Chorea Gravidarum Associated with Poor Perinatal Outcomes: Report of Two Cases

Kötü Perinatal Sonuçlar ile İlişkili Gebelik Koresi: İki Ölgu Sunumu

ABSTRACT Chorea Gravidarum (CG) is a rare syndrome which usually presents in the first trimester of pregnancy with bilateral repetitive, brief, jerky, dance-like, involuntary movements and slurred speech. CG is a self-limiting condition which is probably caused by estrogen which enhances receptor sensitivity to dopamine. Prognosis of CG for pregnant women and fetuses are believed to be good. In this report, we present two cases of CG secondary to rheumatic fever that were associated with adverse fetal outcomes. Both cases had elevated antistreptolysin O titer and mitral valve disease. In both cases, pregnancy was associated with hypertension. In one case, the previous pregnancy was also complicated with CG and fetal demise. Contrary to widespread belief, CG may be associated with adverse fetal outcome.

Key Words: Chorea gravidarum; pregnancy; hypertension


Anahtar Kelimeler: Gebelik koresi; gebelik; hipertansiyon

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Chorea is an involuntary abnormal movement, characterized by rapid, repetitive, non-rhythmic, brief movement of any limb. Chorea gravidarum (CG) is the term given to chorea occurring during pregnancy. The incidence of CG has been reported as ranging from 1/3500 to 1/140,000, with current maternal mortality of 1%. Since CG is a very rare entity, its pathogenesis, natural course, treatment and effects on the outcome of pregnancy have yet to be fully elucidated.

With this background, we report two cases of CG with poor obstetrical outcome, and compare our cases with the available literature.
CASE REPORT

CASE 1
A 28 year, G5 P4, 15 weeks old pregnant was ad -mitted to the clinic with the complaint of slurred speech, and severe unintentional movements of right hand, and foot that started two weeks before. Her neurological evaluation revealed right hemi -chorea, and motor dysr hytmia. She had chronic hypertension treated with alpha methyl dopa (4x 250mg), and mitral valve disease secondary to acute rheumatoid fever. She had also history of the CG in her last pregnancy which diminished slowly during the pregnancy without treatment. Her tests for anti-phospholipid antibody syndrome, inherited thrombophilia, Wilson Disease, Systemic lupus erythamatosus (SLE), coagulation parameters, urine, and blood biochemistry, thyroid function tests were all normal. She had normal first trimester Down syndrome screening test (1/1235). Antistreptolysin O titer was 313 IU/mL (0-200 IU/mL). Her cranial MRI was also normal. Valproic acid (400 mg thrice daily), and prednisolone (10 mg/day) were started at 16th weeks. Ultrasonography revealed fetal diaphragmatic hernia and severe hydro -cephaly at 23 weeks of gestation. The family selected terminat ion of the pregnancy. Autopsy confirmed the ultrasonographic findings. After the six months termination, the woman was completely free of symptoms.

CASE 2
A 18 years old, G2 P1, 22 weeks old pregnant woman was referred to our clinic for hypertension, and intrauterine growth restriction. Her obstetri -cal evaluation revealed, 22 weeks' fetus with intrauterine growth restriction, and oligohydram -nios. Her obstetric history revealed CG and fetal demise due to uteroplacental insufficiency secondary to preeclampsia in her previous pregnancy. Her blood pressure was 150/90mm Hg, and no proteinuria was detected. She was also consulted with neurology, and cardiology because of choreic movements of the left extremities. Neurological evaluation revealed chorea gravidarum (Sydenham Chorea). Her tests for anti-phospholipid syndrome, inherited thrombophilia, Wilson Disease, SLE, co -agulation parameters, urine, and blood biochemistry, thyroid functions tests were all normal. Antistreptolysin O titer was 299 IU/mL (0-200 IU/mL). Her cranial MRI was also normal. She was started on prednisolone therapy (10mg/day). At 25 weeks of pregnancy, in utero fetal demise was detected. Fetal autopsy revealed no congenital anomaly. Six months postpartum, she has still choreiform movement, and declined treatment.

