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Case Report/รายงานผู้ป่วย

Acute Bilateral Visual Loss in Moyamoya Disease after COVID–19 Vaccination: A Case Report

รายงานผู้ป่วยตามัวลงทั้งสองข้างในโรคโมยาโมยาหลังฉีดวัคซีนป้องกันโรคโควิด–19



Orn Tempatarachoke, MD[1]
อร เต็มภัทราโชค, พ.บ.[1]

Abstract

Purpose: To report a Moyamoya patient who presented with acute bilateral visual loss after covid – 19 vaccination.

Case presentation: A 39-year-old Thai man presented with bilateral visual loss three days after the first dose of COVID-19 vaccination (inactivated vaccine - Coronavac or Sinovac). Eye examination revealed total visual field defects in both eyes. The MRI showed acute infarction in the left occipital cortex and old infarction in the right occipital cortex and other areas of the cerebral hemisphere. MRA showed severe narrowing of both ICAs and both MCAs, with collateral supplied with forming a puff of smoke appearance, representing Moyamoya's disease.

Conclusion: Acute bilateral visual loss caused by occipital infarction in Moyamoya's disease after COVID–19 vaccination. It may be a coincidence or triggered by COVID-19 vaccine, Corona Vac.

Keywords: bilateral visual loss, Moyamoya disease, COVID-19 vaccination

[1] *Department of ophthalmology, Samutprakarn Hospital 71 Chakkaphak road, Pak num, Mueang Samutprakarn, Samut Prakarn, Thailand 10270*
Tel : 081-8685905, 02-7018132-9
Email address: orncha@hotmail.com
[1] *แผนกจักษุวิทยา โรงพยาบาลสมุทรปราการ 71 ถนนจักกะพาก ตำบลปากน้ำ อำเภอเมืองสมุทรปราการ จังหวัดสมุทรปราการ 10270*
โทร : 081-8685905, 02-7018132-9

บทคัดย่อ

วัตถุประสงค์: เพื่อรายงานเคสผู้ป่วยโรคโมยาโมยาที่มีอาการตามัวลงทั้งสองข้าง หลังจากได้รับวัคซีนป้องกันโรคโควิด–19

รายงานผู้ป่วย: ผู้ป่วยชายอายุ 39 ปี มาด้วยอาการตามัวลง 2 ข้างหลังได้รับวัคซีนป้องกันโรคโควิด–19 เข็มแรก (ซิโนแวค) 3 วัน ตรวจพบว่าผู้ป่วยสูญเสียการมองเห็น ทั้ง 2 ข้าง ลักษณะเป็น cortical blindness ผลการตรวจ MRI brain มีสมองขาดเลือดจากหลอดเลือดในสมองตีบเกิดขึ้นใหม่ที่ตำแหน่ง left occipital cortex และสมองขาดเลือดจากหลอดเลือดตีบที่เกิดมานานแล้วที่ตำแหน่ง right occipital cortex และตำแหน่งอื่นๆในสมอง MRA แสดงให้เห็นว่ามีการตีบแคบของ internal carotid artery และ middle carotid artery ทั้งสองข้าง ร่วมกับมีหลอดเลือดฝอยผิดปกติคล้ายหมอกควัน เข้าได้กับโรคโมยาโมยา ผู้ป่วยได้รับการรักษาตามอาการ และหลังติดตามอาการ 1 เดือนผู้ป่วยมีลานสายตาดีขึ้น

สรุป: อาการตามัวในผู้ป่วยโมยาโมยารายนี้เกิดจากหลอดเลือดในสมองตีบตำแหน่ง occipital cortex และ visual pathway การได้รับวัคซีนป้องกันโรคโควิด-19 อาจเป็นตัวกระตุ้นให้เกิดการตีบหรือหลอดเลือดหดตัว หรือเป็นสิ่งที่บังเอิญเกิดขึ้นร่วมกันก็เป็นได้

คำสำคัญ: bilateral visual loss, Moyamoya disease, COVID-19 vaccination

Introduction

Thailand began vaccination against the COVID-19 (Coronavirus disease) infection with the inactivated vaccine (Coronavac or Sinovac) on February 28, 2021. This was later followed by the AZD1222 (AstraZeneca) vaccine on June 7, 2021, and then messenger RNA (mRNA)-based vaccine BNT162b2 (Pfizer/BioNTech) in August 2021.

