Hemorrhage and ischemia in different hemispheres in a child with moyamoya disease: Case report and review of literature

Chandan B. Mohanty, Paritosh Pandey

Department of Neurosurgery, National Institute of Mental Health and Neurosciences, Bangalore, Karnataka, India

Abstract

Hemorrhagic type of moyamoya disease (MMD) is extremely rare in children. Ischemia following hemorrhage is very rare in MMD. There are only 11 reports of mixed-type of MMD, with the patient having both hemorrhage and ischemia in the same hemisphere at the time of presentation, or at different time periods. The ischemia is usually secondary to a precipitating cause. However, there are no reports of a child presenting with both ischemia and hemorrhage in different hemispheres. We present a previously unreported phenomenon of MMD, presenting as hemorrhage and ischemia in opposite hemispheres and review the relevant literature.

Key Words

Hemorrhage, ischemia, mixed type, moyamoya disease, pediatric

For correspondence:
Dr. Paritosh Pandey, Department of Neurosurgery, National Institute of Mental Health and Neurosciences, Bangalore, Karnataka, India. E-mail: paritosh2000@gmail.com

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Introduction

Moyamoya disease (MMD) is characterized by stenosis or occlusion of bilateral supraclinoid internal carotid arteries (ICA) or proximal middle cerebral artery (MCA) and anterior cerebral artery (ACA), along with the formation of basal leptomeningeal collaterals, called "moyamoya vessels." MMD is of two types-ischemic or hemorrhagic. The ischemic type is predominant in the pediatric population, while hemorrhagic type is more common in adults. Hemorrhagic type of MMD is rare in children.[1] There are some reports of mixed-type of MMD, with the patient having both hemorrhage and ischemia at the time of presentation or at different time periods. However, there are no reports of a child presenting with both ischemia and hemorrhage in different hemispheres. We present a case of MMD, presenting as ischemia and hemorrhage in opposite hemispheres. This has not been reported, to the best of our knowledge, in English literature.

Case Report

A 6-year-old female presented with sudden onset of weakness of the right side of the body, along with headache. Prior to this, she was asymptomatic, and had normal developmental milestones. On examination, she was conscious, alert and oriented to time, place and person; and had right hemiplegia. Computed tomography (CT) scan of the head revealed hemorrhage in the thalamus, posterior limb of the internal capsule on the left side, without any intraventricular extension [Figure 1]. Magnetic resonance imaging (MRI) brain revealed a small hematoma in the left thalamus, posterior limb of the internal capsule, without any intraventricular extension. Magnetic resonance angiogram revealed occlusion of bilateral supraclinoid ICA, with multiple basal leptomeningeal collaterals. There were no infarcts on diffusion-weighted MRI; however, there were multiple small hyperintensities on T2W1 in the white matter in the fronto-parietal region [Figure 2]. Patient was evaluated with a digital subtraction angiogram, which revealed occlusion of bilateral ICAs, along with leptomeningeal collaterals, suggestive of Suzuki stage 4 MMD [Figure 3]. There were pial-pial collaterals supplying the MCA and ACA territory bilaterally from both PCAs. There were no aneurysms in the leptomeningeal vessels. The hemorrhage was thought to be from the rupture of fragile leptomeningeal collaterals. In view of the severe disease, she was advised bilateral revascularization surgery after 6 weeks, to allow for improvement of her neurological deficits. She improved gradually, and after 2 months, was completely asymptomatic. Her motor power improved to normal, and she was neurologically normal.

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Two months after her hemorrhage, she had a sudden onset of weakness of left lower limb. There was no history of headache or seizures. There was no evidence of prior dehydration, crying or any other precipitating factor. On examination, she was conscious, alert and oriented. Power in the left lower limb was 2/5 proximally and 0/5 distally. Cranial CT scan revealed a well-defined hypodensity in the right medial frontal region, which was not enhancing on contrast, suggestive of infarct in right ACA territory. There was complete resolution of the previous bleed in the left ganglionic region, along with encephalomalacia in the same region.

