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You can dance if you want to: A case of Sydenham's chorea

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ABSTRACT

Isolated motor disturbances in the paediatric population are uncommon presentations to the emergency department. Choreiform movements have a broad differential diagnosis and may present insidiously with progressive worsening of asymmetric clumsiness, hypotonia and dysarthria. The incidence of Sydenham's chorea (SC) caused by acute rheumatic fever (ARF) is very rare in developed countries. We report a previously healthy, vaccinated 9-year old male who presented to our ED with intermittent and progressive right sided clumsiness for four weeks. Physical examination findings showed dysdiadokinesis and dysmetric movements of the right side, which varied in intensity and were less pronounced on serial re-examination during the same ED visit. Basic bloodwork, MRI and MRA/V showed no abnormalities, and the patient was discharged home with urgent neurology follow-up. He re-presented to our ED four days later with worsening gait and inability to hold a pencil at school. He was subsequently diagnosed with chorea by the neurology team. The cause of chorea was later determined to be SC, and the patient's throat swab came back positive for group A-beta hemolytic strep (GAS) infection. We explore current literature regarding the various presentations of ARF, differential considerations in acute chorea, and diagnostic studies needed to determine the etiology of acute chorea. With the low incidence of chorea in developed nations, this diagnosis can be easily overlooked. We highlight the importance of this diagnosis, as well as primary and secondary treatment in ARF.

Paediatric movement disorders are generally of the hyperkinetic type and may develop due to direct injury to the basal ganglia or cerebellum, or be manifestations of systemic disease, such as acute rheumatic fever (ARF), infections, autoimmune disorders, psychogenic disorders, medication side effects, or drug toxicities [1,2]. Specifically, ARF is a multisystem, inflammatory process which can occur as a delayed manifestation of group A streptococcal tonsillopharyngitis infection. Although ARF can occur at any age, most cases occur in children aged 5–15 years [3].

A 9-year-old male presented to our paediatric emergency department (ED) with a 1-month history of progressive right-sided clumsiness and intermittent involuntary movements of his right arm and leg. The patients' parents noted speech slurring as well. Review of systems was unremarkable except for a viral upper respiratory tract infection that started one-week prior to the neurological symptoms. The boy had no previous medical history or significant family history, was not taking any medications, and had up-to-date immunizations.

On examination, the patient had normal vital signs and a mostly benign assessment. Of note, his cranial nerve exam was remarkable for intermittent dysarthric speech without facial droop. His sensory and motor exam was grossly normal. Coordination tests showed right sided finger-nose and heel-shin dysmetria, in addition to right hand dysdiadochokinesis. His gait had elements of unsteadiness with normal base and intermittent episodes of stumbling to the right.

Bloodwork showed a normal complete blood count, extended electrolytes, c-reactive protein, coagulation studies, and renal function. An urgent MRI and MRA/V was normal. The patient was discharged home with urgent neurology follow-up; however, he returned four days later with worsening symptoms.

The paediatric neurology team was consulted and they diagnosed him with chorea. During the patient's admission, thyroid studies, antinuclear and antiphospholipid antibodies, and a metabolic screen were normal. A throat culture detected Group A *Streptococcus pyogenes*, and anti-streptolysin O titers were 114 IU/mL (reference range 166–250 IU/mL). Serum ceruloplasmin was normal, and Ophthalmology's assessment of the patient was unremarkable. His electrocardiogram showed normal sinus rhythm

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Table 1
Components of the Revised Jones Criteria in low, moderate, and high risk populations.

Revised Jones Criteria in low risk populations	
Major manifestations	Minor manifestations
• Carditis and valvulitis (e.g., pancarditis) • Arthritis (polyarthritis only) • Chorea • Subcutaneous nodules • Erythema marginatum	• Polyarthralgia • Fever (≥38.5) • Elevated acute phase reactants (ESR ≥60 or CRP ≥3 mg/dL) • Prolonged PR interval on ECG
Revised Jones Criteria in moderate and high risk populations	
Major manifestations	Minor manifestations
• Carditis and valvulitis (eg, pancarditis) • Arthritis (monoarthritis or polyarthritis) • Chorea • Subcutaneous nodules • Erythema marginatum	• Monoarthralgia • Fever (≥38.5) • Elevated acute phase reactants (ESR ≥30 mm/h or CRP ≥ 3 mg/dL) • Prolonged PR interval on ECG
Evidence of recent GAS infection	
• Throat cultures growing group A beta-hemolytic strep (GABHS) • Elevated anti-streptolysin O titers (ASO > 200)	

with a normal PR interval. An echocardiogram revealed mild mitral regurgitation.