Multiple side effects have been reported, ranging from mild symptoms, such as swelling and redness at the injection site, low-grade fever, nausea, vomiting, and fatigue, to more severity, including high fever, chills, anaphylactic shock, seizures, and loss of consciousness.[1] In addition, the reports about ophthalmological complications of COVID-19 vaccination also arose. The ocular adverse effects include optic neuritis, facial palsy, cranial nerve palsies including third, fourth, and sixth nerve palsies, acute macular neuroretinopathy, central serous retinopathy, uveitis, multiple evanescent white dot syndrome, Vogt-Koyanagi-Harada disease reactivation, etc.[2,3] There were the reported cases of bilateral transient visual field defect or stroke may be triggered by COVID - 19 vaccine.[4,5] However, there is not enough information on the side effects that may occur following vaccination, is just coincidental or really related to the vaccine.

Moyamoya disease (MMD) or "spontaneous occlusion of the circle of Willis" was first reported in 1969 by Suzuki and Takaku.[6,7] MMD is a rare, chronic progressive disorder of the arteries in the brain. The terminal portion of the internal carotid artery becomes narrow or obstructed and produces abnormal collateral vessels at the base of the brain. Inadequate blood supply can cause ischemic stroke, hemorrhagic stroke, or transient ischemic attacks (TIA). Patients often present with symptoms of limb weakness, hemiparesis, headache, dysarthria, aphasia, cognitive impairment, and seizures. MMD is often diagnosed in children in the first decade, or in adults in their 40s. Females have a higher risk. This arteriopathy occurs on an idiopathic or familial. Many studies show genetic links in Asia.

Suzuki and Takaku classified MMD into 6

stages[7] based on the sequential changes in cerebral angiograms. Stage 1: narrowing of the carotid fork. Stage 2: initiation of Moyamoya collaterals. Stage 3: progressive ICA stenosis with the intensification of Moyamoya collaterals at the base of the brain. Stage 4: minimization of Moyamoya, anterior and middle cerebral artery disappear. Stage 5: reduction of Moyamoya, increase the ECA collaterals. Stage 6: total occlusion of ICA and Moyamoya collaterals disappear, and the brain is supplied from the external carotid or vertebral arterial system.

Moyamoya can be classified as Moyamoya syndrome and Moyamoya disease.[9,11] Patients with the characteristic Moyamoya vasculopathy who also have associated conditions such as Down's syndrome, Neurofibromatosis type I, sickle cell anemia, cranial irradiation, etc. are categorized as the Moyamoya syndrome, whereas patients without associated risk factors are said to have Moyamoya disease. Treatment involves managing symptoms with medicine or improving brain circulation by surgery.

This article aims to report a case of Moyamoya disease with acute bilateral visual loss a few days after vaccination against COVID-19.

Case report

A 39-year-old man presented with acute bilateral visual loss, three days after he received the first dose Sinovac vaccine. The patient was a motorcycle taxi driver who had diabetes mellitus and systemic hypertension. He did not have a history of previous neurological deficits and cardiovascular events. He was not obese and stopped smoking 10 years ago. he did not have a history of COVID-19 infection.

The patient was investigated on the fifth post-vaccination day. Ophthalmological examination revealed visual acuity of hand motion in both eyes. Intraocular tensions were normal. Normal pupillary reaction, clear cornea, and clear lens were manifested bilaterally. Fundus examination showed normal macula and no diabetic retinopathy. the optic disc had clear margin, and neither edema nor pallor was shown. The computerized visual field (CTVF 24-2) appeared totally visual field defect in both eyes (Figure 3). Other neurological deficits were not manifested. Fasting blood sugar was 349 mg%, blood pressure 145/94 mmHg and the syphilis screening was negative. The hemogram was normal.

