Occlusive Cerebrovascular Disease with Moyamoya Vessels and Intracranial Hemorrhage during Pregnancy

—Case Report and Review of the Literature—

Kazuhito HASHIMOTO, Kiyotaka FUJII, Kenichi NISHIMURA,
Michihiro KIBE* and Takashi KISHIKAWA**

*Departments of Neurosurgery and **Radiology, Saga Medical School, Saga;*

**Department of Neurosurgery, Fukuoka University, School of Medicine, Fukuoka*

Abstract

A 24-year-old female in the sixth month of pregnancy developed headache and vomiting caused by an intraventricular hemorrhage from moyamoya vessels. Superficial temporal artery-middle cerebral artery bypass surgery was performed when she was in the seventh month of pregnancy in order to prevent intracranial rebleeding or a cerebrovascular ischemic attack during labor and delivery. The patient delivered successfully at full term, without complications. The clinical features and management of patients with such rare occurrences during pregnancy are discussed and seven previously reported cases are reviewed.

Key words: moyamoya disease, intracranial hemorrhage, pregnancy, STA-MCA bypass surgery

Introduction

The incidence of moyamoya disease is slightly higher in females than in males (1 : 1.4),⁴⁾ and young persons are most often affected. Nishimoto *et al.*¹⁶⁾ reported that the frequency of recurrent intracranial hemorrhage increases as moyamoya patients grow older. In their follow-up of 271 cases, they found a mortality rate of 22% among individuals with hemorrhagic moyamoya disease.

Cerebrovascular accidents due to moyamoya disease or "moyamoya phenomenon" of unknown etiology is an uncommon complication of pregnancy. In the future, however, neurosurgeons will encounter increasing numbers of pregnant women with moyamoya disease. In these cases the neurosurgeon and obstetrician must cooperate in formulating the treatment plan, and priority must be established between the delivery and the treatment of a cerebrovascular accident, should one occur during the pregnancy. We encountered such a case, in which we performed superficial temporal artery-middle cerebral artery (STA-MCA) anastomosis to maximize

the chance of a successful delivery.

Case Report

A 24-year-old, right-handed Japanese housewife in the sixth month of pregnancy was hospitalized on July 8, 1982, following sudden onset of severe occipital headache, nausea, and vomiting. On July 12, she was transferred to the neurosurgical unit of Saga Medical School Hospital. She had had renal disease in childhood but the details were unavailable. There was no history of heart disease, drug abuse, migraine, or seizures. The family history was negative.

On admission, her vital signs and general condition were normal. Her blood pressure was 120/74 mmHg. She was alert and well oriented to time and place. Occipital pain continued and moderate nuchal rigidity developed. Pupillary reactions, optic fundi, and ocular movements were normal. The cranial nerves were intact. No motor or sensory abnormalities were noted.

The erythrocyte sedimentation rate was 6 mm/hr. Hematological examination revealed slight anemia but coagulation, including bleeding time, was normal.

Computed tomography (CT) performed on the

Fig. 1 CT scan showing a high-density area in the ventricular system, predominantly in the left lateral ventricle.

day of the attack demonstrated a high-density area in the left lateral ventricle, but there was neither a shift of the midline structures nor enlargement of the ventricle (Fig. 1). Bilateral carotid and left vertebral angiograms were obtained on July 15. The left internal carotid angiogram showed the middle cerebral artery to be occluded at its origin, and there was evidence of a limited moyamoya network in the left basal ganglionic area. The branches of the left middle cerebral artery were slightly delineated *via* the enlarged anterior temporal artery, anterior choroidal artery, and basal moyamoya vessels (Fig. 2A, B). The late phase of the left internal carotid angiogram revealed the branches of the left middle cerebral artery *via* the leptomeningeal anastomoses from the left anterior cerebral artery (Fig. 2C). The right inter-

nal carotid angiogram disclosed no stenosis or occlusion. On the left vertebral angiogram, a portion of the left middle cerebral artery was visualized *via* the leptomeningeal anastomoses (Fig. 3).

