



Sydenham's Chorea in the Age of MRI: A Case Report and Review

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A 9-year-old female with acute chorea was found to have multiple areas of abnormal signal on magnetic resonance imaging of the brain consistent with vasculitis. The child's serology and clinical course were indicative of Sydenham's chorea, and other causes of vasculopathy were excluded. Although magnetic resonance imaging findings similar to those observed in this patient have been rarely reported in Sydenham's chorea; their presence alone should not prompt a search for an alternative diagnosis. © 2002 by Elsevier Science Inc. All rights reserved.

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Introduction

Sydenham's chorea is a major manifestation of acute rheumatic fever [1]. The disorder is characterized by chorea, muscular weakness, and a number of neuropsychiatric symptoms. Sydenham's chorea is thought to be an autoimmune disorder resulting from a prior group A beta hemolytic streptococcal infection [2]. During the latter part of the twentieth century there was an increase in the number of reported cases of rheumatic fever in the United States. The resurgence of rheumatic fever in this country appears to be associated with strains of group A beta hemolytic streptococcal infection that are less likely to cause symptomatic pharyngitis [2,3]. Without clinical

evidence of carditis or other manifestations of rheumatic fever, early diagnosis of Sydenham's chorea may be difficult [4]. Magnetic resonance imaging (MRI) of the brain and computerized tomography (CT) of the head are often obtained to exclude other causes of chorea. Computed tomography scans are typically normal, and MRI studies have not demonstrated consistent abnormalities [2,5,6].

We recently cared for a child with acute chorea whose initial brain MRI demonstrated multiple high-intensity white matter lesions suggestive of vasculitis.

Case Report

A healthy 9-year-old female began to have involuntary purposeless movements of her left arm. The child became irritable and emotionally unstable. Over the next several days her speech became difficult to understand, and she appeared weak on her left side. There had been no recent illnesses or medication exposure. The child was an excellent student, and there had been no decline in school performance. There were no prior medical problems.

Examination revealed quick random movements involving the distal aspects of the left upper extremity. Speech was slurred, and there was an occasional flinging movement of the left arm. Less severe but similar movements were observed in the right arm. Diffuse hypotonia was present and the child could not sustain a fixed posture. There was a mild left hemiparesis. The heart had a normal sinus rhythm without murmur.

Initial laboratory studies included a negative throat culture and antistreptolysin O titer. Erythrocyte sedimentation rate was 48. An antinuclear antibody screen was positive at a dilution of 1:80 with a nonspecific speckled pattern. Electrocardiogram finding was normal. Echocardiogram revealed mild insufficiency of the aortic valve consistent with carditis. Brain MRI revealed multiple foci of increased signal on T₂-weighted and fluid-attenuated inversion recovery images in the cerebral white matter (Fig 1). The MRI abnormalities were consistent with vasculitis or multiple areas of demyelination.

Magnetic resonance angiography of intracranial and extracranial vessels was entirely normal. Lupus anticoagulant, anticardiolipin antibodies, and rheumatoid factor were negative, as were antineutrophil cytoplasmic and anti-DNA antibodies. Antibodies to extractable nuclear antigens were not detected.

The patient received 10 days of oral penicillin. She was placed on low-dose haloperidol (1 mg at night), and her involuntary movements diminished. The child was discharged on penicillin prophylaxis and haloperidol.

Five weeks later the patient was improved. Her erythrocyte sedimentation rate was 28, and streptolysin O antibodies were present with a titer of 1,400 IU/mL. Magnetic resonance imaging studies 4 and 9 months after being released from the hospital revealed no change in the number of white matter lesions. At last follow-up the child was asymptomatic and was no longer receiving haloperidol.

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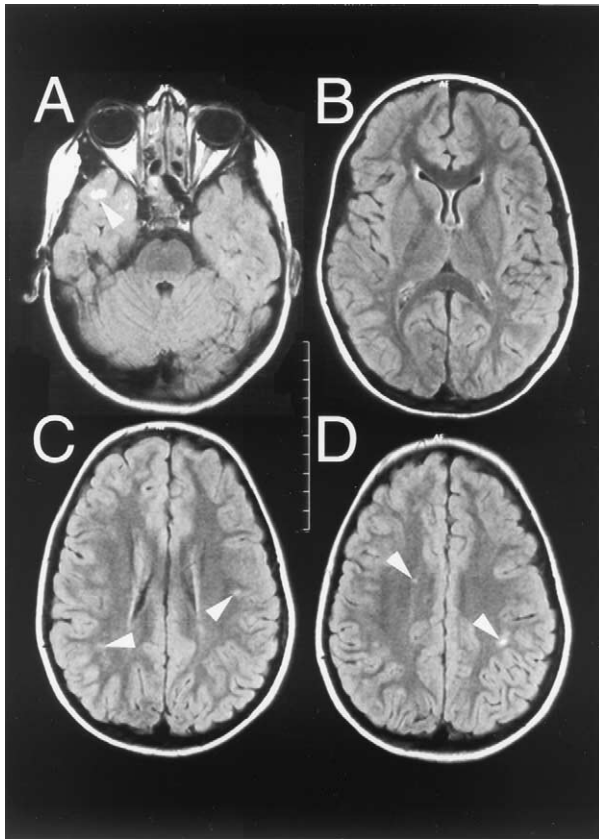


Figure 1. Axial fluid-attenuated inversion recovery magnetic resonance images from child of 9 and 1/2 years of age with Sydenham's chorea demonstrate multiple peripheral white matter hyperintensities (arrowheads) in the right anterior temporal lobe (A) and parietal lobe bilaterally (C, D). Note the absence of hyperintensity or other abnormality in the basal ganglia in B. There were no lesions in the subthalamic nuclei, brainstem, or cerebellum (not shown) (TR = 9000 ms, T₁ = 2500 ms, TE = 105 ms).

