Case Report

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Neurosyphilis with Hydrocephalus: A Case Report

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ABSTRACT

Neurosyphilis is defined as central nervous system involvement by treponema pallidum bacteria. Symptomatic neurosyphilis can be manifested as acute or subacute meningitis (a type of meningitis) that emulates other bacterial infections. Hydrocephalus and cranial nerve paralysis (VII and IX) may occur. In this article, we report a case of congenital hydrocephalus neurosyphilis, with a significant improvement in neurological condition after treatment with penicillin-G. The infant was a 2.5-month-old boy who referred to the emergency department because of fever. On initial examination, the head had been larger than usual. The patient was evaluated with suspicion of sepsis. Cerebrospinal fluid (CSF) analysis was consistent with meningitis and hydrocephalus found in ultrasound. Due to lack of response to antibiotic and anti-tuberculosis (TB) treatments in improvement of CSF analysis, ultimately after positive CSF serology in favor of syphilis, treatment changed into penicillin; then clinical and laboratory findings were improved. The rare manifestation of congenital syphilis as hydrocephalus and the appropriate treatment response to penicillin were interesting points for the introduction of this patient. We presented a case of neurosyphilis, which was characterized by a cognitive and neurological deficits, hydrocephaly, and myoclonus, as well as irritability and hearing loss. Since syphilis is easily diagnosed and treatable, it should be considered and evaluated in patients with cognitive defects and motor disorders. Misdiagnosis of syphilis is a serious medical mistake that may cause long-term consequences.

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Introduction

Neurosyphilis is defined as central nervous system involvement by treponema pallidum bacteria. The annual incidence of neurosyphilis varies from 0.16 to 2.10 per 100000 people (1, 2). Congenital syphilis is caused by the treponema pallidum bacteria, which is passed from mother to child during fetal development or at birth. Infection can lead to prematurity, abortion or multiple clinical features. Only severe cases are seen at birth (3, 4).

Syphilis progresses to four stages without an essential treatment: primary, secondary, late, and late latent (2, 5). Neurosyphilis can be manifested as acute or subacute meningitis (a form of meningitis) that mimics other bacterial infections (5, 6). Hydrocephalus and cranial nerve paralysis (VII and IX) may occur (7). Venereal disease research laboratory (VDRL) test is a nontreponemal test that is used for diagnosis of infection with syphilis. Serum and cerebrospinal fluid (CSF) are usually strongly positive, and the disease responds appropriately to penicillin (2, 4, 5). Neurosyphilis is classified as early and late syphilis. CSF, meninges, and vascular structures are involved in the early stages of neurosyphilis; while in the late stage, CSF and cerebrospinal parenchyma are affected, and sometimes lead to hydrocephalus (8, 9).

In this article, we report a case of congenital hydrocephalus neurosyphilis, with marked clinical improvement in neurologic assessment after treatment with penicillin-G.

Case Report

A 2.5-month-old boy was referred to the emergency department because of fever. He was the first child of the family. His parents were not relatives and had a moderate level of education. He had no history of a specific illness, and the primitive reflexes had been decreased. The vital signs included: temperature (T): 39 ºC, respiration rate (RR): 64 BPM, pulse rate (PR): 150 BPM, blood pressure (BP): 10.l57 mmHg.

On physical examination, the head was larger than normal, the heart's sound was normal, the lung fields appeared clear, the abdomen was soft, the liver felt like edge under right part of the rib cage. Early recognition and management of sepsis was done. Chest x-ray was normal. Antibiotic treatment including ceftriaxone and vancomycin, as well as supportive care was initiated.