DISCUSSION

Chorea gravidarum is broad term encompassing chorea of any causes starting during pregnancy. CG is rare maternal complication which usually presents in the first trimester of pregnancy. Most patients with CG are young; the average age is 22 years. Of initial attacks, 80% of all cases occur during first pregnancies, and, of these, one half starts during the first trimester. The severity of the chorea typically decreases as the pregnancy progresses. Approximately one third of patients go into remission after delivery, and another subset are essentially free of the illness after delivery. In its most severe form, CG can also cause hyperthermia, rhabdomyolysis, myoglobinuria, and eventually death. CG is usually self- limited condition and manageable non-pharmacologically. In severe cases, early treatments include neuroleptics, anti-convulsants, and steroids for symptomatic relief and therapies targeting toward underlying patholo -gies. Drug treatment indicated for patients with unremitting severe chorea affecting daily life. Half the cases of CG are idiopathic, and rheumatic fever and antiphospholipid syndrome are underlying causes of the most of the remainder. The other cause of CG are SLE, Huntington disease, thyrotoxicosis, sub-cortical infarction, drug induc -tions, and moyamoya disease. In both of our cases, the presence of cardiac valvular disease and high titer of anti-streptolysin A strongly suggest rheumatic fever (Sydenham Chorea) as an underlying cause for CG.

Prognosis of CG for pregnant women and fe -tuses are believed to be good, and termination of pregnancy is not recommended. In contrast to this
data, two pregnancies of the case two had complicated with hypertensive diseases of pregnancy (Preeclampsia, hypertension), uteroplacental insufficiency and fetal demises. Similarly, case one had chronic hypertension exacerbated during pregnancy, and fetal anomalies leading to termination of pregnancy. Valproic acid is an antiepileptic drug and has been used to treat movement disorders, including chorea. As in our case 1, valproic acid can be used as a first line agent in chorea due to its superior effect compared with carbamazepine and haloperidol in patients with severe symptoms. Valproic acid is a teratogen and increases the risk of major congenital malformations. However, the use of valproic acid is not associated with diaphragm hernia, and the use of this drug in the second trimester is not associated with major malformations. Therefore, the co-occurrence of diaphragmatic hernia and CG seems to be an incidental, and sporadic finding.

In our study, evaluation of the both cases for antiphospholipid syndrome, inherited thrombophilia, and SLE which may cause chorea and adverse pregnancy outcome were negative. Several pathogenic mechanism for CG have been offered, suggestion is that estrogen enhanced receptor sensitivity to dopamine and induces chorea in individuals who are vulnerable to this complication by preexisting pathology in the basal ganglia. Theoretically, the presence of placental dysfunction accompanying preeclampsia and/or hypertension may cause abnormal interaction between placental hormones, cytokines and central nervous system. At the other hand, the presence of biochemical pathology affecting dopaminergic system at the basal ganglia, may cause dysfunction at other areas of the brain controlling autonomic function, and lead to worsening hypertension, and preeclampsia. Very recently Maia et al, in their series including 66 cases, reported increased abortion rate in women with recurrent CG. In accordance with our findings, Palanivelu reported a case of CG associated with intrauterine growth restriction documented with abnormal Doppler evaluation. Though current literature suggest emotional shocks as the triggering factor for CG, it is not clear whether complicated pregnancy is the emotional cause or result of CG. Taken together, intricate network of interaction between CG, and placental dysfunction secondary to hypertension/preeclampsia may be suggested. Unfortunately, perinatal data of the cases which had been reported previously was not well detailed to permit us to make any further comments.

In conclusion, CG may complicate pregnancy, and may be associated with adverse pregnancy outcome. Pathogenetic mechanism underlying CG and relationship between other pregnancy events and CG is yet to be determined.

REFERENCES