Neurosyphilis Presenting As Status Epilepticus, Successively Hemiparesis And Aphasia

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Abstract

Background: Neurosyphilis can occur anytime and present with a myriad of symptoms. Lissauer form of General Paresis Insane (GPI) is rare. We can learn more about this form of GPI through this case report.

Case Presentation: The patient presented as status epilepticus, successively as hemiparesis and aphasia, which may be considered as the Todd's paresis or stroke. By performing the reactive serum rapid plasma reagent test and cerebrospinal fluid analysis, as well as the brain MRI results, we made the diagnosis as Lissauer form of GPI. The patient was started on intravenous penicillin for a total of 14 days. After that, the patient appeared with marked clinical improvement. Cognitive ability was better than before.

Conclusions: GPI typically has a progressive course and normally presents 10 to 30 years after the initial infection. The manifestations of this patient and his suspicious history of Transient Ischemic Attacks (TIA) may mislead to the diagnosis of Todd's paresis or stroke. The prevalence of syphilis is rising again in recent years. To date, there is no gold standard for the diagnosis of neurosyphilis. Early diagnosis is of great importance as effective penicillin therapy is available.

Background

Neurosyphilis can occur anytime and present with a myriad of symptoms. Status Epilepticus (SE) can be found in neurosyphilis, however, successively presenting as hemiparesis and aphasia is very rare, which may be misdiagnosed as other diseases. To date, there is no gold standard for the diagnosis of neurosyphilis. We report a case of Lissauer form of GPI, which is very rare. Early diagnosis is of great importance as effective penicillin therapy is available. To prevent the Jarisch-Herxheimer reaction, it suggests using hexadecadrol or prednisone before intravenous dripping high dose of penicillin.

Case Presentation

A 56-year-old right-handed Chinese man presented with loss of consciousness and convulsion of limbs. He was sent to the emergency department of Wenzhou Central Hospital. Brain CT showed senile changes, encephalatrophy and leukoaraiosis. Intracranial artery and carotid artery CTA revealed that there were mixed plaques at the bifurcation of the left common carotid artery, multiple calcifications in the intracranial segments of both internal carotid arteries, and localized stenosis in the A1 segments of the right anterior cerebral artery. The status epilepticus were relieved by using diazepam. Then, the patient gradually developed paroxysmal convulsion of limbs four to five times per day, still confused in the interictal stage. Several days later, the patient's consciousness had improved, whose clinic manifestations were characterized by left hemiparesis (Medical Research Council grade [MRC] 1/5) and global aphasia. Then, the patient was transferred to the neurology department of Chongqing Fuling People's Hospital.

The patient had a body temperature of 36.5°C, heart rate of 92 beats per minute, respiratory rate of 20 per minute, and blood pressure of 136/94 mmHg. The patient tested positive for syphilis as evidenced by

reactive serum rapid plasma reagent test (titer, 1:1280). Further tested positive for neurosyphilis as evidenced by cerebrospinal fluid (CSF) analysis (titer 1:5120), with negative antibodies of autoimmune encephalitis. His other laboratory tests showed no significant abnormalities. Brain MRI revealed that: 1. Demyelination of deep brain white matter and encephalatrophy [Fig1, A and B]; 2. acute spotted infarctions in the semiovale center [Fig1, C and D]. Enhanced MRI image of brain showed meningeal enhancement [Fig1, E and F] and cerebral surface enhancement [Fig1, G and H]. The electroencephalogram showed that the EEG was moderate abnormal [Fig2].

The patient had a suspicious past medical history of TIA 8 years ago. After that, he irregularly took medicine of “clopidogrel” and “atorvastatin”.

The members of his family found that his personality changes with deteriorating social interactions, progressive forgetfulness, and apathy gradually. The patient became unable to work and care for his family. He could just handle some simple house work, following other's instructions, such as wash dishes and pick up little children from school to home.

He was started on intravenous (IV) penicillin for a total of 14 days, 4 million units each time, Q4H. On the first day of IV penicillin, the patient presented with transient convulsion of left limbs, without loss of consciousness, which stopped by using diazepam. Clinically, the patient's neurologic examination progressively improved. He appeared with marked clinical improvement of the paraparesis inferior (MRC 4/5) and could walk unaided. He can also communicate with others in vague voice. Faculty of understanding and memory was better than before, while the ability of calculation was still poor.

