NEUROSYPHILIS IN A YOUNG PATIENT PRESENTED AS RAPID COGNITIVE DECLINE

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ABSTRACT

Syphilis is a sexually transmitted infection caused by Treponema pallidum, known to have three distinct phases. The central nervous system involvement may occur as early as the second phase generally with syphilitic meningitis, but neurosyphilis always occurs in the tertiary phase with meningeal, meningovascular or parenchymal manifestations. We present the case of a rapid cognitive decline caused by neurosyphilis in a young adult, in our knowledge a very rare presentation in current practice. Even with a constant reduction of the incidence worldwide, syphilis should always be considered in young patients with rapid cognitive decline and other neurologic manifestations.

Keywords: normal pressure hydrocephalus, neurosyphilis, dementia

INTRODUCTION

Syphilis is a sexually transmitted infection caused by *Treponema pallidum* that can involve the central nervous system usually presenting the syphilitic meningitis or meningo-vascular syphilis especially in the patients with AIDS. Even though the worldwide incidence of parenchymal neurosyphilis has decreased in the last decades, one could suspect it in young patients with cognitive decline accompanied by other neurologic manifestations (1). The clinical picture of the paretic neurosyphilis, historically known as the "general paresis of the insane", is revealed by dementia, seizures, tremor, pyramidal signs and Argyll-Robertson pupils. The onset is usually after 15 to 20 years after a non-treated initial infection. In this paper we report the case of a young patient with neurosyphilis with rapid progression of the disease.

CASE REPORT

We present the case of a 38 years old Caucasian male who was admitted for headache, rapidly progressive mental decline accompanied by bizarre behavior and infrequent myoclonia of the lower limbs. His medical family history reclaims no pathology. In the last months prior to hospitalization his family observed progressive behavior and personality changes. The neurologic examination showed spastic tetraparesis, predominantly in the lower limbs, brisk tendon reflexes, extensor plantar response, asymmetric and infrequent myoclonic jerks in the lower limbs, no sensory or coordination impairment, apparently bradypsychia, temporal and spatial disorientation, amnestic aphasia and ideomotor apraxia. MRI revealed a normal pressure hydrocephalus with no other acute pathological changes (Fig. 1). The neurosurgical consult rec-

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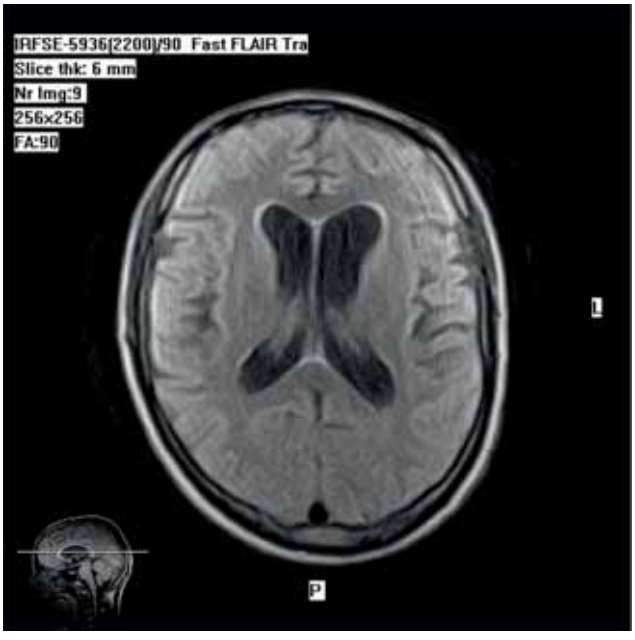

