Received: 6 January 2021 | Revised: 7 June 2021 | Accepted: 11 June 2021

DOI: 10.1002/emp2.12497

JACEP OPEN
JOURNAL OF THE AMERICAN COLLEGE OF EMERGENCY PHYSICIANS OPEN
A GLOBAL JOURNAL OF EMERGENCY MEDICINE
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WILEY

CASE REPORT

Pediatrics

Uncontrollable movements of right upper and lower extremities in a child: A diagnostic puzzle

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Dr Douglas and Dr Rimareva evaluated the patient initially in the emergency department and Dr Patki worked up the patient in the inpatient unit. Dr Patki and Dr Kondamudi wrote the initial draft with contributions from Dr Douglas.

Funding and support: By *JACEP Open* policy, all authors are required to disclose any and all commercial, financial, and other relationships in any way related to the subject of this article as per ICMJE conflict of interest guidelines (see www.icmje.org). The authors have stated that no such relationships exist

Abstract

We describe a case of a 9-year-old child who presented with uncontrollable, involuntary movements associated with a recent streptococcal infection and echocardiographic evidence of valvulitis. These findings are consistent with the diagnosis of Sydenham's chorea, a rare but important movement disorder and one of the major "Jones criteria" for the diagnosis of acute rheumatic fever. Because of its rarity, patients with Sydenham's chorea often are misdiagnosed as having a behavioral or psychiatric illness. Early recognition and appropriate management can prevent the potential severe sequelae associated with acute rheumatic fever.

KEYWORDS
darting sign, dyskinesias, hemichorea, milkmaid sign, rheumatic fever, spooning sign, valvulitis

1 | INTRODUCTION

Evaluating children presenting to the emergency department with abnormal movements could be challenging.[1] Hyperkinetic disorders (dyskinesias) are characterized by excess movements, such as tremors, tics, and dystonia and choreiform, and myocloic movements and are more common than hypokinetic disorders.[2] Chorea is the most frequently encountered movement disorder, and Sydenham chorea (SC), a manifestation of acute rheumatic fever,[3] is the most common cause in school-age children.[4] The clinical presentation can be acute or insidious, characterized by purposeless, irregular, and involuntary movements of extremities accompanied by facial grimacing. The declining incidence of rheumatic fever in developed countries translated to less incidence of SC[5] contributing to reduced awareness and recognition of this condition. The neurological manifestations of chorea are frequently attributed to behavioral or psychiatric conditions and sometimes to central nervous system infections, substance abuse, brain hemorrhages, or tumors. In contrast to rheumatic carditis and arthritis, which typically present within 21 days, the onset of chorea can occur up to 6 months after streptococcal infection.[6] Delays in the diagnosis of Sydenham's chorea, and by extension of acute rheumatic fever, can lead to severe untoward sequelae that would have been otherwise preventable. We report a case of a 9-year-old child who presented with right extremity involuntary movements and was diagnosed as having SC.

2 | CASE PRESENTATION

A 9-year-old African American female with no significant past medical history presented to the emergency department with the chief complaint of involuntary rhythmic movements of her right upper extremity and inversion of her right foot for 1 day. She was alert, oriented, and

otherwise seemed well. Vital signs on presentation were as follows: temperature 98.4°F, heart rate 108 bpm, blood pressure 119/76 mm Hg, respiratory rate 20/min, SpO2 97% in room air. The father reported that while he held her hand, she was alternating between gripping his fingers tightly and letting go in a rhythmic pattern. Her parents initially attributed this to her behavior but became concerned when she was unable to stop this voluntarily.

On further questioning, parents reported that 2 weeks before, the patient was treated at the local urgent care center with amoxicillin for sore throat and a positive rapid Strep test. Physical examination revealed an alert well-appearing child who was somewhat anxious and apprehensive. Ear, nose, and throat examination was remarkable for enlarged tonsils without any erythema, exudates, or lymphadenopathy. Neurological examination revealed an alert, active, anxious child with normal cranial nerves, good muscle tone, and slightly reduced reflexes in both upper and lower extremities. Right-sided non-stereotypical, jerking movements of the face were noted along with involuntary non-rhythmic movements of the right upper extremity. When asked to stretch out hands, the child had hyperextension at the metacarpophalangeal joints (spooning sign) and she was unable to keep her tongue protruded (darting sign). The grip was weak and when asked to hold firm, there was an alternating squeezing and releasing of the finger-like a milking motion (milkmaid sign). Her gait examination was remarkable for intermittent inversion of the right foot exaggerated during ambulation. No sensory deficits were noted in both upper and lower extremities.

Throat culture done on admission was negative, but the antistreptolysin O ttires were elevated (1511 Todd units) along with C-reactive protein (22 mg/dL). Urine toxicology was negative for toxins and drugs. Pediatric cardiology and neurology teams were consulted. Because of the unilateral nature of the movement disorder, neurology recommended imaging studies and electroencephalogram (EEG) to rule out a focal lesion. The non-contrast computed tomography (CT) head was normal. Two magnetic resonance imaging (MRI) studies done 3 days apart showed small T2 FLAIR hyperintense white matter lesions in the right frontal Corona Radiata with partial enhancement. Video EEG showed generalized internment rhythmic delta waves with no epileptiform discharges. Findings were non-specific and were suggestive of mild diffuse cerebral dysfunction. ECG showed sinus rhythm with normal rate, axis, and PR interval. An echocardiogram showed moderate mitral regurgitation without prolapse and mild aortic and tricuspid regurgitation consistent with subclinical carditis. The pediatric cardiology team confirmed the carditis and recommended monthly long-acting penicillin and repeat echo studies to monitor the cardiac function. Patient did not receive any specific therapeutic agent to control the neurological symptoms. The multidisciplinary team of intensivist, neurologist, and cardiologist opted to manage conservatively with watchful waiting rather than initiating any therapeutic drug regimen.