The patient had a 10-day course of penicillin for his strep throat, and was stepped down to prophylactic penicillin to be taken until age 18. In terms of symptomatic management for the chorea, valproic acid was an option offered by the neurology team, however the patients' parents elected to not initiate this treatment as his symptoms started improving.

SC is a neurological disorder which is comprised of abrupt, non-rhythmic, involuntary movements, muscular weakness, and behavioural changes [5]. It is proposed that SC is caused by antibodies against group-A-beta hemolytic strep (GAS) attacking the basal ganglia [6]. It tends to be a delayed complication of GAS infection. SC is diagnosed clinically, and is a component of the modified Jones criteria for ARF (Table 1). Even as a stand-alone component, SC is sufficient to make a diagnosis of acute rheumatic fever in the absence of other major or minor criteria and without diagnostic lab testing [7].

The onset of motor disturbance in SC is often insidious, with a waxing and waning course that ultimately resolves over months [3]. Movements are frequently asymmetric and more profound on one side, may be subtle and intermittent, and cease during sleep. Chorea is often accompanied by dysarthria and hypotonia. There may be associated behavioural changes which can include impulsivity, aggression, emotional lability, and obsessive-compulsive behaviours [3].

Due to the potential latency period between an active GAS infection and chorea presentation, laboratory testing is not always helpful. Elevated ASO titers are seen in up to 80% of patients with SC [8]. In addition, a throat culture may be positive due to GAS carrier status, or could be negative as the course of SC can occur months after an acute infection [8].

SC should be a diagnosis of exclusion for medical providers until other serious causes of chorea are ruled out (Table 2). There are a variety of investigations that may yield the cause of chorea and many of these investigations can be initiated in the Emergency Department (Table 3).

Once the diagnosis is made, treatment for SC is multifaceted. Primary and secondary prevention aids in reducing and eliminating GAS infections while also providing long-term prophylaxis against rheumatic heart disease. The American Academy of Paediatrics recommends penicillin to be used for both primary treatment and secondary prevention of ARF [9]. Recurrence of rheumatic fever episodes tends to occur within the first few years after an acute attack and is more common in individuals with pre-existing heart disease [4]. Typically, intramuscular penicillin is used in preventing recurrence; if the patient is unable to tolerate intramuscular injections, then oral prophylaxis is used. Symptomatic management of the choreiform movements with antiepileptics, neuroleptics or benzodiazepines may be beneficial [4]. There is limited evidence suggesting immunomodulatory agents such as IV steroids, plasma exchange, and intravenous immunoglobulin may shorten the course of illness [1,4,7,10].

Table 2
Differential diagnosis for acquired paediatric chorea.

Differential diagnosis for acquired paediatric chorea
• Autoimmune or inflammatory (antiphospholipid, vasculitis, SLE, Moyamoya) • Cerebrovascular (stroke, seizure) • Hematologic (polycythemia vera) • Drugs (antiepileptic) • Infection (Sydenham's chorea*, encephalitis, Lyme, HIV, EBV) • Metabolic (electrolyte abnormalities, thyroid abnormalities, B12 deficiency, Wilsons disease) • Neoplastic (basal ganglia, paraneoplastic) • Toxins (methanol, carbon monoxide, withdrawal) • Familial (Huntingdon's disease) • Psychogenic (Tourettes) • Ischemia (stroke, perinatal hypoxia-ischemia) • Other (Post cardiac surgery, chorea gravidarum)

Table 3
Suggested work up for acquired paediatric chorea.

Suggested workup for acquired paediatric chorea
• CBC, basic and extended electrolytes, TSH, free T4 • ESR/CRP, ANA, antiphospholipid/anticardiolipin antibodies, serum ceruloplasmin • Throat C + S, antistreptolysin O titer (ASOT), antiDNAse B titer • ECG, echocardiography • MRI brain • +/− bHCG • +/− urine drug screen

This case serves as a reminder to consider both benign and serious causes of acute chorea, and the diagnostic considerations involved in working up new cases of chorea.

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Declaration of competing interest

None.

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