On August 3, 1982, the patient underwent anastomosis of the left superficial temporal artery and the cortical branch of the left middle cerebral artery *via* a left parietotemporal craniotomy. The fetal heartbeat was monitored during the operation, which was performed under general anesthesia. The patient's intraoperative systolic blood pressure ranged from 90 to 120 mmHg. Diuretics and osmotic agents were used as little as possible to avoid dehydration of the fetus.^{10,14} Both patient and fetus endured the procedure well and the patient was discharged on August 14 without neurological deficits.

The pregnancy proceeded uneventfully and the patient was admitted on November 15 for delivery. On November 17 she went into spontaneous labor, which proceeded rapidly and uneventfully to complete cervical dilatation. The right radial artery was catheterized for blood pressure monitoring. A vacuum delivery was performed under epidural anesthesia to avoid a rise in blood pressure through abdominal muscular tension. During the delivery her systolic blood pressure ranged from 90 to 140 mmHg. The infant was a female with a weight of 2726 gm and an Apgar score of 9. The patient had no neurological problems subsequent to the delivery.

On November 29, common carotid angiography was performed. The branches of the left middle cerebral artery were filled through the left superficial temporal artery, which decreased the size of the moyamoya network in the left basal ganglionic area

Fig. 2 Left internal carotid angiogram showing occlusion of the origin of the left middle cerebral artery with evidence of a mild moyamoya network in the basal ganglionic area. Note the branches of the left middle cerebral artery delineated *via* the enlarged anterior choroidal artery, anterior temporal artery, and moyamoya vessels (A, B). The late phase demonstrates the left middle cerebral artery *via* the leptomeningeal anastomoses from the left anterior cerebral artery (C).

Fig. 3 Left vertebral angiogram showing the branches of the left middle cerebral artery *via* the enlarged left parieto-occipital artery.

Fig. 4 Left common carotid angiogram performed 4 months after STA-MCA bypass surgery. The branches of the left middle cerebral artery are well filled through the superficial temporal artery.

(Fig. 4). On December 1, 1982, the mother and child were discharged with no general or neurological problems. The patient has had no further hemorrhagic or ischemic attacks during the 4 years since the operation.

Discussion

A cerebrovascular accident due to moyamoya disease or "moyamoya phenomenon" is a rare complication of pregnancy; only seven such cases have been reported.^{2,3,7,14,17,21} Ours, however, is the first report of "moyamoya phenomenon" in pregnancy in which STA-MCA bypass surgery was performed in an effort to avoid a recurrent cerebrovascular event during delivery.

The diagnosis of moyamoya disease is made on the basis of angiographic findings of intracranial carotid arterial stenosis or occlusion and the presence of a fine network of vessels, frequently bilateral, at the base of the brain.²⁰ "Moyamoya phenomenon," which involves numerous dilated collaterals of the basal perforating arteries and arterioles, is often a disease of the internal carotid fork whose etiologies include von Recklinghausen's disease,¹² arteriosclerosis,⁵ tuberculous meningitis,¹³ sickle cell anemia,¹¹ radiation therapy,¹⁸ and oral contraceptive use.⁶ None of these is likely to be the etiology in our patient.

A summary of symptoms and signs before and during pregnancy in the seven reported cases (as well as our own) is given in Table 1. Four patients had manifestations of cerebrovascular ischemia, which seemed to subside during pregnancy. On the other hand, four patients (including ours) who had intracranial hemorrhages during pregnancy were asymptomatic prior to their pregnancies. All of the intracranial hemorrhages occurred in and after the second trimester. Thus, the time of onset of intracranial hemorrhage may be related to hemodynamic changes of pregnancy.¹¹ In the four patients with the cerebrovascular ischemic type of disease, the symptoms preceded pregnancy, and countermeasures were taken prior to delivery. However, it may be difficult to diagnose the intracranial hemorrhagic type of moyamoya disease before the onset of symptoms and signs. Therefore, in these cases it may be unavoidable that the treatment plan be decided after pregnancy is well established.