Discussion And Conclusions

Neurosyphilis refers to infection of the central nervous system the spirochete *Treponema pallidum*, which can occur anytime between the initial inoculation and the late stage of tertiary syphilis^[1]. Early neurosyphilis may be asymptomatic or include vasculitis, meningitis, stroke, dementia, vertigo, optic neuritis and uveitis. Late or tertiary neurosyphilis is characterized by chronic infection of the brain parenchyma or the posterior columns of the spinal cord. It's easy to misdiagnose the disease, for the lack of specificity and diverse manifestations^[2]. GPI typically has a progressive course and normally presents 10 to 30 years after the initial infection. The onset of age is usually 40-50 years old, which may lead to forgetfulness and personality changes, dementia, impaired proprioception, gait imbalance and epileptic seizures^[3]. It is unclear when our patient was initially exposed to syphilis. To his wife's knowledge, he had worked outside more than 20 years and seldom back home. He had showed personality changes with progressive cognitive decline and apathy since encountered from TIA 8 years ago. The patient started to present as status epilepticus, successively showed as left hemiparesis and global aphasia in this course. These manifestations may mislead to the diagnosis of Todd's paresis or stroke. There is no gold standard for the diagnosis of neurosyphilis up to date, which is dependent on clinical findings and the results of serologic tests and CSF abnormalities. Unfortunately, no consensus has been reached regarding diagnostic criteria about serologic tests and CSF examinations^[2]. The inspection of MRI gave

the hint that he had severe brain atrophy, mainly in hippocampus and temporal lobe. Enhanced brain MRI showed diffused enhancement in cerebral surface and right meninges, while not obvious in hippocampus and temporal lobe. Combining with the other laboratory tests, we considered it as Lissauer form of GPI^[4]. Status epilepticus, successively presenting as left hemiparesis and global aphasia were supposed to be the manifestations of acute exacerbation in the course of chronic disease.

Neurosyphilis is no longer a common disorder, but the prevalence of syphilis is rising again in recent years. Early diagnosis is of great importance as effective penicillin therapy is available. It can also be replaced by tetracycline or ceftriaxone if allergic to penicillin. To prevent the Jarisch-Herxheimer reaction, it suggests that using hexadecadrol or prednisone 3 days before intravenous dripping high dose of penicillin^[5]. It's advised intramuscular injecting benzathine penicillin biweekly for a total of 6 weeks, successively once every month for a total of 6 months, and following up observation regularly after finishing intravenous penicillin.

Abbreviations

GPI: General Paresis Insane; SE: Status Epilepticus; TIA: Transient Ischemic Attacks; IV: intravenous; CSF: cerebrospinal fluid

Declarations

Ethics approval and consent to participate

This study was approved by the Ethical Committee of Chongqing Fuling People's Hospital. Written informed consent was obtained from the patient for publication of this case report.

Consent for publication

Written informed consent was obtained from the patient for publication of this case report.

Availability of data and materials

The dataset supporting the conclusions of this article is included in the article.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

Yunwen Qi: Drafting and revision of the manuscript for content, including medical writing for content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data.

Zongze Jiang: Revision of the manuscript for content; Major role in the acquisition of data; Study concept or design; Analysis or interpretation of data.

Xiong Zhang: Major role in the acquisition of data; Analysis or interpretation of data.

Daixin Xie: Major role in the acquisition of data; Analysis or interpretation of data.

Zhiyou Cai: Revision of the manuscript for content; Analysis or interpretation of data.

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Figures

A and B showed severe brain atrophy, mainly in hippocampus and temporal lobe (arrows); DWI showed high signal (C), ADC showed low signal (D); E, F, G and H showed diffused enhancement in cerebral surface and right meninges.

Figure 1

Brain MRI and Enhanced MRI image

Diffusely low-voltage α waves (10.0-12.0Hz), had no obvious dominance in Occipital. Multiple θ waves with medium-high amplitude (3.0-4.0Hz) were scattered in both leads.

Figure 2

EEG of the the patient upon first admission

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