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Case Report

# Adult-Onset Sydenham Chorea Mimicking a Functional Movement Disorder: A Case Report

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## Abstract

**Background:** Sydenham chorea is a post-streptococcal autoimmune movement disorder and one of the major manifestations of acute rheumatic fever. Adult-onset disease is rare and may be diagnostically challenging because positive streptococcal serology is supportive but not disease-specific.

**Case Presentation:** We report a 38-year-old man with progressive predominantly unilateral choreiform movements of the right shoulder and proximal arm with intermittent facial involvement. MRI and EEG were repeatedly normal. CSF showed no pleocytosis or intrathecal immunoglobulin synthesis. Wilson disease, Huntington disease, autoimmune encephalitis, vascular chorea, infectious, metabolic, drug-induced and paraneoplastic causes were considered and became unlikely after extensive evaluation. Persistently elevated anti-DNase B titres and elevated ASO titres supported antecedent Group A streptococcal exposure. Mild Jack-in-the-box tongue was present, while Milkmaid's grip was absent. Partial distractibility initially raised concern for functional movement disorder.

**Conclusion:** Adult-onset Sydenham chorea should neither be diagnosed solely because ASO or anti-DNase B titres are elevated nor dismissed because the patient is an adult or shows partial distractibility. Diagnosis requires synthesis of movement phenomenology, supportive streptococcal serology and systematic exclusion of competing disorders.

***Keywords:*** *Sydenham chorea; adult-onset chorea; autoimmune chorea; Group A streptococcus; functional movement disorder; anti-DNase B; ASO*

## Introduction

Sydenham chorea is an autoimmune movement disorder classically associated with acute rheumatic fever and remains one of the five major Jones criteria [1]. Acute rheumatic fever is a delayed immune complication of Group A β-haemolytic streptococcal infection and remains clinically relevant worldwide despite declining incidence in many high-income countries [2]. Sydenham chorea is usually a disorder of childhood and adolescence [3,4]. Adult-onset Sydenham chorea is therefore uncommon and has mainly been described in case reports or small series [30-36].

The pathophysiology is thought to involve molecular mimicry between streptococcal antigens and neuronal targets in basal ganglia circuits [6-9]. Patient-derived antibodies have been shown to alter neuronal signalling, including CaMKII activation and dopaminergic pathway effects [6,7]. This mechanism helps explain why MRI and CSF may be normal despite clinically relevant chorea [38,39].

Treatment evidence remains limited, and management is usually based on symptomatic therapy, immunomodulation in selected cases, and prevention of recurrent streptococcal infection [12-14, 41,42]. Diagnostic difficulty is especially high in adults because the differential diagnosis includes functional movement disorder, Wilson disease, Huntington disease, autoimmune encephalitis, vascular chorea, drug-induced chorea and other acquired choreas [15-29]. We present a diagnostically challenging adult case in which unilateral chorea, partial distractibility and persistently positive streptococcal serology required careful interpretation.

## Case Presentation

A 38-year-old right-handed man presented with progressive involuntary movements predominantly affecting the right shoulder girdle and proximal right upper extremity. Symptoms had evolved over several months and interfered with daily activities. He had no known prior neurological disease, no family history of Huntington disease or hereditary movement disorder, and no exposure to dopaminergic medication, neuroleptics or recreational drugs known to induce chorea. Drug-induced chorea was considered because medication-related movement disorders are an important acquired differential diagnosis.

On neurological examination, the movements were irregular, non-rhythmic and choreiform, predominantly involving the right shoulder and proximal arm, with intermittent facial involvement. Strength was preserved throughout. There were no pyramidal signs, sensory deficits, cerebellar syndrome, cognitive impairment or gait ataxia. Milkmaid's grip was absent. Sustained tongue protrusion showed mild motor impersistence consistent with a mildly positive Jack-in-the-box tongue. The movements of the arm and shoulder partially decreased during distraction and trunk stabilization, raising concern for functional movement disorder. However, functional movement disorder requires positive clinical evidence interpreted in the full neurological context, and distractibility alone is not pathognomonic.

Brain MRI was repeatedly normal, without basal ganglia, thalamic, cortical or brainstem lesions. EEG was repeatedly normal and showed no epileptiform activity. CSF analysis showed 1 leukocyte/µL, no oligoclonal bands, negative Borrelia antibody index and no evidence of intrathecal inflammation; protein was mildly elevated at 0.687 g/L, compatible with mild blood-CSF barrier dysfunction rather than inflammatory CNS disease. CSF and serum immune studies in Sydenham chorea may be unrevealing, supporting the concept of immune-mediated functional basal ganglia dysfunction rather than structural inflammation.

Wilson disease was considered as a cause of movement disorders in younger adults. Serum ceruloplasmin and copper were normal, and there was no hepatic syndrome or typical neurological phenotype. Huntington disease was considered because it is the most common hereditary adult chorea, but the patient had no family history, no cognitive or psychiatric decline, no caudate atrophy and no progressive generalized syndrome. Autoimmune encephalitis and autoimmune chorea were considered; however, there were no seizures, encephalopathy, psychiatric syndrome, CSF inflammation or MRI changes suggesting autoimmune encephalitis. Vascular chorea was unlikely because onset was progressive rather than acute and MRI showed no signs of infarction or hemorrhage.