Three causative factors have been proposed concerning the source of intracranial hemorrhage in moyamoya disease. One is a breakdown of the greatly dilated perforating moyamoya vessels that form the extensive basal collateral network.¹⁵ The second is a pseudoaneurysm, which is angiographically identified as a discrete vascular dilatation, usually in the paraventricular region and typically arising from the peripheral portions of the perforating arteries or the anterior and posterior choroidal arteries.^{9,19} The last is a saccular aneurysm arising from the circle of Willis, a feature associated with moyamoya disease.^{9,15,19}

In our case, the CT scan suggested that the hemorrhage was located in the paraventricular area and that the hematoma had penetrated the lateral ventricle. We found no angiographic evidence of aneurysms in our patient, whereas aneurysms were demonstrated in the seven previously reported cases, but we cannot rule out the possibility that there were small aneurysms or pseudoaneurysms near the wall

Table 1 Summary of eight cases of moyamoya disease or "moyamoya phenomenon" in pregnancy

Case No.	Author (Year)	Age (years)	Parity	Symptoms and signs		Angiographical diagnosis
				Before pregnancy	During pregnancy	
1	Matsuoka <i>et al.</i> (1976) ¹⁴⁾	24	primipara	TIA: weakness of extremities, speech disturbance, dysesthesia, headache, syncope	dysesthesia, nausea	moyamoya disease
2	"	34	primipara	episode of unconsciousness, headache, nausea, vomiting, general convulsion	lt. hemiparesis and hemisensory disturbance	moyamoya disease
3	Takagi <i>et al.</i> (1977) ²¹⁾	35	primipara	none	ICH (10 hours after delivery): unconsciousness, general convulsion, miosis, pyramidal signs, rt. homonymous hemianopsia	moyamoya disease
4	Fujita <i>et al.</i> (1978) ³⁾	30	primipara	none	ICH (in 7th month of gestation): headache, vomiting, unconsciousness, lt. hemiparesis, convulsion	moyamoya disease with intracerebral hematoma in rt. frontal lobe
5	Karasawa <i>et al.</i> (1978) ⁸⁾	27	secundipara	unknown	ICH (in 9th month of gestation): nausea, vomiting, rt. hemiparesis, consciousness disturbance	moyamoya disease (CT scan: intraventricular hemorrhage)
6	Bingham <i>et al.</i> (1980) ²⁾	26	unknown	lt. hemiparesis and hemiparesthesia, headache, progressive scoliosis	unchanged	moyamoya disease
7	Sasaki and Mezaki (1984) ¹⁷⁾	30	primipara	TIA: lt. hemiparesis and hemidysesthesia, syncope	rt. sided migraine	"moyamoya phenomenon" (rt.)
8	Present case	24	primipara	none	ICH (in 6th month of gestation): headache, vomiting	"moyamoya phenomenon" (lt.)

TIA: transient ischemic attack, ICH: intracranial hemorrhage.

of the lateral ventricle.¹⁹⁾ Follow-up angiography is advised in such cases.⁹⁾

The neurosurgical and obstetric procedures and outcomes of the seven reported cases and our own are summarized in Table 2. For patients with the cerebrovascular ischemic type of moyamoya disease the outcome was generally favorable. Two of these patients' infants had congenital malformations — one a cleft lip and the other osteogenesis imperfecta.¹⁴⁾ Since little is known about the relationships between these malformations and anticonvulsant medication or moyamoya disease itself, administration of anticonvulsants to pregnant women should be undertaken cautiously.

