

Case Reports

Orofacial Involuntary Movements in Neurosyphilis: Beyond the Candy Sign

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Abstract

Background: Involvement of the central nervous system in patients with syphilis (neurosyphilis) may result in several neuropsychiatric symptoms. Rarely, patients with neurosyphilis may develop movement disorders with different phenomenology. Subtle orofacial dyskinesias have been reported in patients with neurosyphilis, known as the candy sign.

Case Report: We describe a patient with neurosyphilis who presented with severe orofacial involuntary movements.

Discussion: Our patient had orofacial movements at presentation and severity of the movements was much higher than the candy sign that has been reported in patients with neurosyphilis. This report contributes towards the ever-expanding clinical spectrum of neurosyphilis.

Keywords: Syphilis, neurosyphilis, dyskinesia, movement disorders

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Introduction

Syphilis results from infection by *Treponema pallidum* in which multiple organ systems including the central nervous system (CNS) may be affected. There is a great deal of variability in the clinical presentations of syphilis, as a result of which it has also been known as “the great mimicker”.¹ Neurosyphilis seems to occur with higher frequency in individuals co-infected with the human immunodeficiency virus (HIV). The clinical spectrum of neurosyphilis is wide and it may vary with the structures involved in the pathogenesis of the disease (meningeal or vascular or parenchymal).² Subsequently, the longstanding untreated infections may involve the brain parenchyma that may present as general paresis, tabes dorsalis, and the typical ocular finding, the Argyll–Roberston pupil.^{1,3} Rarely, patients with syphilis may present with several movement disorders such as parkinsonism, ataxia, myoclonus, chorea, and dystonia.^{1,4} In addition, the presence of subtle orofacial dyskinesia, which is termed the “candy sign” has been reported in a few cases.^{5,6}

We hereby describe a case of neurosyphilis whose clinical picture was dominated by orofacial involuntary movements and these

movements were much more severe than the previously reported subtle dyskinesias (candy sign).

Case report

A 32-year-old male, a driver by occupation, initially presented to the psychiatry emergency services and he was subsequently referred to the neurology emergency services. The patient was accompanied by his wife, who gave the history of involuntary movements of the face and mouth for 1 month along with abnormal vocalizations. There were no specific aggravating and alleviating factors. Past history revealed the presence of fever and altered sensorium 4 years ago, which had lasted for approximately 2 weeks, and he had one episode of generalized tonic–clonic seizure during that time. Additionally, there were significant behavioral changes characterized by anger outbursts, irritability, and apathy since the last 4–5 years. There was no history of tremor, jerks, imbalance while walking, and abnormal posturing of any other body parts. There was no clear history of any intravenous drug abuse, toxin exposure, and head injury. None of his family members had history of tremor, parkinsonism, memory disturbance, or psychiatric disorders.

On the day of clinical evaluation, he was conscious, and alert, but was not completely cooperative for the physical examination. There were intermittent abnormal vocalizations. There was no neck rigidity suggestive of meningitis. All the cranial nerves were normal on examination, but he had absent light and accommodation reflex. Positive findings during the motor system examination were dominated by involuntary movements in the form of frequent opening and closing of the mouth, continuous perioral movement (chewing movements), and occasional protrusion of the tongue. He also had occasional ballistic hand movements. Rigidity and bradykinesia were minimal in both upper and lower limbs. All the deep tendon reflexes were well elicited and the plantars had flexor response in both sides. We did not find any cerebellar and sensory deficits and his gait was grossly normal (Video 1).

Routine hemogram and biochemical parameters were within normal limits. Considering the fact that subacute onset involuntary movements could be secondary to structural lesions, magnetic resonance imaging (MRI) of the brain was done, which revealed significant bilateral temporal lobe atrophy and signal changes in the thalamus and basal ganglia (Figure 1). The possibility of chronic neuroinfection was considered as the patient had a history of altered sensorium 5 years previously, and there had been behavioral changes over 4–5 years. Infections with human immunodeficiency virus (HIV), herpes simplex virus (HSV), and syphilis were considered. Subsequent examinations revealed that HIV was negative and the cerebrospinal fluid (CSF) profile did not reveal any convincing evidence of bacterial or viral meningitis (cell count, 7 polymorphonuclear leucocytes/mL; protein, 56 mg/dL, and glucose, 84 mg/dL). After ruling out these two common infections (HIV and HSV), which may present with neuropsychiatric symptoms, we checked the serum for Venereal Disease Research Laboratory (VDRL) and Treponema Pallidum Hemagglutination (TPHA); they were both positive in this case. CSF was non-reactive for VDRL as it may happen in ~50% of patients with neurosyphilis (~50%).⁷ Estimation of immunoglobulin G in CSF and a test for rapid plasma reagin could not be done, as these tests are not available in our center. A diagnosis of neurosyphilis was established in this case for the following

