

# Chorea as the presenting feature of neurosyphilis

Serkan Ozben, Canan Erol, Feriha Ozer<sup>1</sup>, Raziye Tiras

Department of Neurology, and <sup>1</sup>Movement Disorders Outpatient Clinic, Haseki Educational and Research Hospital, Istanbul, Turkey

## Address for correspondence:

Dr. F. Feriha Ozer  
Atakoy 9, Kisim A14-B D:138,  
Istanbul/TURKEY.  
E-mail: ffozer@yahoo.com

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## Abstract

Syphilis is still a significant public health problem in developing countries. Although chorea is a very rare manifestation of neurosyphilis, it might be on occasions the initial symptom. This report presents a patient with neurosyphilis who had chorea as the initial presenting symptom.

**Key words:** Chorea, neurosyphilis, *treponema pallidum*

## Introduction

The term “chorea” originates from the Greek word “chorea” meaning dance. Chorea is defined as irregular, unpredictable, brief, jerky hyperkinetic movements that flit randomly from one body part to another.<sup>[1]</sup> Chorea may be a feature of a neurodegenerative disorder such as neuroacanthocytosis, dentatorubropallidolusian atrophy, or benign hereditary chorea and it can also be a rare manifestation of some very common diseases such as vascular diseases or hyperthyroidism.<sup>[2-4]</sup> Other causes of chorea include drugs, central nervous system infections, and antiphospholipid syndrome and correcting the underlying disease should be the first step in the treatment of chorea.<sup>[2]</sup>

Syphilis is still a significant public health problem in developing countries. In the last few years, reports suggest increased incidence of syphilis globally;<sup>[5]</sup> This may partly be due to the simultaneous increase in human immunodeficiency virus (HIV) infection. In the recent studies neurosyphilis with atypical features is being more frequently reported.<sup>[6-8]</sup> Awareness and testing for syphilis serology in an appropriate clinical setting is important. We report a patient with neurosyphilis who had chorea as the initial presenting feature.

## Case Report

A 70-year-old male presented to our outpatient clinic with movement disorder of 10-year duration. The movement disorder consisted of quick, unpredictable, jumping and

jerking of the limbs. The symptoms first started in the right hand and right foot then spread to the left side over a period 3 years. These movements very much qualified to be categorized as chorea. Neurological and psychological examinations revealed no other abnormality. He had no prior history of exposure to neuroleptic drugs. Family history was negative for any movement disorder. He had history of hypertension of one year duration. He was prescribed haloperidol, initially 0.5 mg twice a day and increased to 1 mg three times a day, but there was no observable difference.

Routine biochemical and hematological tests were normal. Brain magnetic resonance imaging (MRI) revealed no abnormality. Fresh blood smear was negative for acanthocytes. Serological tests for syphilis revealed: Venereal Disease Research Laboratory (VDRL) test 1/4 reactive, positive rapid plasma reagin (RPR), and a positive *Treponema-pallidum* hemagglutinations-assay (TPHA). Cerebrospinal fluid (CSF) examination: Protein – 52 mg/dL (normal 15 – 45 mg/dL), normal glucose, and no cells. CSF VDRL was positive. Fluorescent treponemal antibody absorption (FTA-ABS) test was positive in serum (1/640) and positive in CSF, HIV serology was negative.

He was treated with one dose of benzathine penicillin 2.4 million IM and then with penicillin G 2.4 million IM per day for 14 days. His choreiform movements distinctly improved on the 12<sup>th</sup> day of therapy. Repeat serological tests for syphilis in serum revealed 1/4 positive VDRL, 1/4 positive RPR, positive TPHA, positive CSF-VDRL, and positive FTA-ABS both in serum and CSF. He

was subsequently treated with cephtriaxone 2 g per day IM for 10 days. After this therapy, only FTA-AB testing was done, it was positive in serum and negative in CSF. He was followed-up in the outpatient clinic at regular intervals. At six month follow-up he had minimal non-disabling chorea, but he was excited. He was not any medication for chorea.

## Discussion

The exact anatomical substrate of chorea is not defined. Studies in patients with hemichorea suggest lesions involving basal ganglia, mainly the subthalamic nucleus as the substrate.<sup>[9-12]</sup> Vascular lesions,<sup>[9-12]</sup> vascular malformations,<sup>[13]</sup> metastasis,<sup>[14]</sup> infective granulomas,<sup>[15-17]</sup> demyelinating lesions,<sup>[18]</sup> and infectious diseases<sup>[19]</sup> involving these anatomical structures may cause hemichorea. In our patient MRI did not reveal any structural lesion.

Syphilis is a chronic multisystem disease. An estimated 4-9% of patients with untreated syphilis develop symptomatic neurosyphilis. The three broad clinical categories of neurosyphilis are: Asymptomatic, meningovascular, and parenchymatous. Asymptomatic neurosyphilis is characterized by absence of clinical symptoms or signs and abnormal CSF including serology. Meningovascular syphilis results from inflammatory reaction of the perivascular spaces of the meninges, brain, and spinal cord. Tabes dorsalis (and general paralysis of the insane (GPI) are the parenchymatous forms of neurosyphilis.<sup>[20]</sup>

Only a few cases of neurosyphilis presenting with features of basal ganglia involvement have been documented in the recent literature and most of these patients had associated HIV infection.<sup>[21]</sup> Our patient was negative for HIV. The choreiform movement responded to specific therapy for neurosyphilis. We had to give second line drug cephtriaxone as he did not show significant response with benzathine penicillin. Failure to penicillin therapy has been reported in patients with HIV and neurosyphilis. Recent reports suggest that neurosyphilis in patients with HIV may have risk of treatment failure.<sup>[22,23]</sup>

The “gold standard” for the diagnosis of neurosyphilis is not available. CSF leukocytosis and elevated protein are nonspecific and may support the diagnosis in a given clinical setting. A reactive serum treponemal test with a reactive CSF VDRL test is generally considered definitive for neurosyphilis.<sup>[24-27]</sup>

Although neurosyphilis is a very rare cause for chorea, when other diseases are excluded, it should be suspected and the serological test should be done. In the recent years with the availability of effective antibiotic therapy neurosyphilis with more atypical presentations are being

described. Center for Disease Control and Prevention (CDC) recommends a CSF examination for all patients with syphilis infection who have neurological or ophthalmic symptoms and signs or who have active tertiary disease (aortitis, gumma, iritis) or have failed therapy.<sup>[28]</sup>

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