



Case report

TABES DORSALIS: IS THE DIAGNOSIS A CHALLENGE NOWADAYS?

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ABSTRACT

Purpose: To present a rare case of neurosyphilis, the tertiary form of syphilitic infection, following adequate but belated treatment and manifesting with a variety of clinical signs and further progression to tabetic neurosyphilis. Known as the “great imitator”, neurosyphilis is often misdiagnosed and underestimated.

Materials/methods: A 48-year-old male patient was admitted to the Neurology clinic of UMHAT “Dr Georgi Stranski” Pleven, Bulgaria, with progressive neurological deficit including numbness in the lower limbs, imbalance and wide-based gait. The T2W- MRI showed abnormally high longitudinal signals at Th12-L1 level, consistent with transverse myelitis. CSF analysis revealed leucocytic pleocytosis, elevated proteins and normal glucose. The nerve conduction studies confirmed sensory motor polyneuropathy.

Results: After treatment with intravenous aqueous crystalline (IV) penicillin, 2 million intravenously every 4 hours for 20 days, the sensory symptoms reduced. The coordination disturbances persisted, but the gait showed moderate improvement.

Conclusions: The diagnosis of syphilitic myelitis, known as tabes dorsalis, is always complex, not only because of its rarity but because of its unique clinical presentation with multiple clinical symptoms, mimicking other more common neurological disorders. Though confirmation of the diagnosis needs a lot of laboratory and neuroimaging studies, the option of effective specific treatment is worth all the efforts.

Keywords: neurosyphilis, tabes dorsalis, syphilis, sensory ataxia,

INTRODUCTION

Syphilis is a sexually transmitted infection caused by a spirochete, *Treponema pallidum*. Neurosyphilis, the tertiary form of syphilitic infection occurring most often due to inadequate treatment or lack thereof, can manifest either with early symptoms or with further progression as late neurosyphilis. Tabes dorsalis, also called tabetic neurosyphilis, affects the posterior columns of the spinal cord and the dorsal roots. Its clinical presentation includes sensory ataxia, neuropathic pain and less commonly, paresthesia and gastrointestinal disturbances [1].

In the 21st century, with the availability of various antimicrobials and prompt diagnosis and treatment, the progression of syphilis to its secondary and tertiary forms is rarely seen. Since the introduction of penicillin during the 1940s, the incidence of neurosyphilis has declined profoundly. However, with the increasing number of HIV cases, there has been a surge in the cases of neurosyphilis. Worldwide, approximately 30% of the total syphilis cases have a complication with late neurosyphilis, with one-third suffering from tabes dorsalis [2]. Due to the wide spread use of antibiotics, its typical forms have been replaced by atypical ones, making the diagnosis challenging [3, 4]. The incidence of neurosyphilis has been ranging between 0.47 and 2.1 cases per 100,000 [5]. In the early stages of the infection, frequent manifestations include asymptomatic or symptomatic meningitis, gumma and meningovascular syphilis. Late symptomatic neurosyphilis (dementia paralytica and tabes dorsalis) occurs in 10 to 20% of all untreated cases, developing decades after the primary infection [6]. In the modern era, late symptomatic neurosyphilis has been significantly less reported in developed countries, most often observed in untreated patients or in patients with HIV coinfection [7].

The aim of this case report is to raise awareness of this currently rare but very difficult to be clinically recognized condition, especially in a patient who has undergone specific antibiotic treatment.

MATERIALS and METHODS

A 48-year-old male patient was admitted to the Neurology Department at UMHAT “Dr Georgi Stranski” in Pleven, Bulgaria, in May 2023 with increasing weakness and numbness in the lower limbs and impaired balance. The symptoms initially occurred in 2019 with numbness localized in the feet and the palms and gait distur-

bances worsening in the dark and with eyes closed. The patient reported numbness in his chin and left arm and twitching of the calf muscles in the past several months. The patient is heterosexual. In 2021, a diagnosis of secondary syphilis was obtained after the appearance of a skin rash and generalized lymphadenopathy, for which he received Ceftriaxone sodium intravenously for ten days with a recommendation for serological follow-up every three months. Family history revealed a mother with diabetes and diabetic polyneuropathy. At admission, the patient presented with normal vital signs. Neurological examination revealed flaccid paralysis in the lower limbs, positive Romberg's sign, abnormal gait with staggering, and absent patellar and Achilles reflexes. Fasciculations were observed in the lower limbs. Sensory testing uncovered disturbances in the proprioception and the vibration sensation, as well as distal hypoesthesia with hyperpathia. The patient appeared to be apyretic.

Laboratory, serological and immunological tests were performed. Examination of the cerebrospinal fluid (CSF) was also obtained. Magnetic resonance imaging (MRI) of the spine and the head were carried out. The patient was consulted by an ophthalmologist and a dermatovenerologist.