FIGURE 1. *Axial FLAIR MRI suggesting moderate normal pressure hydrocephalus*

ommended a ventriculoperitoneal shunt and the patient was admitted to Neurosurgery Department. On February 2013 surgery was practised with minimum right temporo-occipital craniectomy, with the introduction of a ventricular catheter in the right lateral ventricle. Ventricular catheter was connected to a low pressure delta valve, also connected to peritoneal drainage catheter. Ten days after surgery, Mini Mental Status Scale score was 12 with marked bradypsychia, bradylalia, long latency for verbal responses, temporal and spatial disorientation, without psychotic manifestations, short and medium time memory alteration, amnestic aphasia, ideomotor apraxia. There was no major improvement in the neuropsychological tests as expected after the vetriculoperitoneal shunt. A neurological reevaluation found a light-near dissociation pupil on the right side (Argyll-Robertson pupil), spatial and temporal disorientation, frontal cortical release reflexes, moderate cognitive decline and spastic tetraparesis. An EEG showed pseudoperiodic slow wave discharges with bisynchronous appearance hinting towards an encephalopathy of unknown origin (Fig. 2). Further on, biochemistry and serology showed the following: serum TPHA – positive at 1/20480 dilution, serum VDRL – positive at 1/32 dilution, CSF TPHA – positive at 1/10240 dilution, CSF VDRL – negative, CSF proteins – 0.55 g/L (0.15-0.45), CSF glucose – 52 mg/dL (40-70), CSF chloride – 118 mEq/L (110-130), CSF cells – 12 elements/mm^3 (neutrophils and lymphocytes). CSF and serum HIV were negative, otherwise normal values on laboratory findings. The diagnosis was established as dementia in neurosyphilis and normal pressure hydrocephalus. Our differential diag-

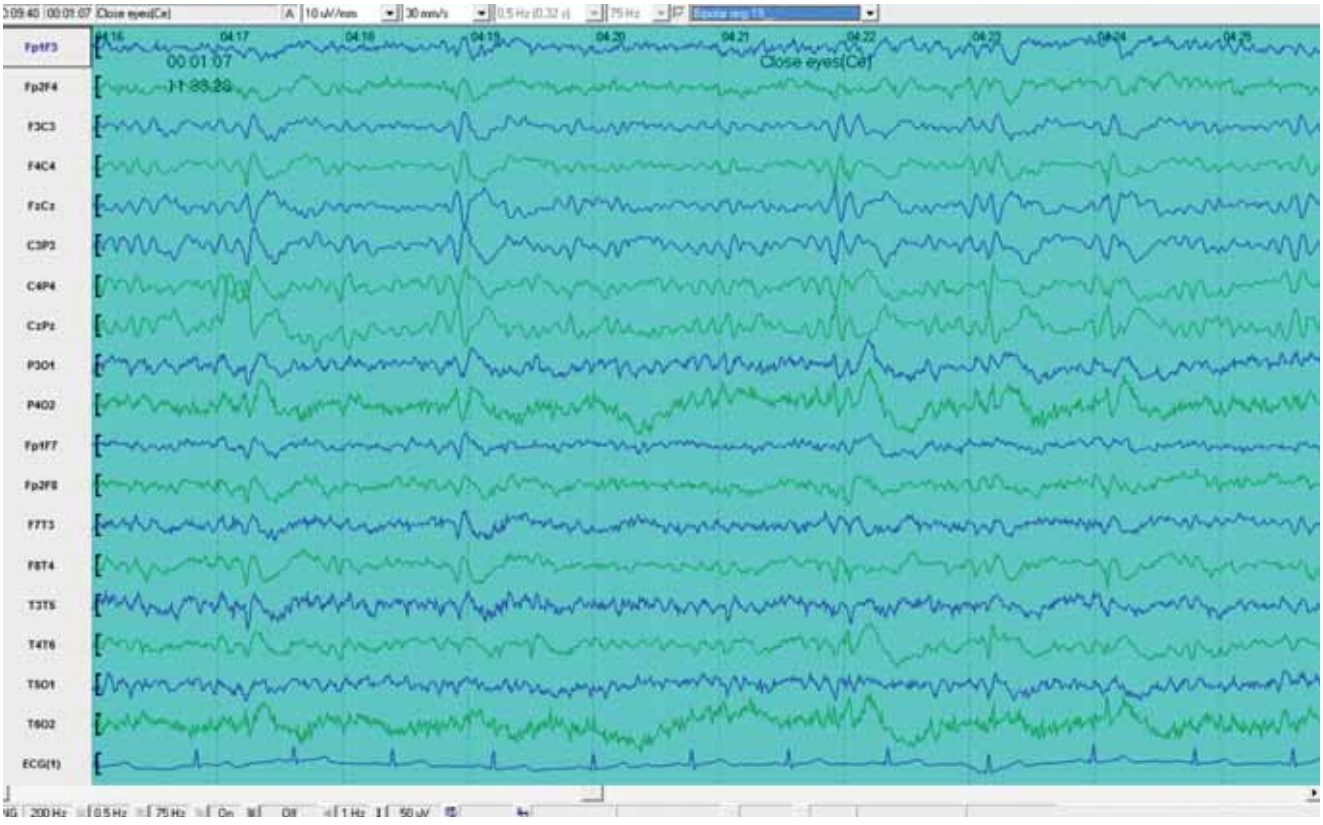

FIGURE 2. *Pseudoperiodic bisynchronous slow wave discharges with triphasic appearance*