The neurology follow-up 3 months later showed significant improvement. At the most recent follow up nearly 9 months after the initial illness, parents reported no residual symptoms and her examination did not reveal any neurological abnormalities. Serial echocardiograms done at 3–month intervals showed progressive improvement of the valvular regurgitations. At the 9-month follow up there was residual mild aortic regurgitation and complete resolution of mitral and tricuspid valve regurgitations and no functional cardiac dysfunction. Patient remains on monthly penicillin prophylaxis and has remained compliant

The overall clinical picture was consistent with right hemichorea and subclinical carditis after a recent streptococcal infection, meeting the revised Jones criteria to diagnose acute rheumatic fever with SC.

3 | DISCUSSION

Clinical presentation of children with abnormal movements often poses a diagnostic challenge when encountered in the ED. Our patient presented with right-sided rhythmic involuntary movements a clinical picture compatible with hemichorea. Hemichorea is a somewhat uncommon condition whose underlying cause may be due to infectious, autoimmune, metabolic, vascular, and toxic causes.[7] Focal lesions[8] affecting the basal ganglia can present with hemichorea. A rare progressive autoimmune disorder in children known as Rasmussen's encephalitis[8] also can be associated with hemichorea. Other neurological conditions in the differential diagnosis include Wilson disease, systemic lupus erythematosus, and drug reactions. It is important to distinguish choreiform movements from other abnormal movements, such as tics, athetosis, conversion reaction, and other forms of hyperkinesis. SC, a manifestation of acute rheumatic fever, is still the most common acquired cause of hemichorea in children.[4] SC more commonly presents with generalized involvement but unilateral abnormalities can occur in 20%–30% of cases.[3] It is unusual that our patient presented within 2 weeks of group A streptococcal infection, rather than the typical time frame of 6 to 8 weeks.[6] Complete resolution of symptoms in our patient at 8 months, instead of the usual 12–15 month period, is also another unusual feature.

The major neurological features of SC are involuntary rhythmic dance-like movements (St. Vitus dance) exacerbated by stress but disappearing during sleep. These often are associated with incoherent speech, dysarthria, muscle weakness, and hypotonia.[9] Clinical signs that can be elicited in affected patients with SC owing to the hyperkinesia include spooning, darting, and milkmaid signs. Chorea could be the sole manifestation of acute rheumatic fever and is one of the Jones major criteria for the diagnosis of acute rheumatic fever.[10] Concurrent carditis as seen in our patient is the most frequent other associated major criteria.[4] The association of SC with the remaining 3 major Jones criteria of arthritis, subcutaneous nodules, and erythema marginatum is rare.[10] Our patient had no overt clinical evidence of carditis but had evidence of valvulitis in both mitral and aortic valves. Subclinical carditis is present when there is echocardiographic evidence of valvulitis without auscultatory findings and is one of the major revised Jones criteria.[4] Because of the unilateral nature of the symptoms, our neurologist recommended a CT scan of the head and brain MRI to exclude a focal lesion. These studies are generally not indicated in a typical patient with SC. When performed, these imaging studies are usually

normal, although reversible T2 hyperintensity[11] in the basal ganglia and cerebral white matter[12] has been described.

Rheumatic fever is believed to be an autoimmune phenomenon triggered by a streptococcal infection resulting in a hypersensitivity-like reaction due to cross-reactivity between the streptococci and human tissue antigens through antigen mimicry.[13] The antibodies target the basal ganglia brain cells in the host and cause a diffuse inflammatory process in the corpus striatum, mainly the caudate nucleus. It is theorized that the symptoms of SC are a result of an imbalance between the dopaminergic system, intrastriatal cholinergic system, and the inhibitory gamma-aminobutyric acid system.[13]

In conclusion, it is important to recognize SC in children and initiate appropriate management for both neurological symptoms and acute rheumatic fever. Agents available to use include dopamine 2 receptor blocking agents such as haloperidol and pimozide and other drugs such as carbamazepine, valproate, phenobarbital, and diazepam.[14] In moderate to severe cases, corticosteroids and intravenous immunoglobulins[15,16] have been shown to decrease the duration of illness through immune suppression. There is no established protocol for the symptomatic treatment for chorea, which is largely determined by the extent of disruption of daily activities. When symptoms are mild or transient, conservative approach with watchful waiting is preferred. Short-term therapy may be indicated if there is significant disruption including moderate gait disturbance.[17] Our neurology team did not recommend pharmacotherapy for this child, which would have been indicated if there were persistent symptoms that interfere with daily activity. Antibiotic therapy to eradicate group A streptococcal pharyngitis should be initiated and secondary prophylaxis with long-acting intramuscular penicillin G benzathine be given monthly to prevent recurrence of acute rheumatic fever and its debilitating complications including cardiac failure.[15,18] SC should be considered in any school-age child presenting with a movement disorder and evidence for recent streptococcal infection sought. Prompt diagnosis can help prevent the serious sequelae of acute rheumatic fever.

CONFLICT OF INTEREST

The authors report no conflicts of interest.

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How to cite this article: Patki R, Douglas R, Rimareva N, Kondamudi N. Uncontrollable movements of right upper and lower extremities in a child: A diagnostic puzzle. *JACEP Open*. 2021;2:e12497. http://doi.org/10.1002/emp2.12497