Brain magnetic resonance imaging (MRI), which was performed on the seventh post-vaccination day, showed multi-stage infarction at both cerebral hemispheres. Acute infarctions at left occipital cortex. Gliotic and encephalomalacia changed at the right temporal lobe, right parietal lobe, right occipital lobe, and left frontal lobe, representing old infarctions. (Figure 1)

Brain magnetic resonance angiography (MRA) showed severe narrowing at cavernous-supraclinoid parts of the right internal carotid artery (ICA), supraclinoid part of left ICA, and M1 segment of both middle cerebral arteries (MCAs), with extensive collateral supplied forming puff of smoke appearance, representing Moyamoya disease. (Figure 2)

The patient was diagnosed with cortical vision loss secondary to occipital stroke. He was hospitalized for a week, under supportive treatment, mainly with aspirin 300 mg and diabetes control. His baseline cortical visual loss persisted, and he still had visual

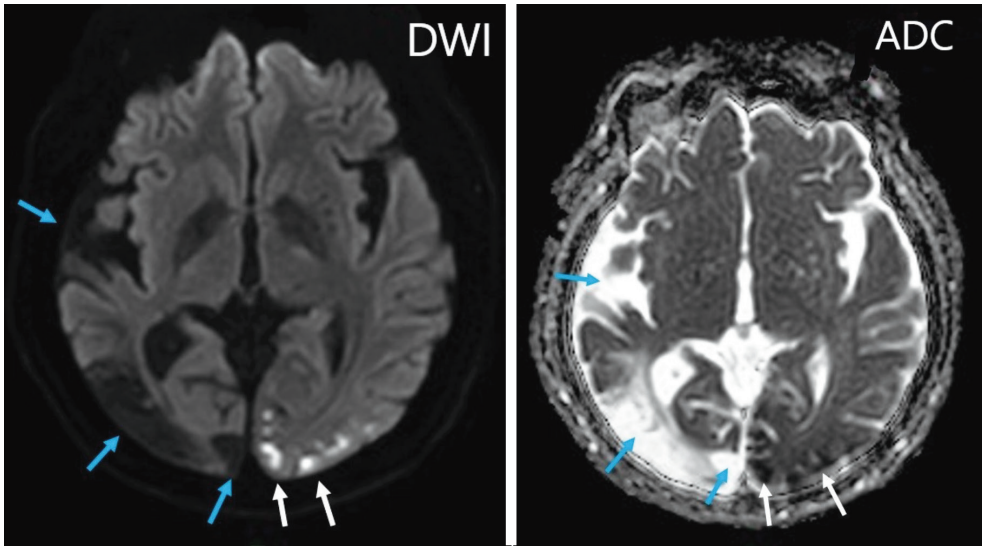


Figure 1 MRI (DWI and ADC): acute infarction at Lt occipital cortex (white arrow). Gliotic and encephalomalacia changed at right temporal lobe and right occipital lobe, representing old infarctions (blue arrow).

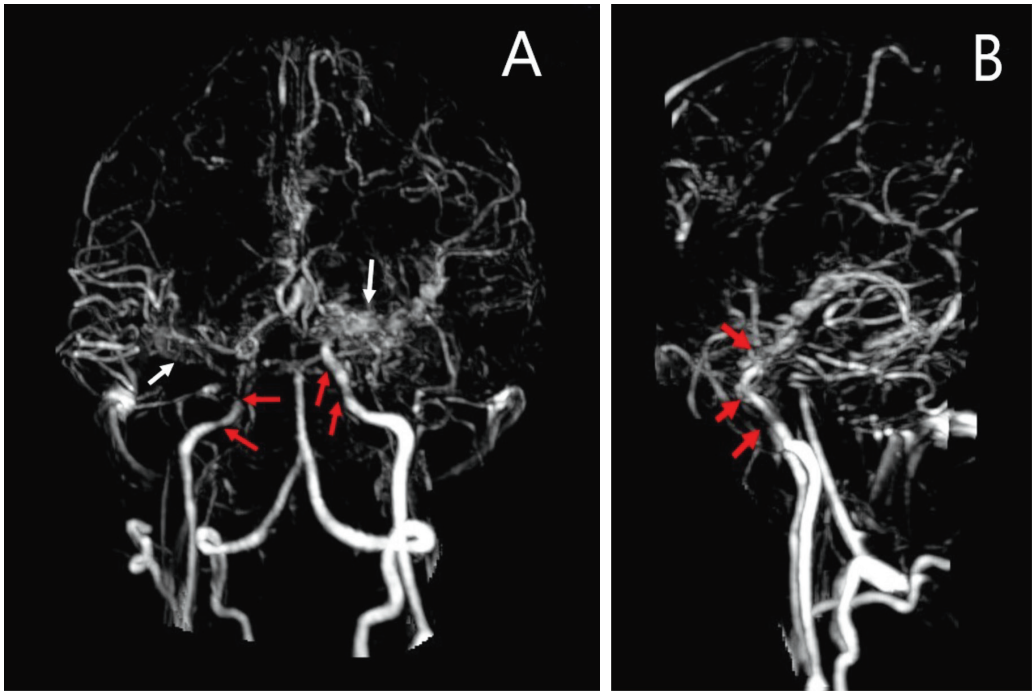


Figure 2 MRA: severe stenosis at bilateral distal internal carotid artery (red arrows) and proximal both middle cerebral arteries. White arrows show extensive collateral vessels.

