Moyamoya disease in pregnancy: maintenance of maternal blood pressure

Helen L Barrett MBBS(Hons) FRACP^{*†‡}, **Karin Lust** MBBS FRACP^{†‡}, **Narelle Fagermo** MBBS FRACP^{†‡},
Leonie K Callaway MBBS(Hons) FRACP^{†‡} and **Lee Minuzzo** MBBS FRANZCOG[§]

^{*}University of Queensland, UQ Centre for Clinical Research, Herston, Queensland, Australia, 4029; [†]University of Queensland, School of Medicine, North, Herston, Queensland, Australia, 4029; [‡]Obstetric Medicine, Internal Medicine and Aged Care Services, Royal Brisbane and Women's Hospital, Herston, Queensland, Australia, 4029; [§]Maternity Services, Royal Brisbane and Women's Hospital, Herston, Queensland, Australia, 4029

Summary: Moyamoya disease is a rare cerebrovascular occlusive disorder characterized by stenosis in the circle of Willis with the development of a compensatory circulation. It has been associated with significant morbidity in pregnancy including intracranial haemorrhage, ischaemic stroke and epilepsy. We present the case of a 26-year-old woman with a previous diagnosis of moyamoya vasculopathy with bilateral superficial temporal to middle cerebral artery bypass grafting. During the second trimester, she developed significant neurological symptoms related to postural hypotension in the presence of a stenosis of the right-sided graft. The hypotension was treated with fludrocortisone therapy with improvement in blood pressure and symptoms. Moyamoya vasculopathy poses unique challenges to obstetric care. This is the first report of use of fludrocortisone for maintenance of blood pressure during pregnancy in this condition.

Keywords: moyamoya disease, pregnancy, cerebral artery bypass grafting, hypotension, fludrocortisone

INTRODUCTION

Moyamoya disease (MMD) is a syndrome of progressive, occlusive cerebral vasculopathy involving bilateral stenosis of the internal carotid arteries, anterior and middle cerebral arteries.¹ We present the case of a 26-year-old right-handed woman with MMD who developed neurological symptoms associated with the normal pregnancy-related decline in blood pressure. This was managed with fludrocortisone.

CASE

The patient's MMD presented with an episode of visual loss, left facial, left upper-limb sensory and motor disturbance in 2009. Magnetic resonance imaging (MRI) scan demonstrated severe MMD involving the internal carotid arteries bilaterally with significant haemodynamic impairment of the right hemisphere. Initial management included aspirin, atorvastatin and bilateral superficial temporal artery to middle cerebral artery bypass surgery in February 2009, with resolution of the presenting symptoms. Prepregnancy blood pressures were 120/80 mmHg in February 2010 and 132/80 mmHg in April 2010. Past obstetric history was of one normal pregnancy with vaginal delivery in 2006, and an early miscarriage in 2010.

Family history included siblings with cutaneous lupus and type 1 diabetes, and a father with type 2 diabetes.

We first reviewed the patient at 15 weeks gestation. At that time she was taking aspirin 100 mg daily. Investigations including connective tissue screen, renal artery duplex and echocardiogram were normal. At 24 weeks of gestation, she experienced left upper-limb, intermittent left lower-limb and left facial sensory disturbance. Symptoms were exacerbated on standing. Blood pressure had declined from prepregnancy levels to ~90/50 mmHg with no postural drop. MRI showed no acute infarct and chronic white matter ischaemic changes were stable (Figure 1). On the right, there was a poor anastomosis from the superficial temporal artery to the middle cerebral artery. This stenosis was present on previous imaging.

It was felt that the symptoms were most likely due to blood pressure decline, right-sided stenosis and inability to autoregulate cerebral circulation. Fludrocortisone was commenced at 50 µg daily and titrated to 150 µg daily over a week, combined with increased oral fluid intake. Systolic blood pressure improved by ~10–15 mmHg (Figure 2). The neurological symptoms improved but remained intermittent for the rest of the pregnancy. Mild peripheral oedema developed. Electrolytes remained normal.

Induction of labour was performed at 38 weeks with dinoprostone. Labour was managed with combined epidural/spinal analgesia. The patient noted intermittent left facial and arm sensory disturbance during Valsalva. Delivery was assisted with ventouse and episiotomy to reduce maternal effort. Blood pressure was monitored during labour and for four hours post-delivery via an arterial line, and remained stable throughout. The infant was well.