nosis included an early onset Alzheimer's disease, Creutzfeldt-Jakob disease, HIV – Dementia Complex, metabolic encephalopathy. Alzheimer's disease has a more slow progression with memory loss, apraxia, aphasia, spatial and temporal disorientation complicated with psychotic manifestations. Such an early onset characterizes the familial form of dementias. CJD is a prion disease manifested with rapid dementia, with psychotic manifestations and the presence of specific MRI appearance and positive 14.3.3 protein in the CSF, but in some stages of the disease the EEG shows a pseudoperiodic pattern of triphasic sharp waves. HIV – Dementia complex was ruled out by the imagistic and virology studies. No metabolic changes were found in order to suspect a metabolic encephalopathy, but virtually every metabolic encephalopathy can manifest as rapid and progressive dementia. The treatment was started with wide spectrum antibiotics (Ceftriaxonum 4 g daily for 14 days). Mild improvement of the symptoms was observed, but the patient must be reassessed periodically. Psychiatric evaluation will be assessed until cognitive stabilization is reached.

CONCLUSIONS

Neurosyphilis is classically subdivided in: syphilitic meningitis, meningo-vascular syphilis, general paresis and tabes dorsalis. The clinical presentation can combine from the following: spinal cord lesions (tabes, Brown-Séquard syndrome, acute or subacute myelopathy), cranial nerve involvement, spinal nerve root involvement secondary to arachnoiditis, encephalopathy with seizures, stroke, neuropsychiatric syndrome, ocular involvement (1). Diagnosis requires a combination of tests. VDRL – CSF, the standard serological test for CSF, is highly specific, but insensitive (2,3). Therefore, a negative VDRL – CSF result does not rule out neurosyphilis. FTA – ABS is more sensitive than VDRL – CSF, but less specific. Therefore, the CSF FTA – ABS test is useful to exclude neurosyphilis (4). Cognitive decline is one of the late syphilis manifestations. However, mild cognitive impairment has been observed in early stages of neurosyphilis and dementia is the most common presentation of neurosyphilis (5,6). Neurosyphilis is associated with cognitive decline and progressive dementia. A small number of cases have been described mimicking Alzheimer's disease on imagistic studies (7). In the differential diagnosis of a subacute dementia in an adult patient neurosyphilis should by ruled out. EEG pattern can include periodic or pseudoperiodic slow sharp waves discharges sometimes PLEDs with potential misdiagnosis to subacute encephalopathy or encephalitis (8). The signs are not pathognomonic and the sensitivity and specificity of supportive laboratory tests are unclear. Widely accepted, a positive VDRL in the CSF is sufficient to diagnose neurosyphilis, although false positive results caused by contamination from serum are possible. Follow-up clinical examination and lumbar puncture should be scheduled at 3-6 months after treatment and every 6 months thereafter until the CSF – VDRL becomes nonreactive.

We consider this case a particular one because of the rapid progression of a complication that usually last for years until it is developed if syphilis is untreated. *Per primam* address to the neurosurgery unit with ventricular drainage for hydrocephalus with good outcome, nonspecific presentation with discussions upon the differential diagnosis and the particular EEG aspect also reflects the particularity. The need of syphilis serology should always be considered in young patients with rapid cognitive decline. Further forensic and psychiatric evaluations are impotant because of the young age of the patients with neurosyphilis who become demented and may be in need of institutionalization or, reversely, who recover after treatment and the reassessment of cognitive tests can quantify the level of personal and social independency.

Abbreviations

AIDS – *acquired immunodeficiency syndrome*, MRI – *magnetic resonance imaging*, EEG – *electroencephalography*, TPHA – *Treponema pallidum hemaglutination assay*, CSF – *cerebrospinal fluid*, VDRL – *venereal disease research laboratory*, FTA – ABS – *fluorescent treponemal antibody – absorbtion*, CJD – *Creutzfeldt-Jakob disease*, HIV – *human immunodeficiency virus*, PLED – *periodic lateralized epileptiform discharges.*

Consent

This case report was approved for publication by the Local Ethical Committee of the Emergency Clinical County Hospital Sibiu, Romania.

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