The patient underwent bilateral superficial temporal artery-MCA bypass, first on the right side, followed by on the left side after 3 weeks. Middle meningeal arteries were preserved bilaterally. She had hyperperfusion syndrome, with transient aphasia following surgery on the left side, which improved completely at the time of discharge. CT following both surgeries did not show any evidence of infarct. At 6-month follow-up, she is asymptomatic, and did not have any further TIA or stroke. Digital substraction angiography carried out after 6 months showed patent grafts with good revascularization to bilateral hemispheres [Figure 4].

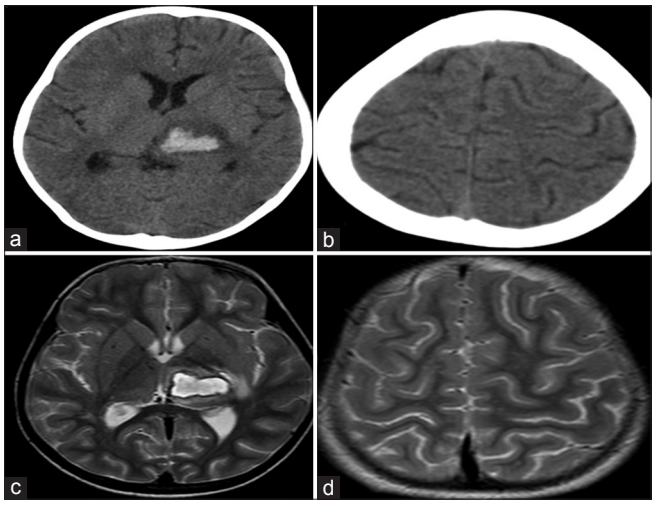

Figure 1: (a and b) Computed tomography head at the time of presentation showed bleed in the left thalamus and posterior limb of internal capsule without any infarct and (c and d) T2-weighted magnetic resonance imaging revealed bleed in the left thalamus and posterior limb of internal capsule without any infarct

Discussion

MMD usually presents as ischemia or hemorrhage; however, true mixed type of MMD is extremely rare. Ischemic type is common in children, however; there are reports of hemorrhagic type in children as well.[1] Only 11 cases of pediatric MMD presenting with ischemia after a prior hemorrhage have been reported in the literature [Table 1]. Ischemic complications have been reported following intraventricular hemorrhage (IVH) in patients with MMD. The pathophysiology of the ischemia is unclear; however, vasospasm following the hemorrhage has been implicated. It is theoretically possible to develop infarct following hemorrhage especially in severe MMD like the present case; however surprisingly, there are no reports of a patient having hemorrhage and ischemia in different hemispheres, at different time periods. Thus this is the first report of a true-mixed type of MMD in a pediatric patient.

Iwama *et al*.[2] reported three patients with cerebral ischemic complications complicating hemorrhage. One of the patients was 10 years while the others were 18 and 43 years old. All the patients had IVH, and were treated with hyperosmolar therapy. The ischemic complications were seen on 14th, 16th, and 11th day of ictus respectively. The ischemia was in the watershed territory in all patients, and was probably due to dehydration following hyperosmolar therapy, along with