acuity of hand motion until discharge. He was given aspirin 300 mg, donepezil 5 mg, antihypertensive and diabetic drugs after discharge.

One month later, the vision improved significantly to 20/160 in the right eye and 20/80 in the left eye. CTVF showed bilateral left homonymous hemianopia (Figure 4) the patient rejected the second dose of COVID-19 vaccination and lost follow-up.

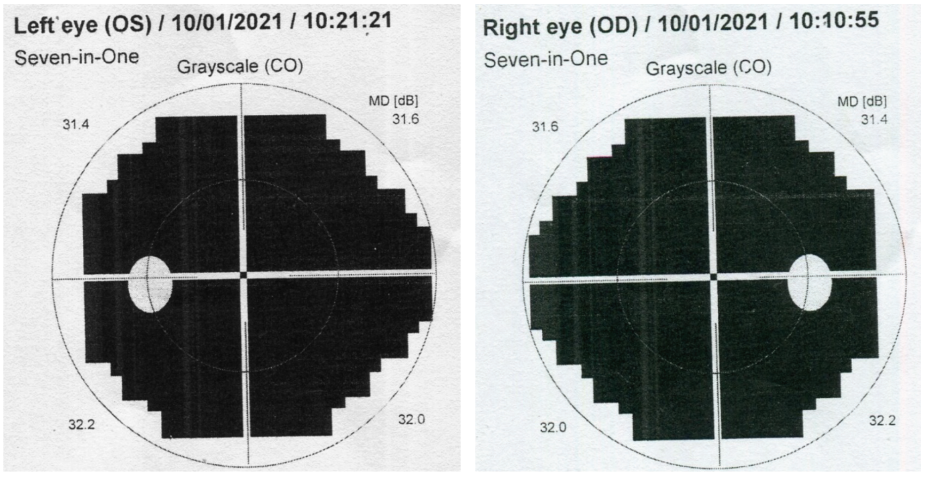


Figure 3 visual field (CTVF 24-2) show total defect in both eyes at first visit.

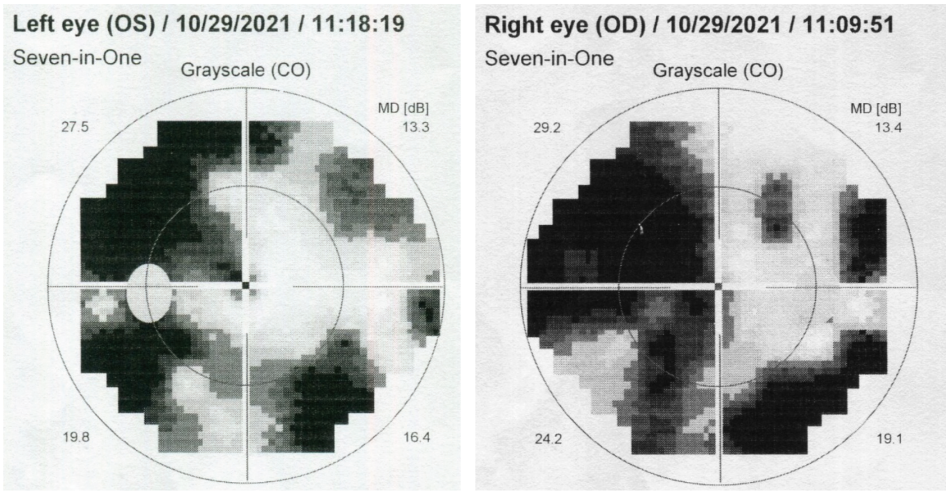


Figure 4 visual field (CTVF 24-2) show improvement at 29 days later (look-like left homonymous incongruous hemianopia and right superior- inferior quadrantanopia).

