Dementia and white matter demyelination in young patient with neurosyphilis

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Study concept and design: Brinar and Habek. Acquisition of data: Brinar and Habek. Analysis and interpretation of data: Brinar and Habek. Drafting of the manuscript: Habek. Critical revision of the manuscript for important intellectual content: Brinar and Habek. Administrative, technical, and material support: Habek.

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In September 2005, a 38-year old male patient from Bosnia and Herzegovina was admitted to our Hospital because of memory impairment and speech disturbance. Symptoms have started in January 2003. At that time his neurological examination was normal except cognitive impairment. MRI of the brain was performed, and multiple white-matter hyperintensities were noted (Figure 1A). The patient refused any kind of medical treatment and further work-up. One year later he was again seen by neurologist in another institution, this time because of moderate spastic paraparesis with mild ataxia and dysarthria. Serum tests for syphilis were positive (Treponema pallidum haemagglutination assay (TPHA) was 1:2560). CSF was obtained and revealed normal cell count with elevated proteins and IgG index. MRI was repeated and showed progression of white matter hyperintensities. He was started on penicillin therapy, which he stopped taking after 10 days of treatment and did not receive any kind of medical attention during the next year.

On admission to our hospital the patient was disoriented with episodes of aggressiveness. His MMSE was 9/30. The rest of neurological examination was normal. Standard laboratory tests, thyroid hormones, vitamin B12 and folic acid were normal. HIV and hepatitis B and C serology was negative. Protein 14-3-3, s-100 and TauAg in CSF were normal. CSF revealed normal cell count and protein level, with positive oligoclonal IgG bands (OCB) indicating intrathecal synthesis. Syphilis serology in serum and CSF was positive as follows: serum Venereal Disease Research Laboratory test (VDRL) was 1:32, TPHA 1:5120 and IgG Fluorescent Treponemal Antibody Absorption Test (IgG-FTA-ABS) 1:320, and CSF TPHA was 1:160, IgGFTA-ABS 1:4. Brain MRI was repeated and revealed numerous white-matter hyperintensities (Figures 1B and C). The lesions did not show post contrast enhancement. MRI of the cervical spinal cord was normal. The patient was started on a 4 week course of 24 million i.v. cristacillin per day, and at last follow-up in March 2006 was without new neurological symptoms with MMSE 11/30.

Syphilis is not one of the major health problems in Croatia. In 1999, twenty new patients with syphilis were diagnosed, however number of infected patients is on an increase in a neighboring countries, especially in Russia and other Eastern European countries.[1,2] On the other hand, syphilis is increasing in the United States and Western Europe as well. However, whereas in Russia it is evenly distributed among men and women, the 2003 epidemic in the US and Western Europe mainly involves men who have sex with men.[3] This increasing number of patients can be partly explained with rising HIV infection and with different sexual behaviour, travel and migration, political, economic and social factors between these regions.

Our patient with neurosyphilis was immunocompetent with numerous white matter hyperintensities resembling leukoaraiosis, which is a rare finding in this disorder. The syphilis is known as a great mimicker, and the same can apply for neurosyphilis. Potential neurological diseases that can be mimicked by neurosyphilis on neuroimaging are herpes simplex encephalitis, mesial temporal sclerosis, leukoaraiosis, normal pressure hydrocephalus and glioblastoma multioforme.[4] But patients with neurosyphilis can also have normal brain MRI or just temporal lobe atrophy. CSF findings may vary from positive OCB and higher IgG index to just pleocytosis with or without positive CSF VDRL. Thus, diagnosis of neurosyphilis can be challenging even with today modern tests. Guidelines for the diagnosis of dementia

published by American Academy of Neurology do not recommend “routine” syphilis screening or CSF examination in the dementia evaluation.[5] However, clinicians should bare in mind that dementia in young patients with temporal lobe atrophy and/or demyelination can be caused by syphilis and thus necessitates CSF and syphilis testing.

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Figures:
Figure 1. A Brain MRI performed in 2003, FLAIR sequences. B and C Brain MRI performed in 2005, FLAIR sequences. White matter hyperintensities (A) are visible on the border of white and gray matter on scans from 2003. The progression of lesions (B), as well as demyelination in the deep white matter of temporal gyri and hypocampal regions (C) are visible in scans from 2005.

Conflict of interest statement
We have no conflict of interest.

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Figure 1.