

A rare case of neurosyphilis presenting as generalized dystonia with bilateral cerebellar infarctions

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Keywords

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Syphilis is a significant public health problem in developing countries. In the last few years, there is a steady increase in cases of neurosyphilis, including atypical forms. Movement disorders (parkinsonism, chorea, ataxia, etc.) have been reported in some patients.^{1,2} Rare cases of focal dystonia (laryngeal, orofacial dyskinesia) have also been described.^{1,3} Generalized dystonia is an unusual and previously undescribed manifestation of neurosyphilis. Here, we present a case study of the patient with meningovascular neurosyphilis with generalized dystonia and partial successful effect after ceftriaxone treatment.

A 57-year-old man had the history of treated syphilis (unspecified) by benzathine benzylpenicillin at age of 42 (lumbar puncture was not performed). After the treatment, cervical dystonia first appeared. Botulinum toxin injections had no effect. At the age of 45 and 46, bilateral

pallidotomy and the left-sided thalamotomy were performed resulting in speech and gait worsening; in addition, dystonia appeared in the right limbs. Levodopa treatment was ineffective. Mutations in DYT1 and DYT6 genes were not found, and family history of movement disorders was negative. The patient had no significant vascular risk factors with the exception of mild treated arterial hypertension. There was no history of neuroleptics use.

On neurological examination (Figure 1, A), he had mild cognitive impairment with frontal lobe signs and paratonia. Pupillary light reflexes were reduced. Severe dysarthria with laryngeal dystonia and mild dysphagia were present. He had asymmetric and decreased deep tendon reflexes with absent Achilles reflex and bilateral Babinski sign. Right torticollis, truncal dystonia, right hand dystonia, and right striatal toe were revealed.

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He also had mild truncal and appendicular cerebellar ataxia as well as sensitive ataxia with decreased vibration sensation in the feet and positive Romberg test.

Brain magnetic resonance imaging (MRI) study (Figure 1, B-G) showed post-stroke lesions in both cerebellar hemispheres, cerebral atrophy (most prominent in the frontal and parietal lobes), periventricular white matter hyperintensities (WMHs) and bilateral postoperative putaminal and left thalamic cysts.

Extracranial ultrasound revealed left vertebral artery occlusion. Positive treponemal and nontreponemal tests were detected in serum, and human immunodeficiency virus (HIV) antibodies were negative. Cerebrospinal fluid (CSF) cell count and glucose were normal, and protein concentration was slightly elevated (573 mg/l). CSF treponemal [treponema pallidum immobilization (TPI) test, fluorescent treponemal antibody absorption (FTA-ABS) test] and nontreponemal [rapid plasma reagin (RPR)] tests were strongly positive. Patient was treated with two courses of intravenous (IV) ceftriaxone [2 g, twice a day (b.i.d.)] with speech improvement and marked torticollis reduction.

The exact cause of dystonia in our patient remains unclear; moreover, we have no

information when cerebellar infarcts (most probably due to meningovascular syphilis) occurred. There are a substantial number of case reports linking dystonia to structural lesions of the cerebellum. Some cases document the emergence of late-onset oromandibular dystonia (OMD) or blepharospasm with torticollis after bilateral cerebellar infarction or hemorrhage.^{4,5} In our patient, the combined lesions in cerebellar hemispheres, putamen, and thalamus could lead to disruption of the multisynaptic links between cerebellum, basal ganglia, and cortex, resulting in the dystonia development. Nevertheless, considering the positive effect of ceftriaxone treatment, we can assume a role of the direct treponema pallidum (*T. pallidum*) impact or small vessel syphilitic vasculitis (Nissl-Alzheimer arteritis) in the dystonia progression. This case demonstrates that early diagnosis and treatment of neurosyphilis is extremely important in patients with secondary movement disorders.

Conflict of Interests

The authors declare no conflict of interest in this study.

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None.

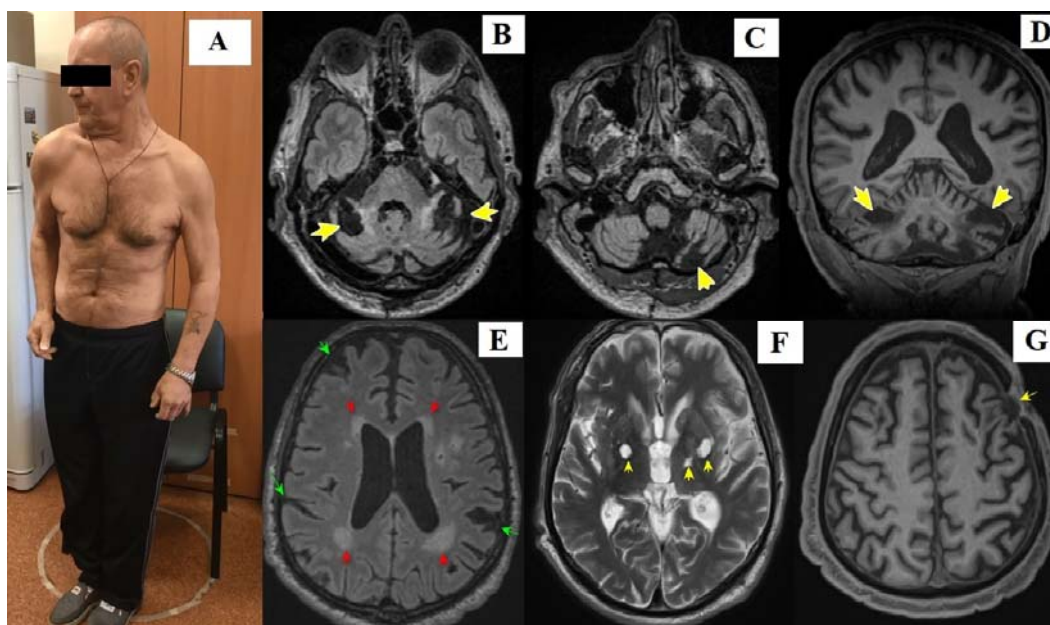


Figure 1. Clinical features (A) and brain magnetic resonance imaging (MRI) study (B-G) of the patient; B-C [T2-weighted-fluid-attenuated inversion recovery (T2-FLAIR)] and D (T1-WI): Bilateral cerebellar infarctions (yellow arrows); E (T2-FLAIR): Periventricular white matter hyperintensities (WMHs) (red arrows), frontal and parietal lobes atrophy (green arrows); F (T2-WI) and G (T1-WI): Postoperative changes including putaminal and thalamic cysts (yellow arrows)

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