RESULTS

Hemogram, renal function and electrolytes were normal. Thyroid function tests and levels of vitamin B12 were within normal ranges. Serological testing for Lyme diseases and Rapid HIV were negative. The immunological test of aquaporin 4 (AQP4)-IgG was also negative. Serum Venereal Disease Research Laboratory (VDRL) test was weakly positive, and the Treponema Pallidum Haem Agglutination (TPHA) score was 4+. CSF analysis showed leukocytic infiltration and elevated proteins with normal glucose. The CSF electrophoresis demonstrated an increased alpha1-alpha2 ratio, a marked increase in the beta-immunoglobulins and a slightly diffuse increase in the fraction of alpha1 globulin. A contrast-enhanced MRI of the lumbar region showed abnormally high signals longitudinally in T2W- at the level Th12-L1 along the anterior and posterior spinal cord columns, which is consistent with transverse myelitis at this level. [Fig. 1] The electromyography (EMG) for motor neurography showed a reduced amplitude (A) of CMAP (Compound Muscle Action Potential) for peroneal nerve bilaterally with borderline conduction velocities (CV) and expected latencies. Sensory neurography revealed a slight decrease in the conduction velocity (CV) for the left sural nerve, but it was otherwise normal. The EMG findings were consistent with that of an inflammatory polyradiculoneuropathy. After a consultation with a dermatovenerologist, treatment with Benzyl penicillin 4 x 2 MIU IV/day for 15-20 days was initiated. In addition, the patient was also administered corticosteroids, non-steroid anti-inflammatory drugs (NSAIDs), pentoxifylline and vitamin B, which resulted in substantial clinical improvement. No complications ensued.

Fig. 1. MRI of the lumbar region with abnormally high signals longitudinally in T2W- at the level Th12-L1 along the anterior and posterior spinal cord columns, consistent with transverse myelitis.



At this point, based on the patient history, clinical course and the wide diagnostic work-up, the diagnosis was consistent with neurosyphilis - tabes dorsalis. At discharge, it was recommended that the patient continued treatment at home with Penicillin 4 x 2MIU/day for the next ten days, regular serological testing every month and follow-up by a dermatovenerologist.

DISCUSSION

Historically, neurosyphilis was common in the pre-antibiotic era, occurring in 25 to 35 percent of all syphilis patients. Currently, it is an exotic diagnosis, mostly seen in the early stages of the sexually transmitted infection, predominantly in the risk group of HIV co-infected patients or in low-developed countries [2, 8]. In routine clinical practice, neurosyphilis should be suspected and ruled out in elderly or immunocompromised patients even without a prior history of treponemal infection, presenting with sensory ataxia and involvement of the spine on neuroimaging studies.

As previously mentioned, tabes dorsalis is a rarity in the modern world due to the accessibility of antibiotics in cases of timely diagnosis. It has been observed mainly in cases where the patient did not receive treatment, or it was inadequate, leading to the progression of the disease and, over 15-20 years, morphing into neurosyphilis. Broadly, neurosyphilis can manifest as early or late forms. The former mainly affects mesodermal structures such as the meninges and vessels, while the latter affects the parenchyma of the brain and spinal cord [6]. One of the earliest manifestations of neurosyphilis are neuropsychiatric symptoms such as mild personality changes, amnesia, later evolving into hallucinations, delusions, delirium or aggression. These symptoms improve upon the addition of antipsychotics to the antibiotic regimen [4]. Asymptomatic cases are the most common ones in neurosyphilis, and thus, proper screening at the primary

stage may facilitate timely diagnosis and prevent progression [9]. The differential diagnosis for neurosyphilis includes a variety of disorders, such as spinal tumors, vit. B12 insufficiency, transverse myelitis, longitudinally extensive transverse myelitis (LETM), as well as neuromyelitis optic spectrum disorders [9-12]. This warrants the need for an extensive diagnostic panel.

This report presents the unconventional clinical case of a patient who progressed to late-stage syphilis despite having received the required dose and duration of treatment and regular follow-up serology tests. It highlights the importance of proper screening tools and regular follow-ups to arrest the disease progression early on, thus preventing any loss in the patient's quality of life.

All other infectious, autoimmune and paraneoplastic diagnostic hypotheses were excluded through imaging results and extensive CSF and serum analysis.

CONCLUSION

Highlighting the features of rare conditions such as tabes dorsalis will contribute to a better understanding of the disease and its clinical course in the modern world. Although less common, late forms of neurosyphilis must always be searched for in cases where there is the slightest suspicion because early recognition and effective treatment are crucial for better clinical outcome of the patients.

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Please cite this article as: Danovska M. Tabes dorsalis: is the diagnosis a challenge nowadays? *J of IMAB*. 2024 Jul-Sep;30(3):5642-5644. [Crossref - <https://doi.org/10.5272/jimab.2024303.5642>]

Received: 05/03/2024; Published online: 15/07/2024



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