Chorea Gravidarum

A Case Report Including Magnetic Resonance Imaging Results

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Chorea gravidarum is a rare movement disorder that occurs in pregnancy. Although once quite common, it is now considered to be an unusual finding. Several illnesses are associated with this disorder including rheumatic fever, systemic lupus erythematosus, and Huntington's disease. The following is a case report and discussion of the clinical aspects of this disorder.

CASE REPORT

The patient was a 26-year-old, gravida 1, para 0, woman at 26 weeks' gestational age by dates, who was admitted to the hospital with complaints of weakness and numbness in her right hand and occasional involuntary movements of the same hand and wrist. Her symptoms had progressed over the 3 weeks prior to admission.

The patient's past medical history was significant for the diagnosis of rheumatic heart disease, which was made on angiographic study for workup of a heart murmur when she was 3 years old. There was no history of Sydenham's chorea or St. Vitus' dance. In addition, the patient was hospitalized in 1983 for suspected subacute bacterial endocarditis caused by infection with *Streptococcus viridans*. At that time, she was noted to be mildly encephalopathic.

The patient had no history of drug or alcohol abuse or exposure to environmental toxins. Her medical history also did not suggest a collagen vascular process. Family history was negative for Huntington's disease.

Physical examination revealed the patient to be afebrile with blood pressure 120/70 mmHg, pulse 80 beats per minute and regular, and respirations 16/min. The patient did exhibit some choreoathetoid movements of the right face, arm, and leg. Findings on the remainder of the head, eyes, ears, nose, and throat examination as well as the respiratory examination were unremarkable. Cardiovascular examination revealed a grade 3/6 pansystolic murmur, which was heard loudest at the apex. Abdominal examination showed a gravid uterus consistent with dates. There was no hepatomegaly or splenomegaly. Extremities showed no clubbing, edema, cyanosis, or joint abnormalities.

Neurologically, the patient exhibited repetitive, involuntary, brisk, snakelike movements, which were made worse when she became anxious. She had some decreased sensation to light touch and pinprick in the right forearm. Her deep tendon reflexes were symmetric. Decreased muscle strength was noted in the right upper extremity. The patient did have poor finger-to-nose and rapid alternating movements in the right upper extremity secondary to the underlying involuntary movements. Heel to shin testing was normal, and Romberg sign was negative.

Admission laboratory tests showed the patient's chemical profile to be within normal limits. Complete blood count showed a hemoglobin of 110 g/L (11 g/dL), hematocrit of 0.31, white blood cell count of 8.6×10^9/L ($8.60 \times 10^3/\mu$L), and platelets of 274×10^9/L ($274 \times 10^3/\mu$L). Antistreptolysin 0 titer was 1:125. Rheumatoid factor was negative. Westergren method sedimentation rate was 82 mm/h. Thyroid function studies were within normal limits. Urinalysis was noncontributory. A screening test for *Streptococcus* was negative, and three blood cultures were negative. Antinuclear antibody test was negative. An electrocardiogram showed a normal sinus rhythm with normal ST and T waves.

Echocardiogram showed thickened anterior and posterior leaflets of the mitral valve with mitral valve prolapse and mild to moderate mitral regurgitation. Ultrasound of the abdomen showed a single viable fetus with vertex presentation and 25 weeks' gestational age by biparietal diameter.

Magnetic resonance imaging (MRI) showed focal high-signal regions in the left coronal radiata at the level of the

Submitted, revised, May 1, 1989.

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left lateral ventricle and the left cerebellum, compatible with infarction, demyelination, or inflammation.

DISCUSSION

Chorea gravidarum is a rare movement disorder involving the limbs and face that occurs in pregnancy.[1-3] This syndrome was once relatively common, but now is an extremely unusual finding.[3,4] The movement disorder is choreiform in type, characterized by involuntary, repetitive movements that may be either unilateral or bilateral and often worsen in stressful situations.[3-5] Loss of muscle tone and lack of coordination may occur in association with the movement disorder.[2] Acute psychosis may also accompany these symptoms.[2,3]

The onset of chorea gravidarum usually occurs during the second or third trimester of pregnancy.[4] Although it commonly presents as a movement disorder, slurred speech may be noted as the initial clinical finding.[2] This condition usually resolves within a few days of delivery and has a strong tendency to recur with subsequent pregnancies.[2,4] Although once considered to carry a high mortality, the risks from chorea gravidarum are now considered negligible.[2,3,6]

Before the availability of antibiotics, the occurrence of chorea with pregnancy was largely due to acute rheumatic fever. Now that acute rheumatic fever has become rare, the majority of instances of chorea gravidarum appear to be secondary to previous rheumatic fever rather than an acute episode.[2,4,5] Jonas et al[7] showed that chorea gravidarum was not necessarily related to a recent or current infection caused by *Streptococcus* but rather could occur in patients with a history of rheumatic fever in the past but no evidence of an acute recurrence. Chorea may or may not have occurred with the original episode of rheumatic fever in patients who later develop chorea gravidarum during pregnancy. Recent studies have shown that chorea gravidarum may also be a manifestation of systemic lupus erythematosus or Huntington's disease.[4,8-12] These patients probably represent the 30% with chorea gravidarum who have no history of rheumatic fever who were reported in the original studies by Willson and Preece.[13]

The cause of chorea gravidarum appears to be related to a preexisting neuropathological lesion activated by increased estrogenic effects of pregnancy.[3,4,8] Most investigators suggest a lesion in the area of the basal ganglia and corpus striatum that consists of an absolute or relative increase in dopamine activity.[3,4,6,13-15] Estrogen evaluation of pregnancy may stimulate dopamine receptor activity in this area of the brain.[4,15]

Various forms of therapy for chorea gravidarum have been recommended throughout the years. In treating this disorder, one must remember that this condition is self-limited and mortality is negligible.[8] Termination of pregnancy is not considered a primary mode of treatment for this disorder but, rather, is reserved for exceptionally severe, progressive, and resistant cases.[8,13]

Medical treatment of chorea gravidarum should probably be considered only for severe cases occurring in the second or third trimester in patients for whom the chorea is severe enough that the risk of injury in high.[15] Although phenobarbital and chloral hydrate have been used in the past, haloperidol now appears to be the drug of choice for the treatment of chorea gravidarum.[4,5,15] Symptoms can also be controlled with diazepam and chlorpromazine. Obviously, the risks of these medications in pregnancy have to be considered in light of the relatively benign, self-limited nature of chorea gravidarum. Haloperidol, however, appears to have little, if any, risk of birth defects if used during the second and third trimester.[6,15] The recommended dose for haloperidol is 2 to 6 mg/d. Severe cases can require as much as 20 mg/d.[15]

As previously mentioned, the prognosis for chorea gravidarum is excellent. Mortality is very rare and is often the result of the underlying disease rather than the chorea itself.[3,16] Although patients suffering from chorea gravidarum are at risk for relapse, it is not considered a contraindication to future pregnancies.[2,4,5]

The case presented here is unique in that a recent review of the literature was unable to locate any reports in which MRI was performed on a patient with this disorder. The patient's clinical presentation of unilateral choreoathetoid movements may have localized as a result of multiple additive insults to the coronal radiata. The abnormal findings noted on MRI may represent an area of significant edema and hemorrhage from rheumatic fever or previous infarction from septic emboli and may not be the direct result of her present illness. These lesions could have become clinically evident under the stress of pregnancy. Further MRIs in patients with chorea gravidarum will help to further describe the radiographic pathology associated with this disease.

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