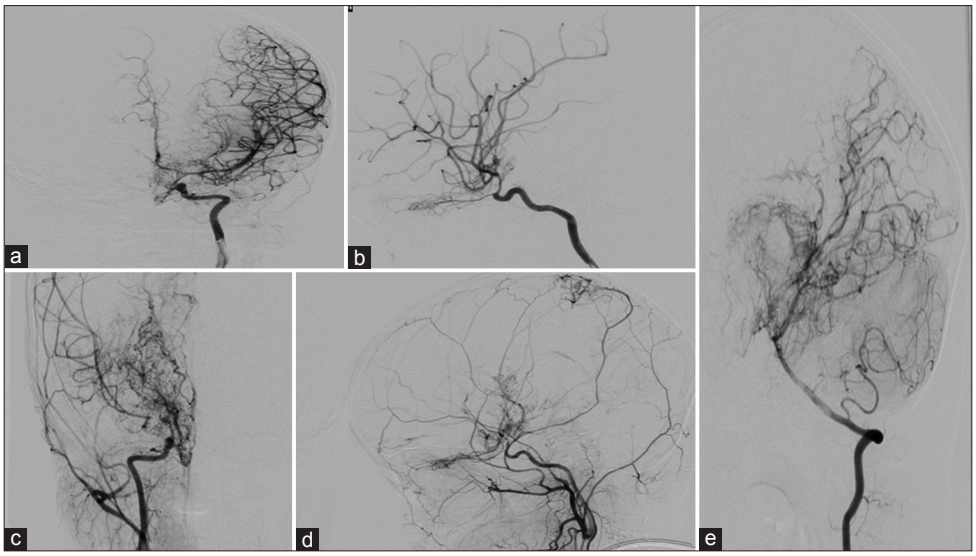

Figure 2: Digital substraction angiography – (a) Left internal carotid AP view, (b) Left internal carotid lateral view shows extensive basal collaterals, supraclinoid internal carotid arteries stenosis and poor filling in the ACA distribution, (c) Right internal carotid anteroposterior view, (d) Right internal carotid lateral view shows extensive basal collaterals, supraclinoid ICA stenosis and poor filling in the ACA and MCA distribution, (e) Right vertebral artery, lateral view shows extensive collaterals. No aneurysms were noted in the posterior circulation

raised ICP. Two of these patients improved and underwent revascularization while the third patient died due to the infarct. Su *et al*.[7] reported a 29-year-old woman with bilateral MMD, who had IVH. She had hypoperfusion of bilateral frontal cortices on CT perfusion, which went on to progress to infarct on 15th day following conservative treatment for the IVH, which included hyperosmolar agents. Nagasaka *et al*.[3] reported a 9-year-old girl, who presented with right caudate bleed and IVH, and MRI showed restricted diffusion and infarct in the right frontal lobe. They hypothesized that ischemia occurred in the vascular territory of the vessel which had ruptured, causing IVH. Nakai *et al*.[4] reported patients who experienced cerebral infarction 11 days after hemorrhage, presumably due to vasospasm of the collateral circulation. Patra *et al*.[5] presented a similar patient, who had ischemia 5 days after IVH. Rafay *et al*.[6] reported two patients with MMD presenting with hemorrhage, and having infarct. The first was a 10-year-old girl, who had IVH due to MMD, and then had right frontal and occipital infarct 15 days following the hemorrhage. The second patient was 11-year-old boy, who had right temporal hemorrhage with intraventricular extension, and had a right parieto-occipital infarct 4 days after the hemorrhage. Suyama *et al*.[8] reported two patients with MMD with hemorrhage followed by infarct. The 7-year-old girl developed infarct 16 days after an episode of right paraventricular and intraventricular bleed while the 9-year-old girl developed infarct 7 days after an episode of right paraventricular and lateral ventricular bleed.

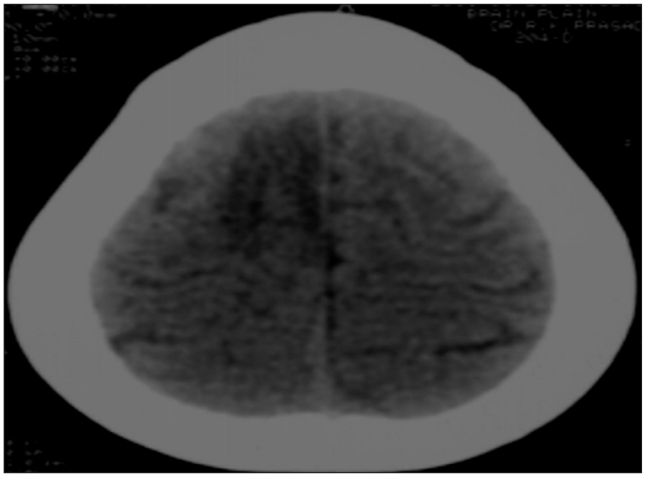

Figure 3: Computed tomography head 2 months after episode of bleed reveals right ACA territory infarct

