Neurosyphilis manifesting as spinal transverse myelitis

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Summary. Spinal myelitis caused by neurosyphilis is an extremely rare disease, and there are only few visual examples of magnetic resonance imaging scans. We present a clinical case of neurosyphilis, which is of great importance concerning diagnostic, differential diagnosis, and tactics of management. A patient complaining of progressive legs weakness, numbness, and shooting-like pain in the legs as well as pelvic dysfunction was admitted to the hospital. Neurological examination revealed spinal cord lesion symptoms: legs weakness, impairment of superficial and deep sensation together with pathological symptoms in the legs. Hernia of intervertebral disc or tumor was suspected, and myelography with computed tomography of the spine was performed. No pathological findings were observed. More precise examination of the patient (a small scar in the genitals and condylomata lata in anal region were noticed) pointed to possible syphilis-induced spinal cord lesion. Serologic syphilis diagnostic tests (Treponema pallidum hemagglutination assay, reagin plasma response, serum enzyme-linked immunosorbent assay) and cerebrospinal fluid tests (general cerebrospinal fluid test and Venereal Disease Research Laboratory test) confirmed the diagnosis of neurosyphilis. Spinal cord lesion determined by magnetic resonance imaging was evaluated as spinal syphilis or syphilis-induced myelitis. Conventional treatment showed a partial effect.

Introduction
Syphilis is a chronic infectious disease caused by the bacterium Treponema pallidum. It is usually acquired by sexual contact with another infected individual. It progresses, if untreated, through primarily, secondary, and tertiary stages. In about 30% of untreated patients late disease of the heart, central nervous system or other organs develops (1). Neurosyphilis can be asymptomatic or cause different disorders, like tabs dorsalis, general paresis, and meningovascular syphilis. There are also two types of spinal syphilis other than tabs dorsalis: syphilitic meningomyelitis and meningovascular syphilis. Neurological symptoms can appear in any stage of syphilis. During secondary syphilis lesion of central nervous system develops in up to 40% of the patients.

The incidence of syphilis decreased significantly with the introduction of penicillin in the 1940s but rose sharply again with the advent of human immunodeficiency virus (HIV) infection in the 1980s. Despite the overall decreases, outbreaks of syphilis have recently been reported in men who have sex with men. Some epidemiological studies suggest that the incidence of primary syphilis is rising (2, 3). The prevalence of syphilis in Lithuania decreased over the last years from 101.4 cases per 100,000 in 1996 to 9.9 cases per 100,000 in 2004 (4).

This clinical case report is important not only because of its clinical value, but also it has scientific value. Despite significant scientific achievements in biology, not all biological features of treponemes are known. Nervous system lesions caused by syphilis are not so frequent to draw inferences about them from larger patient groups. This makes every case important for the future conclusions.

Presentation of the case
A 38-year-old man was admitted to the hospital because of shooting low back pain, radiating to the legs, legs weakness, numbness, and retained urination with constipation. The patient had been well until four months ago, when he felt that his legs became weaker.
and he could not walk as good as before. He thought that he got cold while digging a well because of working in cool and wet environment. Weakness and numbness in his legs deteriorated, and one month before admission to the hospital the patient could not walk. Sharp shooting pain radiating to his legs appeared. Couple days before the admission patient began having difficulty emptying his bladder and had constipation. There was no history of a tick bite, head injury, seizure disorder, recent viral illness, diabetes, hypertension, and intravenous drug abuse. Patient denied having accidental sexual relations. His girlfriend died one and a half years ago.

The patient did not have fever, chills, cough, rash, weight loss, or neurological symptoms related to the arms. Patient was jobless and got only some occasional utility works. The temperature was 36.7°C, the pulse was 72 beats/minute, and the respirations were 24 times per minute. His blood pressure was 120/80 mmHg. On general physical examination, the patient appeared well.

Neurological examination showed normal motor function in the arms. Muscle strength in the legs was 3/5 in the proximal and distal muscle groups bilaterally. The deep-tendon reflexes were ++ in the arms; the knee jerks were brisk ++ +; ankle jerks were diminished, + bilaterally. Abdominal reflexes were absent. Pinprick and thermal sensations were absent below the costal margins. Joint-position sense and vibratory sensation were absent in the legs. Pathological Babinski signs were observed in both feet. Straight-leg rising to an angle of 70 degrees provoked hamstring pain on both sides. Coordination was normal in the arms but poor in the legs. The patient could barely walk unsupported. A small scar was noticed on the dorsal surface of the penis. Later on, condylomata lata in anal region were observed. Neurological symptoms (loss of pain and thermal sensations bellow costal margins, impaired deep sensations in the legs as well as muscle weakness in the legs, pathological Babinski symptoms, and impaired urination together with bowel function) pointed to spinal cord lesion localized in the thoracic area. Intervertebral disc herniation of this area was suspected with further differentiation of other possible causes of spinal cord injury (spinal tumor, abscess, multiple sclerosis (MS), HIV induced myelopathy and others).

Myelography with subsequent computed tomography of lumbar part was done. These tests showed no herniation of the disc or neoplasm. Further examination was targeted to find the etiological basis of the myelopathy. The urine was normal as were the results of routine hematological, blood chemical findings. A lumbar puncture was performed (Table). Cerebrospinal fluid (CSF) showed pleocytosis (18 cells in mm³) with domination of lymphocytes (13 cells) and elevated protein level (0.88 g/l). A CSF white blood cell count greater than 10 per mm³ or a CSF protein level greater than 50 mg per dl (0.50 g/l) indicates possible neurosyphilis.

