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Sydenham Chorea: Rare Consequence of Rheumatic Fever

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CASE REPORT

Sydenham Chorea (SC) is an acute rheumatic fever complication. SC is the most common acquired childhood chorea, usually affecting children five to fifteen years of age.¹ It occurs following an untreated group A streptococcal infection and a latent period of one to six months.^{1,2} Despite rheumatic fever diminishing, 18% to 36% of acute rheumatic fever patients develop SC.³ Full recovery often takes several months; some patients suffer permanent neurologic sequelae.¹

An 11-year old male presented to the Emergency Department with two days of uncontrolled body twitching. The movements affected his right arm and leg, with occasional lip twitches; he experienced intermittent confusion and hyperactivity. The patient denied recent illness, but recalled a fever with headache and vomiting several months prior. Besides the above findings, his physical examination was normal.

The patient's rapid streptococcal antigen test was negative, but his throat culture was positive for group A beta hemolytic Streptococcus. An anti-streptolysin O (ASO) titer resulted at 503 (reference range, <240 IU/mL). Symptoms progressed to include slurred speech, head jerking, awkward gait, and decreased right eye vision.

Several motor manifestations are pathognomonic: "Milkmaid's grip" occurs when patients are unable to clench their fists, displaying as intermittent relaxation and tightening of hand grip.¹ "Choreic hand" is "spooning" of the hand by wrist flexion and extension of the digits.¹ The linked video demonstrates this patient's motor manifestations (Video).

Evaluation of chorea in pediatric patients should include testing for group A streptococcal infection with throat culture and ASO titers. SC is key to diagnosing rheumatic fever and should

prompt evaluation for rheumatic heart disease. This patient's brain magnetic resonance imaging and electrocardiogram were normal, but his echocardiogram showed mild mitral regurgitation. Treatment included penicillin for ten days, instructions to get monthly bicillin injections through age twenty-one and a recommendation for lifelong antibiotic prophylaxis.

Video. Motor manifestations of patient.

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