Streptococcal serology showed persistent evidence of antecedent Group A streptococcal exposure. Anti-DNase B was elevated on two occasions over a three-month interval, measuring 496 U/mL and later 476 U/mL. ASO titres were also elevated, with values including 272 IU/mL and later 222 IU/mL. These findings were interpreted as supportive but not diagnostic, because ASO and anti-DNase B indicate previous exposure rather than prove Sydenham chorea.

During the second admission, the movement disorder remained clinically coherent: predominantly unilateral chorea with intermittent facial involvement, preserved strength and mild motor impersistence. No alternative structural, infectious, autoimmune, hereditary, metabolic or toxic cause became apparent. Adult-onset Sydenham chorea was therefore considered the most coherent diagnosis. Symptomatic treatment with tiapride was initiated.

## Discussion

This case illustrates several diagnostic pitfalls in adult-onset Sydenham chorea. First, adult age should not exclude the diagnosis. Although Sydenham chorea is typically paediatric, adult cases have been reported, including presentations with psychiatric symptoms, reversible striatal hypermetabolism, diagnostic confusion with drug-induced movement disorder, recurrence in adulthood and very late-onset treatable disease [30-36]. Pregnancy-associated recurrence and cognitive sequelae in adult patients have also been described [36,37].

Second, positive streptococcal serology must be interpreted cautiously. ASO and anti-DNase B titres support antecedent Group A streptococcal infection but do not prove autoimmune basal ganglia disease [1,3,4].

In the present patient, serology became meaningful only because the movement phenomenology was compatible with chorea and competing diagnoses had been systematically assessed. This distinction is essential to avoid overdiagnosis in patients with unrelated functional or somatoform movement disorders.

Third, normal MRI and CSF do not exclude Sydenham chorea [38,39]. The proposed pathophysiology involves antibody-mediated dysfunction of basal ganglia networks rather than destructive structural disease [6,9]. Functional imaging abnormalities, including reversible striatal hypermetabolism, have been reported in adult-onset disease even when conventional imaging is unrevealing [31].

Although antibodies against dopamine D2 receptors (and, in research settings, D1 receptors) have been described in patients with Sydenham chorea and may provide insights into disease pathophysiology, their diagnostic utility remains insufficiently established. Consequently, these assays are currently not part of routine clinical diagnostics, are available only in a limited number of specialized research laboratories or university centers, and neither a positive nor a negative serum result is considered sufficient to confirm or exclude the diagnosis [43-45].

Fourth, partial distractibility should not automatically lead to a functional diagnosis. Functional movement disorder is diagnosed by positive signs of inconsistency and incongruence, not merely by normal investigations [15,16]. Organic and functional disorders may also coexist or overlap diagnostically [17]. In our patient, partial distractibility was outweighed by repeated observation of coherent choreiform phenomenology, intermittent facial involvement, motor impersistence and supportive serology.

The differential diagnosis was broad. Wilson disease was important because of age and treatability but became unlikely after normal copper metabolism studies and absent typical clinical features [24]. Huntington disease was unlikely because there was no family history, cognitive decline, psychiatric syndrome or caudate atrophy [25-27]. Autoimmune chorea and autoimmune encephalitis were less likely because there was no encephalopathy, seizures, CSF inflammation or supportive MRI pattern [19,23]. Vascular chorea was unlikely because there was no acute onset or MRI lesion [28,29].

Therapeutic evidence in Sydenham chorea is limited but supports symptomatic therapy in functionally relevant chorea and immunomodulation in selected severe cases [12,14]. Corticosteroid treatment has been reported to shorten disease duration in some cases [41,42]. Antibiotic prophylaxis is recommended in rheumatic fever frameworks to prevent recurrent streptococcal exposure rather than to treat active CNS infection [1,2]. The present patient was managed symptomatically with tiapride, reflecting a pragmatic approach in a clinically stable adult with persistent chorea.

The main limitation of this report is the absence of a disease-specific biomarker. The diagnosis remains probabilistic and based on clinical synthesis. Nevertheless, the combination of compatible chorea, mild motor impersistence, repeated normal MRI/CSF, exclusion of major alternative diagnoses and persistent streptococcal serology makes adult-onset Sydenham chorea the most coherent explanation.

## Conclusion

Adult-onset Sydenham chorea is rare but should remain within the differential diagnosis of adults with unexplained chorea. Elevated ASO or anti-DNase B titres should support, but never replace, clinical reasoning. Conversely, adult age, normal MRI, normal CSF or partial distractibility should not automatically exclude the diagnosis. This case emphasizes that diagnosis requires integration of movement phenomenology, supportive evidence of antecedent streptococcal infection and systematic exclusion of competing disorders.

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## Author Contributions

**Dalibor Mihajlovic:** Conceptualization, patient care, diagnostic work-up, study design, literature review, data interpretation, manuscript drafting, manuscript revision, supervision, corresponding author.

**Yazeed Bani Hamad:** Literature review, data interpretation, critical revision of the manuscript.

**Christoph Baumsteiger:** Clinical supervision, critical revision of the manuscript, intellectual contribution.

**Zaid Bani Hamad:** Literature review, data interpretation, manuscript revision, intellectual contribution.

All authors read and approved the final manuscript.

## Conflicts of Interest

The authors declare that they have no competing interests.

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## Ethics Statement

According to institutional policy, ethics committee approval was not required for publication of a single anonymized case report.

## Patient Consent

Written informed consent for publication of this case report and accompanying images was obtained from the patient.

## Data Availability

The data supporting the findings of this study are available from the corresponding author upon reasonable request.

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