Laboratory tests' results were as follows: white blood cell (WBC): 10.8 x 10^3/µl, hemoglobin (Hb): 10.3 g/dl, platelet (PLT): 331 x 10^3/µl, polymorphonuclear (PMN): 33.4%, lymphocyte: 56.9%, blood urea nitrogen (BUN): 6 mg/dl, Cr: 0.5 mg/dl, Ca: 9 mg/dl, P: 3.5 mg/dl, Mg: 2.2 mg/dl, Na: 135 meq/l, K: 3.9 meq/l, blood sugar (BS): 109 mg/dl, urine analysis (U/A): normal, specific gravity (SG): 1010

CSF Analysis: WBC: 35 cells, PMN: 30%, lymphocyte: 70%, red blood cell (RBC): 180 cells, glucose of CSF (GLU): < 20 mg/dl, protein: 890 mg/dl

Cranial sonography showed hydrocephalus, CSF collection retested:

WBC: 45 cells, PMN: 30%, lymphocyte: 70%, RBC: 200 cells, GLU: < 20 mg/dl

Seizure was added to the patient's symptoms, and anticonvulsant drug was added to treatment. In the consultation with neurosurgery, ventriculoperitoneal (VP) shunting was suggested.

However, antibiotic was suggested because of the presence of low glucose in spinal fluid analysis, and also evidence of brain infections. In order to cover resistant pneumococci, rifampin was added to the patient's treatment. For completing the duration of antibiotics treatment, CSF analysis was performed after 10 days. The fluid sugar was still below 20 mg/dl, and no change was observed in the analysis. To cover probable listeria monocytogenes, ampicillin was added to the treatment. Bacteria and viruses polymerase chain reaction (PCR) composition, evaluated by PCR.TORCH study, was negative. The immune system was evaluated. After antifungal treatment, analysis of the CSF showed treatment failure:

WBC: 38 cells, PMN: 40%, lymphocyte: 60%, RBC: 10 cells, GLU < 20 mg/dl, protein: 900 mg/dl

Despite the negative history of tuberculosis (TB) in his family, the patient was treated with anti-TB, but there was no response in the clinical condition and CSF.

Finally, CSF-VDRL test was reported with a high titer, and the patient was treated with intravenous penicillin. The family was referred to an adult infectious disease specialist.

After two weeks treatment with intravenous penicillin, CSF analysis proved to be acceptable. VP shunt placement was done; seizures and neurological symptoms improved.

In follow-up, the patient was able to sit and nip, and CSF-VDRL test was negative.

Discussion

The uncommon manifestation of congenital syphilis such as hydrocephalus, and the appropriate therapeutic response to penicillin were the interesting points to introduce this patient.

Hydrocephalus at birth can be caused by intrauterine infections, TORCH infections, genetic disorders, and anatomical defects of the brain. The most common cause includes TORCH and genetic disorders that are extremely fascinating in our case (10, 11). Congenital infection with treponema pallidum causes a wide range of symptoms, from unusual infection to severe cases seen at birth. A baby or child may have symptoms like hepatosplenomegaly, rash, condylomalata, snuffles, jaundice (non viral hepatitis), pseudoparalysis, anemia or edema (nephrotic syndrome and/or malnutrition) (10, 12). The CSF-VDRL has high diagnostic specificity. This technique can be used to monitor response to drug treatment (13-15). In this case, the diagnosis and response to treatment with the serological test were consistent with those documented in other studies, which confirmed the diagnosis. At the same time, the importance of the CSF-VDRL test was noted.

In our patient, clinical manifestations began with hydrocephalus (Figures 1-3), and then gradually worsened with cognitive decline and seizures. His past medical history did not reveal any clinical and skin signs and symptoms that referred to the primary and secondary stages of syphilis (16), which is very rare and important in this regard. We live in time of antibiotic production, due to which prevalence of syphilis has declined, although resistance to penicillin has been identified rarely.