Correspondence to: Dr Helen Barrett, Department of Internal Medicine, Level 3, James Mayne Building, Royal Brisbane and Women's Hospital, Butterfield Street, Herston, Queensland, Australia, 4029
Email: Helen_Barrett@health.qld.gov.au

Figure 1 Magnetic resonance image showing the abnormal cerebral vasculature, with narrowing of several of the major vessels and the 'puff of smoke' appearance more distally. There are bilateral superficial temporal to middle cerebral artery bypass grafts. The white circle shows the site of poor anastomosis from the superficial temporal artery to the middle cerebral artery on the right side

The patient was initially well postpartum, fludrocortisone was weaned over the following four weeks to 50 µg/day, but upon cessation the neurological symptoms experienced during the pregnancy recurred, and the patient developed headaches. The fludrocortisone was restarted at 50 µg/day with improvement, although intermittent symptoms have continued. It has been ceased four months later, with blood pressure and symptoms remaining stable at the present time. The patient is undergoing further evaluation.

DISCUSSION

Moyamoya, in Japanese, means 'puff of smoke'. This label arose because of the characteristic radiological appearance of the cerebral arteries due to the proliferation of small perforating vessels. MMD can result in both ischaemic events and intracranial haemorrhages. The Ministry of Health and Welfare in Japan has defined four types of MMD: ischaemic (63.4%), haemorrhagic (21.6%), epileptic (7.6%) and 'other' (7.5%).² The highest rate is in Japan with an incidence of 0.54/100,000/year,

prevalence 6/100,000.³ In a Californian study, incidence varied by ethnicity: Caucasians 0.06/100,000 per year, Asian American 0.28, African American 0.13, Hispanic 0.03.⁴ There is a female preponderance (female:male ratio ~1.8-2.7:1).^{3,5} Children primarily present with ischaemic events and adults haemorrhagic events. Those diagnosed with MMD in America (excluding immigrants from Japan) have a greater prevalence of ischaemic events and more benign course than the Japanese cohorts.⁶ Most cases of Moyamoya are sporadic. In Japan, ~12% are familial with an autosomal-dominant inheritance.³ Genomewide linkage studies have suggested associations with chromosome 17q25, chromosomes 3 and 6.⁷⁻¹⁰

MOYAMOYA IN PREGNANCY

There are many sporadic reports of undiagnosed MMD presenting with haemorrhage or stroke in pregnancy, often in the setting of preeclampsia or eclampsia. It is not clear that pregnancy itself alters the risk of intracerebral events, but there are suggestions that preeclampsia may. There are two large reviews: the first in 1998 by Komiyama found 23 cases where MMD was diagnosed during pregnancy – these presented with cerebral haemorrhage or ischaemia. Three of the 23 women died and eight had ongoing neurological impairment. By contrast, those who were diagnosed with MMD before pregnancy – 31 case reports, had good outcomes with only one adverse event.¹¹ A nationwide survey of 280 centres in Japan, reporting MMD outcomes over five years found 64 deliveries, 59 to women with previously diagnosed MMD (58% had extra-cranial-intracranial bypass surgery before pregnancy) and five newly diagnosed.¹² There was only one case of cerebral haemorrhage with poor outcome among those with known MMD.¹²

Cerebral autoregulation remains intact in pregnancy.¹³ This autoregulation is impaired in MMD. Presumably, while the cerebral blood flow improves post bypass surgery, the autoregulatory mechanisms will not be restored by a direct extra-cranial-intracranial bypass. Severe hypertension as with preeclampsia, hypotension, hyper- and hypocapnia (as with hyperventilation during labour) may impair cerebral perfusion and autoregulation.^{14,15}

Figure 2 Graph of maternal blood pressure during pregnancy and early postpartum period

Reported mode of delivery in MMD is primarily by caesarean section: 76% in the Japanese nationwide survey.¹² No adverse events were reported in that survey among women with known MMD, or with extracranial-intracranial bypass surgery regardless of delivery mode. There are rare reports of intracranial haemorrhage during vaginal delivery in women not previously diagnosed.¹²

Both general and neuraxial anaesthesia have been used successfully in MMD. Neuraxial techniques appear safe and have the advantage of allowing neurological assessment but risk haemodynamic instability. Most reports mention arterial blood pressure monitoring and use of blood pressure support with fluids or inotropes.¹⁶

CONCLUSIONS

We present the first report of fludrocortisone use during pregnancy to maintain blood pressure and hence cerebral perfusion in a woman with an extracranial-intracranial bypass for MMD disease. MMD disease presents unique challenges for management in pregnancy with blood pressure maintenance being paramount. MMD should be considered in women with intracranial haemorrhage or infarct during pregnancy. Maternal outcomes in previously diagnosed MMD are usually good. The use of fludrocortisone in this case ameliorated the neurological symptoms induced by pregnancy-related hypotension, allowing avoidance of neurosurgery and continuation of the pregnancy.

DECLARATIONS

The authors have no conflicts of interest to declare. The patient has given her permission for the publication of this case study.

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