucose of 5.0. Microscopy showed a RBC of 600, a WBC of 5 and negative culture growth. A viral encephalitis was presumed and intravenous antiviral treatment was started (Acyclovir 700mg TDS for 10 days).

Syphilis serology had been requested at admission. This revealed a serum TPHA level of 1 in 80,000 and a VDRL titre of 1 in 128. A repeat lumbar puncture showed 1 in 5,125 and 1 in 8 respectively. Penicillin treatment, 1.2M units daily for 3 weeks, with steroid cover, was commenced immediately.

The patient went on to make an almost complete recovery; it was very difficult to decide which of his residual deficits were due to his prior head injury, which may have been the cause of an old left frontal infarct, and which were secondary to his acute syphilitic meningitis. His residual deficits were slurred speech, mild disinhibition, impairment of executive function on the WAIS-R and Wisconsin Card Sorting Test (tests of frontal lobe function), and impaired forward planning.

Following recovery we were able to elicit the following sexual history. ZZ had no memory of genital lesions, nor had he received any treatment in the past for any sexually transmitted disease. Over the last four years since his prior partner's death he had engaged in an episode of receptive anal intercourse and visited prostitutes 1-2 times per month. He insisted that he had used condoms all of the time. Testing for HIV gave a negative response. At discharge he agreed to follow-up with his local GUM clinic, they were able to undertake contact tracing. He had no further sexually transmitted diseases.

Discussion

The prevalence of syphilitic infections of the central nervous system has dropped markedly over the century. In the United Kingdom between 1936–1939 there were 1,629 deaths ascribed to general paresis or tabes. By 1969 the three-year cumulative death toll had dropped to 224 [1]. It was feared after World War II that unwitting partial treatment with penicillin, when given for other conditions, would result in late manifestation. What appears to be happening is that the attenuated form presents in a different manner. There has been a change in the form of the illness. Previously parenchymatous neurosyphilis (tabes dorsalis or general paresis) was the most common form; nowadays meningovascular syphilis accounts for the majority of presentations [2].

Traditionally syphilis has been divided into four stages: the primary stage with lesions appearing at the site of inoculation: the secondary stage with a generalised rash appearing around weeks 4-8: the tertiary stage with destructive lesions such as gummata and bone changes: and the quaternary stage of parenchymatous changes in the central nervous system. Nowadays a division is drawn between early and late syphilis. This is arbitrarily placed at 2 years as during this period the lesions contain many trepanemes and are infectious. Early syphilis can include a spirochaetaemia which can be responsible for systemic and neurological symptoms. The perception that neurosyphilis is a late manifestation of infection is incorrect.

Meningovascular disease usually present(s) within 1–5 years of the primary infection. Diffuse inflammation, leading to thickening and necrosis, and vascular

pathology occurs. When vascular pathology is predominant the presentation tends to be neurological. Otherwise there is a mix of emotional instability and irritability, impaired concentration and judgement, with increasing forgetfulness. As the condition progresses fleeting delusions and clouding of consciousness can occur [3]. Some sources state that a Lumbar puncture will reveal a high white cell count; it has also been reported that pleocytosis is uncommon (10%) in HIV seronegative patients [4,5]. In slightly more cases (10-30%) protein is present [4,5]. In this case some potein was present but it was thought to be secondary to blood contamination.

Acute syphilitic meningitis is characterised by pyrexia, headache, delirium, somnolence and neck stiffness. It occurs within 2 years of infection. However the presence of Tonic-Clonic seizures leads to a diagnosis of diffuse meningo-encephalitis. A grandiose, fatuous affect can be a presentation of general paresis, but the long-standing existence of these traits in this patient, confirmed by collateral informants, precludes this diagnosis. This patient's presentation could have been due to drug intoxication, mania, an acute polymorphic psychotic disorder or a combination. It was only due to a comprehensive history coupled with a high index of suspicion that appropriate investigations were performed. This case highlights that with the decline of a once-common condition, treatable causes of delirium and dementia might be missed. It is vital that practitioners recognise acute confusional states and investigate them appropriately and that patients with a sexually transmitted disease are referred to specialists for follow-up and contact tracing.

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