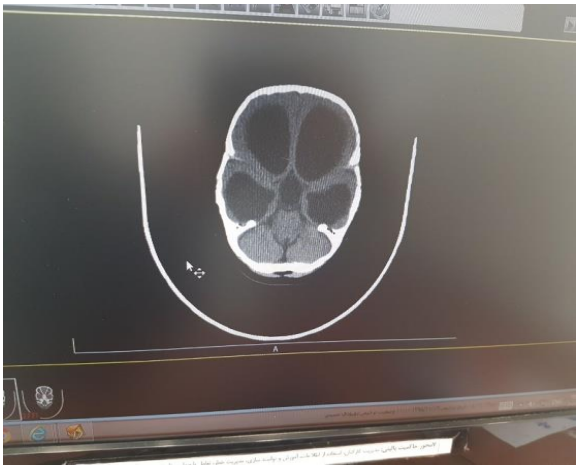

Figure 1. Brain computed tomography (CT) scan

Neurosyphilis requires 3-4 million penicillin G units every 4 hours for 10 to 14 days, and follow-up every 6 months with a CSF analysis. After 6 months, the cell count should be declined in CSF, and the number of cells and protein in the CSF should be normal after 2 years. In our patient, follow-up after 6 months is appropriate to this point (2, 4, 5, 7-19).

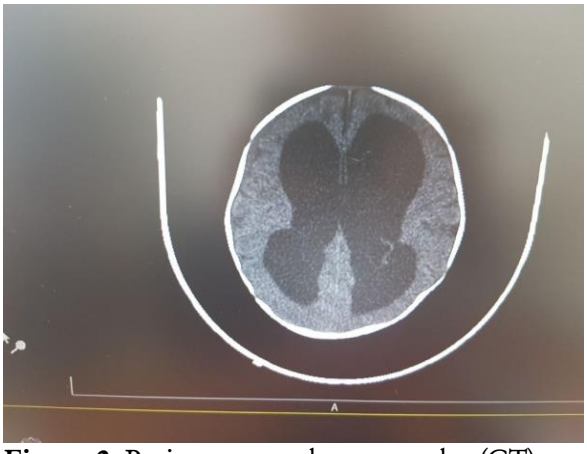

Figure 2. Brain computed tomography (CT) scan

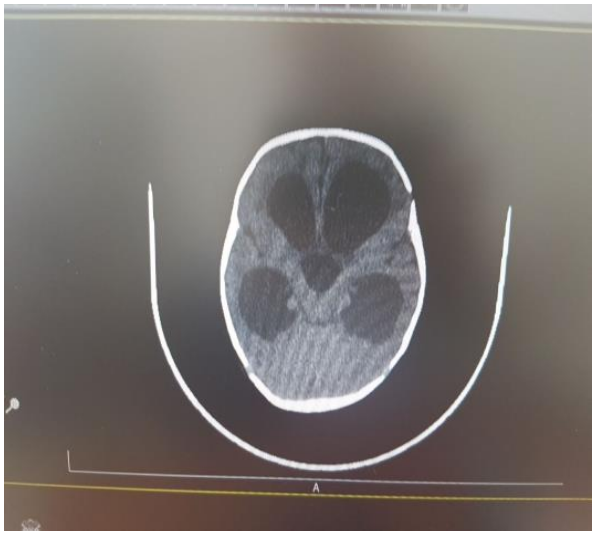

Figure 3. Brain computed tomography (CT) scan

It is significant that congenital syphilis is a lethal but curable disease. Serial tests should be performed because early and timely diagnosis and initiation of appropriate therapy are crucial (20).

We presented a case of neurosyphilis, which is characterized by a cognitive decline, neurologic disorder, seizures, hydrocephalus, and myoclonus, as well as irritability and hearing loss. This was more likely to be the type of cerebral parenchymal infection that is the clinical type of congenital syphilis.

The patient was treated with penicillin and his symptoms (seizures and neurologic dysfunction) greatly improved. Since syphilis is easily diagnosed and treatable, it should be considered and evaluated in patients with cognitive defects and motor disorders. Misdiagnosis of syphilis is a serious medical mistake that may cause long-term consequences.

Conflict of Interests

Authors have no conflict of interests.

Acknowledgments

None.

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