CLINICAL VIGNETTE

The Great Pretender

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Case

An 83-year-old male presented to neurology for left lower extremity weakness. Past medical history included osteo-arthritis and bilateral hernia surgery over forty years ago. He reported that he started having mild weakness in both legs shortly after the bilateral hernia repair. The weakness worsened in the left lower extremity over the last twelve months and is now accompanied by pain. He also notes atrophy and occasional numbness and tingling of the left leg.

The neurologist performed a detailed neurological exam. The patient was alert and oriented to person, place, time, and situation and had occasional difficulty following complex commands. Strength was 5 out of 5 in both upper extremities and the right lower extremity. The patient's effort when testing strength of the left lower extremity was limited due to pain. Strength was 2 out of 5 on left hip flexion and 4+ out of 5 on left knee flexion and knee extension. Dorsiflexion and plantar flexion of the left ankle was 5 out of 5. There was decreased vibration sensation in both lower extremities, but light touch, pinprick, and temperature senses were intact throughout. Reflexes were brisk but symmetric throughout and toes were upgoing bilaterally. Romberg, gait, and heel-to-shin tests were not performed due to left hip pain. He ambulated with a front-wheel walker.

Electromyography (EMG) showed distal axonal sensorimotor polyneuropathy affecting the lower extremities greater than the upper extremities. Magnetic resonance imaging (MRI) of the thoracic and lumbar spine showed spinal stenosis, most notably at the lumbar level, but corroborating findings were not seen on the EMG. Blood tests, including hemoglobin A1c, serum protein electrophoresis pattern (SPEP) with immunofixation electrophoresis (IFE), B12, folate, rapid plasma reagin (RPR), thyroid-stimulating hormone (TSH), and hepatitis C antibody were obtained. The only notable lab result was a positive RPR. The neurologist attributed the patient's symptoms of left lower extremity weakness to degenerative hip disease. The patient subsequently saw an orthopedic surgeon and underwent a left hip arthroplasty.

Thirteen months after the neurology consult, the patient established care with a new primary care physician (PCP). He reported ongoing balance problems. The new PCP noted his past positive RPR and ordered a *Treponema pallidum* particle agglutination (TPPA) test. The TPPA returned positive. When advised of the test results, he reported that he engaged in high risk sexual behavior when he was younger but has not had any sexual activity recently. His diagnosis was reported to the local county of public health and a referral was placed for him to see Infectious Disease for further evaluation and treatment options. The patient initially declined the referral as he did not think treatment was indicated.

Four weeks later the patient agreed to Infectious Disease consultation. Additional history showed that he never had sexual intercourse without a condom and had sex with both men and women. He had unprotected oral sex. His last sexual exposure was in his 40s. He used alcohol in the past but did not have any intravenous drug use or tobacco use. He has tried marijuana. Physical exam showed decreased left hip flexion, decreased vibratory sensation, hyperreflexia of the left patella and upgoing Babinski reflex. Infectious Disease physician felt the patient's presentation was concerning for tabes dorsalis and recommended he undergo a lumbar puncture to establish the diagnosis and be treated for syphilis regardless of the result. The patient declined both the lumbar puncture and treatment.

Two weeks after the Infectious Disease consult, the patient was hospitalized for syncope. He was ruled out for acute coronary syndrome. There was concern that syphilis may have contributed to his presenting symptoms. Both Ethics and Psychiatry were consulted during the hospitalization regarding his capacity as he continued to refuse both the lumbar puncture and treatment for presumed syphilis. He was found to have capacity as he did not have waxing and waning mental status and he was able to understand the implications for not receiving treatment.

Two months after his hospitalization for syncope, the patient fell and was admitted to an outside hospital for intracranial hemorrhage and pneumonia. Upon hospital discharge he went to a long-term acute care hospital for three weeks before he was transferred to a skilled nursing facility (SNF). While he was at the SNF, his friend contacted the patient's PCP to update her on recent events and informed her that he had been deemed incompetent. However, this friend reported to the PCP that he did not agree with this assessment as the patient seemed his usual "quirky" and "strange" self.

Two months later, approximately two years since his initial presentation to the neurology, the patient spoke with his PCP by phone. Per his PCP, the patient was unable to have a coherent conversation. He displayed paranoia and delusions in his speech akin to the "rantings of a mad man."

Discussion

Syphilis is a sexually transmitted infection caused by *Treponema pallidum*. The rates of infection have been increasing in recent years.[1] Syphilis is known as the "Great Pretender" as it can mimic many different diseases. There are four stages of the disease: primary, secondary, latent and tertiary. Neurosyphilis and ocular syphilis can occur at any time.[1-4]

There are five types of neurosyphilis: asymptomatic, meningeal/acute, meningovascular, general paresis, and tabes dorsalis. Early in the disease, patients maybe asymptomatic. For those who are symptomatic early in the disease course, the symptoms consist of syphilitic meningitis, ocular syphilis, and/or otosyphilis. Rarely, vascular complications can arise from syphilitic meningitis and lead to an ischemic stroke as syphilitic endarteritis can cause infarction in the brain. Late disease symptoms manifest as general paresis and tabes dorsalis. Both of these late presentations of neurosyphilis are now relatively rare and usually occur at least 15 years after initial infection. In general paresis, syphilis infects the meninges and cortex to cause personality changes, paranoia, and emotional lability. It can eventually lead to memory loss and dementia. In tabes dorsalis, the posterior columns of the spinal cords are affected, which manifests in abnormal gait, paresthesias, lightning pain of extremities, loss of proprioception on exam, and a positive Romberg test. Tabes dorsalis often co-exists with general paresthesias. Argyll-Robertson pupils, which are bilateral small pupils that do not constrict when exposed to light but accommodate, with convergence may be seen with both tabes dorsalis and general paresis.

Most patients with neurosyphilis have cerebrospinal fluid with lymphocytes and elevated protein. While it can be helpful to obtain a lumbar puncture in those who have neurologic symptoms, treatment is often recommended in those with symptoms regardless of this test. A lumbar puncture is not indicated in those with a diagnosis of syphilis who do not have neurologic symptoms. There are alternative treatment options for early syphilis, but the treatment of neurosyphilis remains benzathine penicillin.[5] Those with penicillin allergies should be desensitized.

The patient in our clinical vignette likely had neurosyphilis with initial exposure over 40 years ago. Lack of treatment led to the patient developing symptoms seen in both tabes dorsalis and general paresis. Patients with neurological symptoms and positive syphilis serologies should be treated as the sequelae of not treating can be detrimental.

REFERENCES

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