Common in all the previously reported patients was IVH, and that infarct occurred 1-2 weeks following the hemorrhage. The causative factors may be raised ICP, dehydration, and shrinkage of ruptured vessel, poor intake or hypotension following treatment of these patients with hyperosmolar agents. Vasospasm may also play a role, in subacute stages. The present patient is unique from all the above reported patients, because she had a hemorrhage in one hemisphere and ischemia in other, without any precipitating factors. This is the first such case in the literature.

Hemorrhage is a rare presenting feature in children with MMD, reported in case reports and small case series. The incidence of hemorrhage in the pediatric population is reported to be about 3% while in adults it is about 25-30%.[1] Ahn *et al*.[1] presented a series of 13 children presenting with hemorrhage. Majority of the patients were in angiographic stage-3 with abundant MMV. Rupture of a microaneurysm or rupture of the friable moyamoya vessels are the two causes of hemorrhage in MMD. It has been proposed hemodynamic stress on the MMV causing rupture of these friable vessels is the main cause of bleed in the pediatric age-group.[1] The prognosis of hemorrhagic MMD is poorer than ischemic MMD. Revascularization reduces the risk of further ischemia in these patients; however, the role of revascularization in reducing future hemorrhage is unclear.

In conclusion, ischemia following hemorrhage in patients with MMD is a rare event, and is probably related to changes in cerebral hemodynamics following hemorrhage, increase in ICP and dehydration related to hyperosmolar therapy. It is important to manage these patients carefully to avoid hypotension and dehydration, and treat raised ICP. Our patient

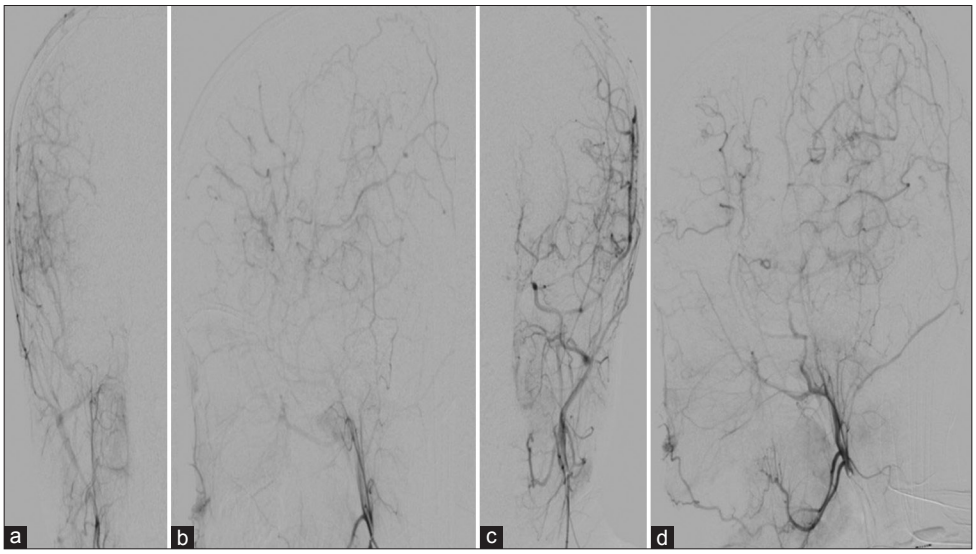

Figure 4: Postop digital substraction angiography (a-d) patent grafts with good revascularization to bilateral hemispheres