Serologic syphilis diagnostic reactions were made, and reagin plasma response (RPR) was positive (1:128) showing recent infection. Treponema pallidum hemagglutination assay (TPHA) was positive (+). Serum enzyme-linked immunosorbent assay (ELISA) test was positive. Venereal Disease Research Laboratory (VDRL) test in CSF was positive, rate 1:16 (+). HIV test was negative. Direct examination for spirochetes was not performed because perianal syphilitic lesions were not erosive.

### Table. Findings of cerebrospinal fluid on lumbar puncture

<table>
<thead>
<tr>
<th>Variable</th>
<th>Findings</th>
<th>Normal range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Appearance of CSF</td>
<td>Clear, colorless</td>
<td>Clear, colorless</td>
</tr>
<tr>
<td>Cells (per mm³)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Red blood cells</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>White blood cells</td>
<td>18</td>
<td>&lt;5</td>
</tr>
<tr>
<td>Differential count</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lymphocytes</td>
<td>13</td>
<td>0–5 (70% of WBC)</td>
</tr>
<tr>
<td>Granulocytes</td>
<td>5</td>
<td>0</td>
</tr>
<tr>
<td>Monocytes</td>
<td>0</td>
<td>0–5 (30% of WBC)</td>
</tr>
<tr>
<td>Glucose (mmol/l)</td>
<td>2.9</td>
<td>60–70% of plasma level</td>
</tr>
<tr>
<td>Total protein (g/l)</td>
<td>0.88</td>
<td>0.15–0.4</td>
</tr>
</tbody>
</table>

CSF – cerebrospinal fluid; WBC – white blood cells.
A magnetic resonance imaging (MRI) examination of the cervical and thoracic spine (Fig. 1–5) showed a T2 hyperintense signal lesion intramedullary in the ventral part of the spinal cord on the level of T6–7 vertebra, accumulating contrast material in T1 regime. This is a lesion typical of myelitis.

Neuro-ophthalmological testing revealed no edema in the retina and hyperemic veins. Argyll Robertson symptom was not observed. Otoneurological examination and audiogram showed no abnormalities.

Adequate treatment was started. The recommendations of Centers for Disease Control and Prevention for the treatment of neurosyphilis include 18 to 24 million IU of intravenous penicillin each day for 10 to 14 days (5). Treatment for neurosyphilis approved by Lithuanian Ministry of Health is: benzylpenicillin sodium 12–24 million IU/day i/v (2–4 million VV every 4 hours for 10–21 days) or procaine benzylpenicillin 1.2–2.4 million IU i/m once a day for 14–21 days. Patient received treatment with intravenous penicillin 4 million IU five times a day, 16 days together with prednisolone starting at 30 mg, reducing the dose and discontinuing administration of the medication in order to prevent occurrence of Jarisch–Herrxheimer reaction. This reaction is an inflammatory response to the destruction of treponemes (6). Symptoms of Jarisch–Herrxheimer reaction are myalgia, headache, chills, tachycardia, increased respiratory rate, and malaise. Jarisch–Herrxheimer reaction occurs in 50 to 75% of people with early syphilis who receive treatment with antibiotics (7). Despite prednisolone administration, the patient complained of malaise, myalgia, and headache. Patient’s temperature went up (37.1°C) after the first injection of penicillin. Treatment showed positive effect, and patient could walk short distances with some nursing personnel help.
Urinary dysfunction remained. Condylomata lata in anal region diminished. The patient continued treatment in another hospital where he got treatment for another four days (4 million IU five times a day in all 20 days). RPR level diminished to 1:64 after treatment. The patient did not show up for repeated control after treatment.

**Discussion**

Neurosyphilis is a rather rare disease in our practice, especially manifesting as spinal myelitis (8). We found only a few cases on the Internet described. Asymptomatic syphilis occurs in 8 to 40% of infected patients (9, 10). After evaluating clinical symptoms (papular lesions in the anus region, a scar on the penis surface, which could possibly be after chancre, and neurological symptoms), positive serological tests and positive VDRL in the cerebrospinal fluid, lesions found in the MRI of the spinal cord, diagnosis of early secondary syphilis complicated with neurosyphilis was made. Because epidemiological history was not clear (patient’s girlfriend died one and a half year ago), we could not be sure about the duration of the disease. But specific skin lesions and our hypothesis that the patient had sexual contact 1.5 years ago let us think about early syphilis, because the duration of the disease is less than 2 years. Favorable effect of the treatment also supports the stage of secondary syphilis. As we can see in this case, neurosyphilis can cause some diagnostic, differential diagnostic and therapeutic difficulties. Spinal neurosyphilis has to be distinguished from other causes of myelitis, e.g. spinal tumor, intervertebral disc herniation, abscess, MS, HIV induced myelopathy and others (9, 11).

The prevalence of syphilis in Lithuania decreased over the last years (4), but there are still many things to do to make it lower. Many centers of sexually transmitted diseases work anonymously, and when new cases of syphilis are diagnosed, contact persons are not usually determined. Under such circumstances more cases of undertreated and undiagnosed diseases appear (12). Cases like this as well as all unclear cases should be investigated in order to exclude neurosyphilis; serological tests should be done. All patients with secondary syphilis or syphilis of more than one year should be evaluated for neurosyphilis. Patients should be monitored closely for signs and symptoms of recurrence.

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Nugaros smegenų mielitu pasireiškės neurosifilis

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Raktažodžiai: mielitas, neurosifilis, magnetinio rezonanso tomografija.


References