In the women with the intracranial hemorrhagic type of moyamoya disease the outcomes varied. The limited experience indicates that patients with severe moyamoya disease or "moyamoya phenomenon" who develop intracranial hemorrhage and for whom the prognosis would be poor without surgery should undergo evacuation of the clots, external ventricular drainage, or other surgical intervention. In cases of intracranial hemorrhage due to the rupture of a sacular aneurysm, direct surgery should be performed

Table 2 Treatment and outcome of in eight cases of moyamoya disease or "moyamoya phenomenon" in pregnancy

Case No.	Treatment	Delivery		Outcome	
		Procedure (gestation)	Anesthesia	Mother	Baby
1	medication only	cesarean (40 w)	general	good	cleft lip
2	medication only	cesarean (40 w)	general	good	osteogenesis imperfecta
3	medication only	cesarean (40 w)	spinal	fair	good
4	rt. frontal lobectomy and evacuation of blood	abortion		poor (akinetism)	died
5	unknown	vacuum (35 w)	epidural	good	unknown
6	no surgery	forceps (?)	pudendal	good	good
7	medication only	cesarean (38 w)	spinal	good	good
8	lt. STA-MCA bypass	vacuum (39 w)	epidural	good	good

STA-MCA: superficial temporal artery-middle cerebral artery.

regardless of the stage of pregnancy.¹⁰⁾ It must be remembered, however, that most aneurysms are located in the vertebrobasilar system, and the presence of moyamoya vessels may limit the field of exposure and increase the risks of ischemic complications.¹⁵⁾ If a patient with a massive hemorrhage is deeply comatose with faltering vital signs and the fetus is not expected to survive, therapeutic abortion may be the only option (Table 2, Case 4³⁾).

When a patient has recovered from an acute cerebrovascular event, attention should be directed to ensuring a safe delivery. If the fetus is sufficiently mature, early induced labor should be considered.¹⁸⁾

At present there is no definitive treatment for moyamoya disease, and the indications for cerebral revascularization have not yet been firmly established. We performed STA-MCA bypass surgery on our patient for the purpose of decreasing the stress on the moyamoya vessels, which were functioning as collaterals to the cerebral circulation.^{8,16)} The surgery was undertaken at what we judged to be the most suitable time, *i.e.*, three months before the expected date of the delivery, which is usually long enough for a STA-MCA bypass to become a good collateral.

Among the reported cases and ours, cesarean sections were performed in four and vaginal deliveries were accomplished under general and spinal anesthesia in two cases each. Assisted cephalic deliveries were necessary in three instances, under epidural anesthesia in two cases and pudendal anesthesia in one.

Sasaki and Mezaki¹⁷⁾ emphasized that cesarean section under general anesthesia is an effective means of avoiding hyperventilation-induced cerebral ischemia or hypertension provoked by active labor. An assisted cephalic delivery with good pain control is also possible if the birth canal is flexible enough for the infant's passage and the labor proceeds rapidly, as occurred in our case.¹⁷⁾

One reported case (Case 3) is noteworthy in that the patient had an intracranial hemorrhage due to markedly fluctuating blood pressure just after undergoing cesarean section under spinal anesthesia.²¹⁾ Kubota *et al.*¹⁰⁾ suggested that in such a case a nearly full-term fetus may be delivered by cesarean section under lumbar epidural anesthesia just prior to intracranial operation under general anesthesia, provided that the mother's blood pressure be kept low. Whichever method of delivery is selected, it is important to control the pregnant patient's blood pressure, both during and after pregnancy.

Acknowledgment

The authors wish to thank Miss Junko Narikiyo for her secretarial assistance.