Table 1: Summary of reported cases in literature of cerebral infarction following hemorrhage in pediatric moyamoya disease

Author	Age/ sex	Presentation	Repeat episode	Time after bleed	Outcome	Pathogenesis
Iwama *et al*., 2001[2]	10/F	Headache, CT showed IVH, DSA showed b/l MMD	Lt hemiparesis, aphasia, CT showed Rt parieto-occipital infarct	14 days	Good after Lt STA-MCA bypass	Dehydration, hyperosmolar agent
	18/M	Loss of consciousness, CT showed IVH, DSA showed b/l MMD	Rt hemiparesis, CT showed Lt parietal infarct	16 days	Good after bilateral STA-MCA bypass	Dehydration, hyperosmolar agent
	46/F	Headache, CT showed Lt caudate bleed with IVH	Rt hemiparesis, CT showed Lt parieto-occipital infarct	11 days	Died due to hemorrhage and infarct	Dehydration, hyperosmolar agent
Nakai *et al*., 2002[4]	29/F	Headache, drowsiness, CT showed IVH	Worsening of sensorium, CT showed infarct	16 days	Died	Vasospasm, hypercoagulable state
Rafay *et al*., 2006[6]	10/F	Trisomy 21, headache, drowsiness, incontinence. CT showed IVH, DSA showed b/l MMD	Recurrence of headache, Lt hemiplegia, CT showed Rt frontal and occipital infarct	15 days	Residual moderate hemiparesis, no surgery	Raised ICP
	11/M	Headache, vomiting, CT showed Rt mesial temporal hemorrhage with IVH, DSA showed b/l MMD	No fresh symptoms, MRI showed Rt parieto-occipital infarct	4 days	Good after bilateral pial synangiosis, left upper quadrantanopsia	Raised ICP
Nagasaka *et al*., 2007[3]	9/F	Headache, vomitings, Lt hemiparesis, CT showed Rt caudate hemorrhage with IVH, DSA showed b/l MMD	MRI showed Rt frontal infarct	Concomitant	Good	Vasospasm, possible occlusion of afferent vessel
Su *et al*., 20080[7]	29/F	Headache, drowsiness, CT showed IVH, DSA showed b/l MMD	Worsening of sensorium, CT showed b'l frontal infarct	15 days	Good after Left STA-MCA bypass	Dehydration, hyperosmolar agent
Suyama *et al*.,2008[8]	7/F	Headache, vomiting, drowsiness. Cerebral angiography showed b/l MMD. CT showed Rt paraventricular white matter bleed and IVH	Severe headache, left hemiparesis. MRI showed multiple right frontal and b/l parietal subcortical infarcts	16 days	Good. Initially treated with edaravone, then STA-MCA bypass and encephalo-myo-synangiosis	Dehydration, hyperosmolar agent
	9/F	Headache, drowsiness. Prior history of headache and transient rt. Upper limb monoparesis. CT showed bleed in right paraventricular white matter and IVH. Cerebral angiography showed b/l MMD	Headache, transient rt hemiparesis. MRI lt. frontal infarct	7 days	Good. Initially treated with edaravone, then STA-MCA bypass and encephalo-myo-synangiosis	Dehydration, hyperosmolar agent
Patra *et al*., 2012[5]	12/M	Headache, vomiting. CT showed IVH, MRA showed b/l MMD	Rt. hemiparesis, MRI showed infarcts in right and left posterios paraventriclar area, centrum semiovale	5 days	Good, no intervention	Dehydration, hyperosmolar agent

CT=Computed tomography, IVH=Intraventricular hemorrhage, DSA=Digital substraction angiography, STA=Superficial temporal artery, MCA=Middle cerebral artery, MMD=Moyamoya disease, ICP=intracranial pressure, b/l=Bilateral, MRI=Magnetic resonance imaging

is unique because the two events seem unrelated, and is the first example of both types of MMD in the same patient, in opposite hemispheres.

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