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Cerebral Autosomal Dominant Arteriopathy with Subcortical Infarcts and Leukoencephalopathy (Cadasil) in Pregnancy

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Abstract

Cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy (CADASIL) is a genetic condition. This is known to be caused by NOTCH3 gene mutation. These mutations cause arteriopathy of small vessels characterized by recurrent subcortical lacunar infarcts and white matter degeneration along with grey matter micro-haemorrhages. A review of studies from patients with CADASIL showed that all patients, including the asymptomatic group, had MRI alterations [1].

The prevalence of CADASIL in the UK is 4.15 per 100 000 adults. CADASIL is characterized by neurologic manifestations such as migraine with severe long lasting aura, psychiatric issues (mainly severe depression), vascular dementia, cognitive impairment and stroke. It is also noted to shorten life expectancy [2]. Lifestyle and environmental factors can change the clinical manifestations. Simple measures like improving blood pressure and HbA1c can potentially decrease the risk of haemorrhagic stroke [1,3].

Keywords: pregnancy, stroke, preeclampsia, subcortical infarcts, leukoencephalopathy, acute ischaemia.

Case Report

We present the case of a low risk 24yr old patient, G0P0, booked for midwifery led care. Ex-smoker (Stopped in pregnancy), no alcohol or drugs. She had a past medical history of frequent migraine, with photophobia and visual changes with partial visual field loss.

She underwent a post-dates induction of labour with a Foley catheter at 41+3weeks gestation, followed by ARM and syntocinon infusion the following day. During the induction process and the labour she had a borderline high BP (max 150/96mmHg) which settled without treatment.

The labour progressed normally and she delivered by vacuum extraction for pathological CTG in the same day. The vacuum delivery was not difficult but it was complicated by atonic primary PPH. She developed secondarily a hypotensive episode which lasted about 9-10 minutes. She was treated with intravenous fluids, intravenous syntocinon infusion and misoprostol. Her blood pressure initially increased briefly to 150/100 mmHg then normalised again.

She developed a secondary postpartum haemorrhage a few hours later and required carboprost and further syntocinon infusion. Her total blood loss was 2500ml and she was transfused 2 units packed red blood cells.

Day 1 – she developed a new onset of intermittent left sided sensory disturbance/weakness, headache & dysphasia. Differential diagnosis considered included Posterior Reversible Encephalopathy Syndrome, Cerebral Venous Sinus Thrombosis, haemorrhagic or ischaemic stroke and a space occupying lesion. After discussion with neurology she had a Computerized Tomography scan and angiogram of her brain which were normal.

Collateral history revealed that the patient`s father had been diagnosed with CADASIL and he had suffered strokes starting in his late 40`s. Magnetic Resonance Imaging on revealed small changes in keeping with CADASIL (ischaemic and chronic white matter changes) but no evidence of acute stroke. She was commenced on Aspirin 300mg OD and her symptoms gradually improved. This was on the basis of a specialised stroke team recommendation as per their guidance.

Day2 she developed an acute episode of agitation and became aggressive. She was reviewed by the Psychiatrist who suspected an organic cause but considered her at high risk of puerperal psychosis.

Day 3 she had a new onset of a severe headache followed by speech difficulties and a pyrexia. As there were no signs of gynaecological infection she was commenced on Tazocin empirically & 3 days of acyclovir to cover potential encephalitis and transferred to the stroke unit.

On the same day she had a tonic-clonic seizure lasting 2 min which was managed with Lorazepam, Labetalol & Keppra. The neurology team recommended an EEG which was markedly abnormal (slow wave activity over the left hemisphere) so she was transferred to ICU for more intensive monitoring. An MRI on the same day showed acute ischaemia in the left hemisphere and a CT angiogram indicated widespread vascular irregularity probably caused by vasospasm.

Day 5- She had a repeat MR angiogram suggested irregularity of the left Middle Cerebral Artery. A CT angiogram the following day showed diffuse irregularity of intracerebral vessels and a CT perfusion scan showed no perfusion which was interpreted as vasospasm. Her condition slowly improved and speech recovered over the next few days.

Day 9- The patient was transferred back to the Stroke ward and her Aspirin was reduced to 75mg. She had two LP attempts which were unsuccessful due to her agitation on day 11 and 12. A further repeat MRI/MRA on day 11 showed a new small acute infarction adjacent to the left caudate, but marked improvement of the majority of intracranial vessels. A repeat EEG also improved with no epileptiform changes. She was discharged home Day 12 on prophylactic Clexane and Aspirin for 6 weeks, having ongoing cognitive issues. The patient was reviewed 6 weeks postpartum and was considered to have made a complete recovery.

Conclusion and Discussion

CADASIL is an important cause of ischemic cerebrovascular accident in young population. Common manifestations include transient cerebral ischemic attack, decreased cognitive status, and vascular dementia. The severity is higher in symptomatic patients and increases dramatically with age.

CADASIL may potentially be a more frequent diagnosis in pregnancy and postpartum as the median age of pregnant women is gradually increasing. These patients should be managed by appropriate multi-disciplinary team comprising of experienced obstetricians within a high risk obstetric service, neurologists and anaesthetists. There may also be a role for preimplantation genetic diagnosis consideration with these patients.

There is a paucity of data about pregnancy outcome in CADASIL. One retrospective case series of 25 women with CADASIL and history of prior pregnancies reported 80% of complications including neurological symptoms and preeclampsia, mostly postpartum. The mean ages of complicated and uncomplicated pregnancies were 31.6 and 24.7 years, respectively. Hemi-paraesthesia, aphasia, hemiparesis and visual disorders were also common [1].

The prevalence of preeclampsia in CADASIL patients (10%) was greater than in normal pregnancies (3-5%) [1]. Another short study suggest that CADASIL does not seem to be associated with unfavourable pregnancy outcomes for either the woman or the fetus [4].

There are no current clinical guidelines or recommendations on antenatal care or management in labour or postpartum. There is no specific treatment for CADASIL and aspirin efficacy is unproven but it is widely prescribed. These women should receive antihypertensive treatment if indicated and positive lifestyle changes should be encouraged (e.g. stop smoking).

During the intra-partum period important considerations include care fluid balance to avoid hypovolaemia and maintaining a stable blood pressure. It`s important to maintain normocapnia during general anaesthesia. CADASIL is not a contraindication for regional anaesthesia. During the postpartum period it is safe to use NSAIDs for analgesia as well as opioids.

Potential exacerbating factors include angiography and anticoagulants along with smoking. Thrombolytic therapy (intravenous thrombolysis) is contraindicated because of the presumed increased risk for cerebral haemorrhage [5].

CADASIL is usually diagnosed on MRI (CT scan and CT angiogram might be normal) and it is important to use sequential imaging to identify changes that develop.

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