

## Case Report

# Hemichorea as a presentation of acute rheumatic fever: a case report

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### ABSTRACT

Chorea is a major manifestation of acute RF and is the only evidence of RF in approximately 20% of cases. We report on a 15-year-old boy who presented with transient right side involuntary jerky movements, apical systolic murmur, sinus bradycardia, arthralgia, elevated antistreptolysin O titer and ESR, who was diagnosed with acute rheumatic fever and improved with haloperidol, prednisolone, digoxin, aspirin and furosemide and was given benzathine penicillin prophylaxis for future RF. Patient is faring well in follow up visits. We present our case because of its rarity.

**Keywords:** Hemichorea, Jones Criteria, Rheumatic fever, Sydenham's chorea

### INTRODUCTION

Rheumatic Fever (RF) is a public health concern due to carditis and heart damage, which may be aggravated by late diagnosis and poor penicillin prophylaxis adherence. Early diagnosis of acute RF though difficult is very important to prevent the serious consequences in young. Sydenham chorea (SC) is the most common cause of acquired chorea in the young. The incidence of RF is higher in developing countries, where the absence of consistent and early antibiotic treatment makes RF a endemic problem. According to the 1992 modified Jones Criteria, chorea (or indolent carditis) alone is sufficient for the diagnosis of RF, provided the other causes have been excluded.<sup>1</sup> Chorea is characterized by involuntary movements of extremities, muscular hypotonia, dysarthria, gait disturbance, and tics, as well as behavioral manifestations. The choreic movements interfere with volitional movements and result in a clumsy gait, dropping and spilling. Symptoms are usually self-limited and on average it resolves spontaneously in 3-6 months and rarely lasts longer than 1 year. SC is usually bilateral and female predominant.<sup>5,8,11</sup>

### CASE REPORT

A 15-year-old boy presented with a 3-days history of episodic involuntary movement of his right limbs. On examination, he twisted his right hand and leg and had a clumsy gait and frequent jerking movements. He had noticed deterioration in his handwriting skills and had increasing difficulties with speaking, walking, and daily activities such as manipulating combs, spoons and drinking from a glass. The involuntary movements disappeared during sleep and became apparent when the patient was anxious. He was emotionally labile during that period of time. He had a mild fever and cough and sore throat 12 days prior to the onset of the involuntary movements and it subsided on its own. The medical history and family history were unremarkable. Review of systems revealed palpitations and bradycardia. Arthralgia was noted. The initial physical examination revealed a slightly decreased heart rate about 56 per minute and a grade 4/6 high-pitched blowing systolic heart murmur at the apex. No skin rash, subcutaneous nodules or joint swelling was found. His neurological examination was remarkable for right hemichorea and slurred speech. He

had frequent facial grimaces and choreic movements of his shoulders and neck. Deep tendon reflexes were slightly reduced but muscle power were preserved. CRP was elevated. Renal, liver and thyroid function test were normal. Due to the chorea, raised ASO and ESR, and arthralgia, belonging to endemic region, age, poor socio-economic condition, rural location and poor sanitation and hygiene, acute rheumatic fever was suspected, although hemichorea is not a common presentation of acute rheumatic fever. Albeit possibility of focal seizure with possible intracranial space occupying lesions like tuberculoma, neurocysticercosis were kept however MRI brain revealed no abnormality. Laboratory studies revealed an elevated antistreptolysin O titer of 794 IU/ml and an increased erythrocyte sedimentation rate of 42 mm/hr. A chest xray showed borderline cardiomegaly. Electrocardiogram revealed sinus bradycardia. Echocardiography showed moderate mitral regurgitation and dilatation of the left atrium and left ventricle with mild ventricular compromise and electroencephalogram were within normal limits. A diagnosis of acute rheumatic fever with Sydenham's chorea was made based on the presence of chorea, apical murmur, concordant echocardiographic findings, elevated acute phase reactant, arthralgia, conduction disturbance of bradycardia and presumptive evidence of prior streptococcal infection. Haloperidol 0.03 mg/kg/day was given and the chorea improved 2 days after therapy.

After 2 days bradycardia improved. Treatment of the carditis included prednisolone 2 mg/kg/day for 2 weeks followed by a tapering of the dose and aspirin 100 mg/kg/day (after prednisolone tapering) for 1 month followed by gradual withdrawal. Digoxin 4 mcg/kg/day and furosemide 1.5 mg/kg/day were used for mild heart failure. After 4 weeks echo and heart size was normal. He was given benzathine penicillin prophylaxis, and patient is faring fine in follow up visits.

## DISCUSSION

There is still no single symptom, sign, or laboratory test that is pathognomonic or diagnostic of acute rheumatic fever. The guidelines for the diagnosis of acute rheumatic fever are based on the 1992 updated Jones criteria.<sup>1</sup> Chorea consists of rapid, involuntary movements observed on physical examination, is worsened by stress and anxiety and subsides during sleep. Sydenham's chorea (SC) is a cardinal feature of rheumatic fever and is sufficient alone to make the diagnosis.<sup>1</sup> SC is a disease of childhood or early adolescence mostly reported in patients 5 to 15 years old. SC was reported in about 10-15% of patients with rheumatic fever but the incidence was relatively low, around 1-8%, in Africa, the Pacific, South and East Asia and the Arabian Peninsula.<sup>2-10</sup> In most patients, Sydenham's chorea is bilateral, and hemichorea has been reported in only 15-20% of patients.<sup>5,7</sup> It occurs considerably more often in girls than boys in the teen years, occurring at a ratio of approximately 2:1.<sup>3,5</sup> Because the onset of chorea is

usually 1 to 6 months after the inciting streptococcal infection, other rheumatic manifestations may not be found at presentation.<sup>5,12</sup> In our patient, the hemichorea was suspected as focal seizures as the second differential diagnosis which was withdrawn upon normal MRI of brain. MRI findings in SC are not consistent and may be normal. Bradycardia was not recognized as an early sign of myocarditis. Diagnosis of SC may be difficult, because no single, established diagnostic test is available. The diagnosis of acute rheumatic fever was made as patient had chorea, arthralgia, bradycardia, raised ASO titre and ESR and preceding history of fever with sore throat. Although hemichorea in rheumatic fever is an unusual presentation, we emphasize the importance of the differential diagnosis of focal seizures and hemichorea. SC is the most common cause of acquired chorea in children.<sup>12</sup> There are other diseases that cause chorea in childhood. The differential diagnosis is mainly between SC, lupus-associated chorea and drug-induced chorea.<sup>10</sup> Sometimes, chorea induced by another disease is misdiagnosed as SC. The common causes of chorea include atypical seizures, brain tumor, cerebrovascular accident, collagen vascular disorders such as systemic lupus erythematosus and Behcet's disease, drug intoxication, endocrine disorders such as hyperthyroidism and hypoglycemia, hereditary disorders such as Huntington's chorea and Wilson's disease, pregnancy and viral encephalitis.<sup>11-13</sup> The causes of hemichorea are the same as those of bilateral chorea.

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