Discussion

This article reports a patient with Moyamoya disease who had bilateral occipital infarctions resulting in bilateral visual loss after COVID-19 vaccination. Previously, the patient did not have any neurological disorder. Although the old infarction occurred in the right temporal, parietal and occipital cortex causing left homonymous hemianopia, the patient did not aware of his visual problem. Until ischemia in the left occipital lobe leads to loss of vision in the half of the visual field.

"Moyamoya" is a Japanese expression for something hazy just like a puff of cigarette smoke drifting in the air, because the disease produces abnormal net-like vessels at the base of the brain. MMD is a very rare disease, the prevalence appears about 1 per million in Europeans. In Asia, it is more common, especially in Japan and Korea. In Japan, it is found about 1 per 200,000 cases.[8] MMD is chronic, progressive cerebrovascular stenosis characterized by the terminal portion of the internal carotid artery becoming narrow or obstructing and

producing abnormal collateral vessels at the base of the brain. The exact cause is unknown. But it is believed to be caused by genetics or infection.

This abnormal cerebrovascular system can easily lead to ischemic stroke or hemorrhagic stroke. There are two peaks of incidence, the peak in the pediatric group occurs in the first decade and the peak in the adult group occurs at age 30-40 years. TIAs or ischemic strokes are common in children. Whereas most adult patients present with intracranial hemorrhage. Symptoms are associated with the regions of the brain supplied by the internal carotid arteries and middle cerebral arteries, including frontal, parietal, and temporal lobes. Patients may also have syncope, convulsion, visual deficits, or personality changes. The symptoms may be triggered by hyperventilation, exercise, crying, eating hot food, coughing, stress, fever, or dehydration.

No treatment can reverse or prevent cerebrovascular stenosis. The treatment involves managing symptoms and improving blood flow to the brain. Improvement in cerebral blood flow may prevent future ischemic strokes.[9,10] Since MMD affects the internal carotid artery while sparing the external carotid artery, surgical treatment typically uses the external carotid artery as a new source of blood supply to the ischemic areas. There are two methods of revascularization: direct and indirect. Direct revascularization uses a branch of the external carotid artery (usually the superficial temporal artery) directly anastomosed to a cortical artery. This method immediately increases the blood supply to the brain but is often difficult to perform in children because of small vessels. Indirect techniques use vascularized tissue supplied by the external carotid artery (e.g., dura, temporalis muscle) or the superficial temporal artery in direct contact with the brain, in order to stimulate blood vessel growth. This method is less difficult but takes several weeks for the new blood supply.

Medical therapy has been used when the patient has a mild disease or has a high risk for surgery. Antiplatelet agents have been used for stroke prophylaxis. Anticoagulants such as warfarin are rarely used. Calcium-channel blockers may be useful in headaches or migraines, which are common in MMD patients.

This case may have an unusual presentation of MMD such as posterior circulation involvement and only visual loss symptom. The main vascular supply to the occipital cortex is provided by branches of posterior cerebral arteries (PCAs). PCAs originate from posterior circulation (vertebrobasilar system) which is less effective in the early to middle stage of MMD. From MRA, this patient has no stenosis at PCAs, basilar artery, and vertebral arteries. But he may have abnormal vessels that cannot be detected by angiogram (more periphery or very small). He had a previous cerebral infarction without detected symptoms until the new infarction occurred. Many possible mechanisms could explain the cause of this acute infarction such as poor control of blood sugar, stress, and fever after getting the vaccination. The other mechanism that could be related to cerebrovascular stenosis, was triggered by COVID-19 vaccination. Because there were many studies showing the correlation between COVID-19 vaccination and vascular obstruction such as cerebral venous thrombosis[15], stroke[5], or vasospasm[4].

Conclusion

For patients present with occipital stroke in children or middle-aged groups, MMD should be

considered in differential diagnosis although it is a very rare disease. Although MMD usually affects the anterior circulation (ICAs and MCAs), the posterior circulation may also be affected.[12,13,14] MMD patients should be aware when getting vaccination because they can be triggered by stress, crying, fever, or the side effect of the vaccine itself.

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Footnotes and Financial Disclosures

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Address for correspondence: Orn Tempatarachoke, Department of ophthalmology, Samutprakarn Hospital 71 Chakkaphak road, Pak num, Mueang Samutprakarn, Samut Prakarn, Thailand 10270, Tel : 081-8685905, 02-7018132-9

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