

Pregnancy and Delivery in a Patient with CADASIL: A Case Report

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ABSTRACT

Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is an autosomal dominant arteriopathy, caused by mutations in a gene called *Notch3* on long arm of chromosome 19. It is very rare in pregnancy. This case report is presented to create awareness regarding investigating patients presenting with such illness for a better perinatal outcome.

Keywords: Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy, *Notch3*, Subcortical infarcts.

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BACKGROUND

Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is an autosomal dominant arteriopathy, caused by mutations in a gene called *Notch3* on long arm of chromosome 19. Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy is characterized by recurrent subcortical ischemic infarcts that can lead to migraine, with or without aura; cognitive problems; seizures; psychiatric symptoms; dementia; and urinary incontinence,¹ which usually occur between 40 and 50 years of age, although MRI is able to detect signs of the disease years prior to clinical manifestation of disease. The definitive test is sequencing the whole *Notch3* gene. Skin biopsies are often used for the diagnosis.² No specific treatment for CADASIL is available.

The occurrence of CADASIL in pregnancy is very rare. Our current knowledge of the disease is based on a few reports. CADASIL women are frequently considered at high risk of systemic vascular events during pregnancy and often prescribed with antithrombotic drugs. There is no agreement about the optimal supportive care and treatment regime for CADASIL in pregnancy.

CASE DESCRIPTION

We report a case of 34-year-old primigravida, second marriage, married for 9 months, who is a known case of CADASIL, bipolar affective disorder (BPAD) with history of gestational diabetes mellitus (GDM) admitted in view of uncontrolled blood sugar in August 2019 to our institution, Institute of Maternal and Child Health, Government Medical College Kozhikode, Kerala. She had history of BPAD at the age of 25 with an episode of mania and was on sodium valproate and olanzapine. Detected to have CADASIL in September 2018 and at that time she was presented with history of fever and cough for 1-week duration, had occasional episodes of headache, developed altered sensorium in the form of word output, hallucinations, and had one episode of urinary incontinence. She was evaluated with MRI, which showed bilaterally symmetrical multiple punctate T2 flair hyperintensities in subcortical and deep white matter region; and gene study proved *Notch3* gene mutation positive. Since then she was on ecosprin 75 mg daily. There was history of similar illness in brother and history of paraplegia in father. She was managed with insulin, sodium valproate, and folic acid.

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Ecosprin continued till 38 weeks. At 39 weeks, she was induced with prostaglandins in view of GDM on insulin and had a normal labor on August 16, 2019, a male baby of weight 2.8 kg. Postnatally, she continued with sodium valproate and ecosprin restarted 12 hours and injection LMWH 6 hours after delivery and given for 5 days as thromboprophylaxis. She was discharged in stable condition on postnatal day 7.

CONCLUSION

In patients with CADASIL, there are no data describing its effects on pregnancy. Donnini *et al.*³ described 50 pregnant patients with CADASIL and concluded that there have been no major adverse events during pregnancy. They noted that roughly half of the patients were on antithrombotic therapy and concluded there was no evidence supporting a specific preventive antithrombotic regimen.

Women with CADASIL commonly develop preeclampsia and neurologic symptoms while pregnant and during the postpartum period. In a recent study, 12 of 25 mothers with CADASIL confirmed by *R133C Notch3* mutation developed neurologic symptoms in their pregnancies. In 82%, the first symptoms of disease were during pregnancy.⁴ CADASIL is associated with a history of migraine⁵ and should be taken into consideration as a possible cause of headache during pregnancy, especially if there are focal neurologic symptoms. Pregnant mothers with suspected or established diagnosis of CADASIL should be monitored carefully.⁶

This case is being reported for its rarity and need for proper evaluation of personal and family history in cases with such

presentation. Our patient was evaluated due to strong family history in her first-degree relatives.

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