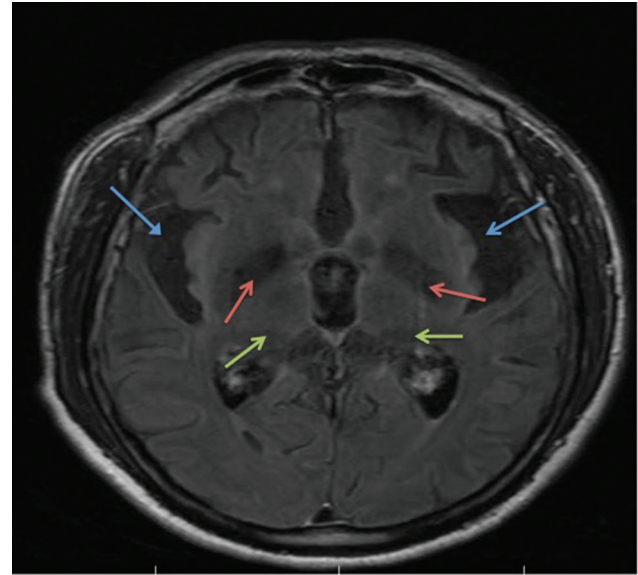


Figure 1. Axial T2 Fluid-Attenuated Inversion Recovery (FLAIR) Magnetic Resonance Sequence of the Patient. The image shows bilateral temporal lobe atrophy (blue arrows), and hypointensity in the globus pallidus (red arrows) and thalamus (green arrows).

reasons: (1) subacute onset neuropsychiatric symptoms, (2) MRI changes that are commonly observed in neurosyphilis, (3) CSF pleocytosis, and a mild rise in CSF protein, and (4) serum reactive to both VDRL and TPHA. The diagnosis gained further support from the fact that the patient had notable improvement in both motor and behavioral symptoms after 6 weeks of treatment with penicillin.

Discussion

As mentioned above, patients with neurosyphilis may present with a wide range of neuropsychiatric symptoms. Rarely, involuntary movements may dominate the clinical presentations. Movement disorders in neurosyphilis usually represent meningovascular syphilis, and involuntary movements are the result of the ischemic lesions secondary to vascular involvement.⁴ The spectrum of movement disorders in neurosyphilis is large and it may encompass parkinsonism, myoclonus, dystonia, and ataxia. We are aware of two case reports describing subtle orofacial dyskinesia in neurosyphilis, which was later called the candy sign.^{5,6} However, in our case the magnitude of the orofacial movements was more than what has been referred to as the candy sign. We observed frequent involuntary opening and closing of the mouth along with abnormal vocalizations. In addition, our patient had occasional ballistic hand movements. To the best of our knowledge, orofacial involuntary movements of such high magnitude have never been reported in patients with neurosyphilis and our case certainly contributes towards the ever-expanding clinical spectrum of this complex disease. The learning points from this case include (1) subacute onset involuntary movements in a background of behavioral abnormalities may indicate a chronic neuroinfection such as neurosyphilis and (2) orofacial involuntary movements much more severe than the candy sign may be presenting symptoms.



Video 1. Patient with Abnormal Vocalizations along with Continuous Orofacial Involuntary Movements. Continuous orofacial involuntary movements appear in the form of frequent opening and closing of the mouth and chewing movements.

References

1. Drago F, Merlo G, Ciccarese G, Agnoletti AF, Cozzani E, Rebori A, et al. Changes in neurosyphilis presentation: a survey on 286 patients. *J Eur Acad Dermatol Venerol* 2016;30:1886–1900. doi: 10.1111/jdv.13753
2. Nagappa M, Sinha S, Taly AB, Rao SL, Nagarathna S, Bindu PS, et al. Neurosyphilis: MRI features and their phenotypic correlation in a cohort of 35 patients from a tertiary care university hospital. *Neuroradiology* 2013;55:379–388. doi: 10.1007/s00234-012-1017-9
3. Timmermans M, Carr J. Neurosyphilis in the modern era. *J Neurol Neurosurg Psychiatry* 2004;75:1727–1730. doi: 10.1136/jnnp.2004.031922
4. Shah BB, Lang AE. Acquired neurosyphilis presenting as movement disorders. *Mov Disord* 2012;27:690–695. doi: 10.1002/mds.24950
5. Marto JP, Borbinha C, Lampreia T, Alves L, Viana-Baptista M. Teaching video neuroimages: candy sign: the clue to the diagnosis of neurosyphilis. *Neurology* 2017;88:e35. doi: 10.1212/WNL.0000000000003544
6. Martinelli P, Rizzo G, Scaglione C, Capellari S. Neurosyphilis orofacial dyskinesia: the candy sign. *Mov Disord* 2013;28:246–247. doi: 10.1002/mds.25281
7. Harding AS, Ghanem KG. The performance of cerebrospinal fluid treponemal-specific antibody tests in neurosyphilis. *Sex Transm Dis* 2012;39:291–297. doi: 10.1097/OLQ.0b013e31824c0e62