Discussion

Sydenham's chorea usually develops between 3 and 13 years of age and is believed to follow a streptococcal infection. Diagnosis, however, is not dependent on serologic evidence of preceding exposure [7]. Streptococcal infections can be subclinical and often precede the development of neurologic symptoms by 1-6 months. At least 25% of patients with Sydenham's chorea will fail to have serologic evidence of prior infection [3].

Sydenham's chorea is a major manifestation of acute rheumatic fever (rheumatic fever) and is the only evidence of rheumatic fever in 20% of cases [7]. The combination of Sydenham's chorea and clinical evidence of another major manifestation of rheumatic fever remains uncommon; however, echocardiography can detect cardiac involvement in over 60% of patients with Sydenham's chorea. Nevertheless, isolated chorea is sufficient for diagnosis of rheumatic fever provided other causes of chorea have been excluded (Table 1) [1,8].

Magnetic resonance imaging findings in Sydenham's chorea are not consistent, and studies may be normal. Published abnormalities include areas of increased signal

Table 1. Diagnostic criteria for first attack of acute rheumatic fever*

1. Evidence for a prior group A streptococcal injection plus
2. Either two major or one major and two minor manifestations

Major Manifestations	Minor Manifestations
Carditis	Arthralgia
Polyarthritits	Fever $\geq 39^{\circ}\text{C}$
Chorea	Elevated acute-phase reactants
Erythema marginatum	Sedimentation rate
Subcutaneous nodules	C-reactive protein
	Prolonged PR interval

*According to the 1992 modification of the Jones criteria, chorea or indolent carditis alone are also sufficient for diagnosis provided other causes for the conditions have been excluded.

intensity on T₂-weighted images that usually involve the basal ganglia or cerebral white matter [4,6,7,9]. Occurrences of multiple white matter lesions suggestive of ischemic foci have been rarely reported. One study reported an increase in basal ganglia volume consistent with localized swelling [2]. Follow-up studies may demonstrate improvement, although some residual abnormality is common [4,5,7,9].

Kienzle et al. in 1991 [9] reported MRI findings in two children with unilateral choreiform movements whose clinical course and evaluation were consistent with Sydenham's chorea. Magnetic resonance imaging in both patients revealed increased signal intensity in the contralateral striatum [9]. In their review of the physical and psychologic symptoms of Sydenham's chorea, Swedo et al. [6] also described brain MRI findings in 11 patients. Imaging in seven patients was normal, and studies in two other patients demonstrated only congenital abnormalities (a right parietal schizencephalic cleft and minimal ventricular asymmetry). The two children with the most severe chorea had foci of increased signal on T₂-weighted images. Brain MRI in one of these patients revealed a single area of increased signal in the left centrum semiovale. The other child's study revealed a small focus of increased signal in the posterior left thalamus [6].

In 1995, Traill et al. [5] described a patient with Sydenham's chorea with findings on cranial CT of bilateral loss of attenuation in the striatum. Their patient's MRI demonstrated increased signal intensity on T₂-weighted images involving the putamen, globus pallidus, and head of the caudate nucleus bilaterally [5]. Emery and Vieco [4] later described a patient with hemichorea secondary to Sydenham's chorea. Their patient's initial MRI revealed swelling and increased signal on T₂-weighted images of the contralateral caudate and putamen [4].

In 1999 Ryan and Antony [7] reported a patient with Sydenham's chorea with brain MRI findings of multiple areas of increased T₂ signal within the cerebral white matter. Their patient's MRI findings suggested foci of ischemia or demyelination. Subsequent magnetic resonance angiography and cerebral angiography revealed

abnormalities in the left middle and left anterior cerebral arteries consistent with vasculitis [7].

Evidence suggests Sydenham's chorea may result from the production of immunoglobulin G antibodies that cross-react with antigens in the membrane of group A streptococci and antigens in the neuronal cytoplasm of the caudate and subthalamic nuclei [2,3]. The few available neuropathologic studies have demonstrated inflammatory vasculitis, neuronal swelling and degeneration, and petechial hemorrhages in the basal ganglia, cerebral cortex, and cerebellum [3,9,10,11]. Although these findings were evident after severe illness, less severe cases of Sydenham's chorea could also be associated with inflammatory changes and vasculitis of sufficient magnitude to produce abnormal findings on brain MRI.

Causes of chorea in the young include moyamoya disease, Wilson's disease, medication and drug toxicity, systemic lupus erythematosus, and other causes of inflammation of the cerebral vessels [1,7]. The presence of increased signal intensity on T₂-weighted and FLAIR images in our patient resulted in a diagnostic evaluation for vasculitis. Other causes for vasculitis were excluded, leading us to conclude that Sydenham's chorea may be associated with abnormal MRI findings suggestive of an ischemic vasculopathy.

Our experience supports the findings reported by Ryan and Antony [7] that Sydenham's chorea may be associated with diffuse areas of increased signal within the cerebral white matter. Although such abnormalities are nonspecific and could be secondary to demyelination, we believe these findings may be secondary to small-vessel vasculitis. The occurrence of such findings in an otherwise typical case of

Sydenham's chorea should not prompt an extensive investigation for other vasculitides.

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