References

- 1) Amias AG: Cerebral disease in pregnancy. *J Obstet Gynaec Brit Comm* 77: 100-120, 1970
- 2) Bingham WF, Beguin EA, Ramirez-Lessepas M: Moyamoya disease in pregnancy. *Wis Med J* 79: 21-25, 1980
- 3) Fujita K, Yamasaki S, Tamaki N, Fujita S, Shirakata S, Matsumoto S: Cerebro-vascular accident during pregnancy. *No Shinkei Geka* 6: 989-995, 1978 (in Japanese)
- 4) Handa H, Yonekawa Y: Date filling of 1500 cases with spontaneous occlusion of the circle of Willis ("moyamoya" disease). Including the results of long-term follow-up of 200 cases. *No Socchu* 7: 477-480, 1985 (in Japanese)
- 5) Hinshaw DB, Thompson JR, Hasso AN: Adult arteriosclerotic moyamoya. *Radiology* 118: 633-636, 1976
- 6) Iivanainen M, Vuolio M, Halonen V: Occlusive disease of intracranial main arteries with collateral networks (moyamoya disease) in adults. *Acta Neurol Scand* 49: 307-322, 1973
- 7) Karasawa J, Kikuchi H, Furuse S: Subependymal hematoma in "moyamoya" disease. *Surg Neurol* 13: 118-120, 1980
- 8) Karasawa J, Kikuchi H, Furuse S, Kawamura J, Sakaki T: Treatment of moyamoya disease with STA-MCA anastomosis. *J Neurosurg* 49: 679-688, 1978
- 9) Kodama N, Mineura K, Suzuki J, Kitaoka T, Kurashima Y, Takahashi S: Cerebrovascular moyamoya disease associated with aneurysm at the peripheral portion of the posterior choroidal artery. *No Shinkei Geka* 4: 985-991, 1976 (in Japanese)
- 10) Kubota M, Yamaura A, Nishiyama H, Makino H: Intracranial hemorrhage during pregnancy. *Neurol Med Chir (Tokyo)* 25: 640-644, 1985 (in Japanese)
- 11) Kurt HHM: Cerebrovascular disease in sickle cell anemia. A clinical, pathological and radiological correlations. *Stroke* 9: 45-52, 1978
- 12) Lamas E, Diez Lobato R, Cabello A, Abad JM: Multiple intracranial occlusions (moyamoya disease) in patients with neurofibromatosis. One case report with autopsy. *Acta Neurochir (Wien)* 45: 133-145, 1978
- 13) Mathew NT, Abraham J, Wandy J: Cerebral angiographic features in tuberculous meningitis. *Neurology (Minneapolis)* 20: 1015-1023, 1970
- 14) Matsuoka M, Kamata T, Okuba H: Pregnancy and delivery of the patient with moyamoya disease. *Sanfujinka Chiryō* 33: 572-575, 1976 (in Japanese)
- 15) Nagamine Y, Takahashi S, Sonobe M: Multiple

- intracranial aneurysms associated with moyamoya disease. Case report. *J Neurosurg* 54: 673-676, 1981
- 16) Nishimoto A, Ueta K, Onbe H: Cooperative study on moyamoya disease in Japan, in Kawafuchi J, Nukui H (eds): *Proceedings of the 10th Japanese Conference on Surgery of Cerebral Stroke (Gunma)*. 1981, pp 53-58
- 17) Sasaki J, Mezaki N: Considerations concerning the management of pregnancy and delivery in "moyamoya" disease. *Sanka To Fujinka* 51: 109-116, 1984 (in Japanese)
- 18) Servo A, Puranen M: Moyamoya syndrome as a complication of radiation therapy. *J Neurosurg* 48: 1026-1029, 1978
- 19) Suzuki J, Komada N: Moyamoya disease. A review. *Stroke* 14: 104-109, 1983
- 20) Suzuki J, Takaku A: Cerebrovascular "moyamoya" disease: Disease showing abnormal net-like vessels in base of brain. *Arch Neurol (Chicago)* 20: 288-299, 1969
- 21) Takagi T, Ushimaru N, Hirata M: A case of "moyamoya" disease found out after cesarean section. *Sanka To Fujinka* 44: 949-954, 1977 (in Japanese)

Address reprint requests to: K. Hashimoto, M.D., Department of Neurosurgery, Saga Medical School, Sanbonsugi, Nabeshima-cho, Saga 840-01, Japan.