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Angiographic findings of Moyamoya

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Background : *Moyamoya is a rare cerebrovascular occlusive condition of unknown etiology which has rarely been described in Thai population. We report angiographic findings and clinical features from a single institution in Thailand.*

Objective : *Describe angiographic findings and clinical features of moyamoya in patients.*

Design : *Retrospective descriptive study.*

Setting : *Department of Radiology, Faculty of Medicine, Chulalongkorn University.*

Materials and Methods : *Twenty-five patients diagnosed with moyamoya and moyamoya-like by cerebral angiography between 2003-2009 were retrospectively reviewed for clinical manifestation and stenoocclusive lesions according to Suzuki Grading System.*

Results : *Of the total 25 patients,19 were female (76%), 6 were male (24%), F:M = 3:1. Their median age was 38 years. Five of 6 pediatric patients (83%) presented with ischemic stroke, only one patient (17%) presented with hemorrhagic stroke, Thirteen of 19 (68%) adult patients presented with hemorrhagic stroke, whereas 6/19 (32%) presented with ischemic stroke. From 48 examined cerebral hemispheres, none was grade as stage I (0%), 2 (4.2%) as stage II, 14 (29.2%) as stage III, 19 (39.6%) as stage IV, 12 (25%) as stage V,1(2%) as stage VI.*

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Conclusions : *Clinical presentation and angiographic findings of moyamoya in this study is consistent with prior observations, especially from Japan. However, it differs from the studies in the United States, particularly in clinical manifestation.*

Keywords : *Moyamoya, stroke, cerebral blood vessels, stenosis or occlusion, collateral circulation, cerebral angiography.*

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เหตุผลของการทำวิจัย : *Moyamoya เป็นภาวะที่มีการอุดตันของหลอดเลือดสมองโดยไม่ทราบสาเหตุแน่ชัด ซึ่งมีอุบัติการณ์ต่ำและมีการรายงานค่อนข้างน้อยในประเทศไทย ผู้วิจัยได้ศึกษาลักษณะภาพทางรังสีหลอดเลือดและลักษณะทางคลินิกของผู้ป่วย moyamoya*

วัตถุประสงค์ : *บรรยายลักษณะภาพทางรังสีหลอดเลือดและลักษณะทางคลินิกของผู้ป่วย moyamoya*

รูปแบบการวิจัย : *การศึกษาแบบย้อนหลังเชิงพรรณนา*

สถานที่ทำการศึกษา : *ภาควิชารังสีวิทยา คณะแพทยศาสตร์ จุฬาลงกรณ์มหาวิทยาลัย*

ตัวอย่างและวิธีการศึกษา : *ศึกษาผู้ป่วย moyamoya 25 คนที่วินิจฉัยจากการตรวจทางรังสีหลอดเลือดสมองในโรงพยาบาลจุฬาลงกรณ์ ตั้งแต่ปีพ.ศ. 2546 - 2552 โดยบรรยายลักษณะทางคลินิกและความรุนแรงของภาวะการตีบหรืออุดตันของหลอดเลือดโดยใช้ Suzuki Grading System*

ผลการศึกษา : *ในจำนวนผู้ป่วยทั้งหมด 25 คน ประกอบด้วยเพศหญิง 19 คน (76%), เพศชาย 6 คน (24%), อัตราส่วนเพศหญิง:ชาย = 3:1, อายุเฉลี่ย 38 ปี, ผู้ป่วยเด็ก 5 คน (83%) แสดงอาการของสมองขาดเลือด, มีเพียง 1 คน (17%) แสดงอาการมีเลือดออกในสมอง ผู้ป่วยผู้ใหญ่ 13 คน (68%) แสดงอาการมีเลือดออกในสมองในขณะที่อีก 6 คน (32%) แสดงอาการของสมองขาดเลือด ในจำนวน 48 ซีกสมองที่มีพยาธิสภาพแบ่งตามความรุนแรงออกเป็น stage 1 = 0%, stage 2 = 2 (4.2%), stage 3 = 14 (29.2%), stage 4 = 19 (39.6%), stage 5 = 12 (25%) และ stage 6 = 1 (2%)*

สรุป : *จากการศึกษาพบว่าลักษณะภาพทางรังสีหลอดเลือดและลักษณะทางคลินิกของผู้ป่วย moyamoya ที่พบในโรงพยาบาลจุฬาลงกรณ์มีความสอดคล้องกับการศึกษาอื่น ๆ โดยเฉพาะในประเทศญี่ปุ่น แต่แตกต่างกับการศึกษาในประเทศแถบตะวันตกเช่น สหรัฐอเมริกา โดยเฉพาะในแง่ของการแสดงทางคลินิก*

คำสำคัญ : *Moyamoya, ภาวะสมองขาดเลือด/เลือดออกในสมอง, หลอดเลือดสมองตีบ/อุดตัน, การไหลเวียนสำรอง,การตรวจทางรังสีหลอดเลือดสมอง.*

Moyamoya is a rare cerebrovascular occlusive condition associated with progressive stenosis of the intracranial internal carotid arteries (ICAs) and their proximal branches, especially the anterior and middle cerebral arteries (ACAs and MCAs).[1-3] This process may involve posterior circulation, including basilar and posterior cerebral arteries (PCAs).[4,5]

Etiology of moyamoya is unknown. However, it also occurs secondary to various underlying diseases, known as "moyamoya-like" or "moyamoya syndrome".[2,6] Their manifestations vary among geographical regions. Studies in Asian populations indicate that adults have much higher rates of hemorrhage as presenting symptom than children.[1,2] In the United States, the majority of affected adults and children present with ischemic symptoms.[7,8]

This study set out to investigate whether differences exist between moyamoya at King Chulalongkorn Memorial Hospital (KCMH) and other studies, with regarding to angiographic findings and clinical features.

Materials and Methods

Patients

Twenty-five patients diagnosed with moyamoya and moyamoya-like by cerebral angiography at KCMH, between Jan 01,2003 and Dec 31, 2009. Inclusion criteria included unilateral or bilateral angiographic identifications of severe stenosis or occlusion of the distal ICA and/or proximal ACA or MCA (definite and probable cases according to diagnostic criteria of moyamoya disease by the Research Committee on Spontaneous Occlusion of the Circle of Willis in Japan).[9] Patients with previous revascularization surgery were excluded.

Imaging Examinations

All patients underwent cerebral angiography by transfemoral catheterization technique. Angiography was performed including bilateral internal carotid arteries (ICAs), bilateral external carotid arteries (ECAs), bilateral common carotid arteries (CCAs) and unilateral or bilateral vertebral arteries (VAs) anteroposterior (AP), lateral and oblique projections. Images were obtained with digital subtraction technique.

Image analysis

All cerebral angiograms were retrospectively reviewed by an interventional neuroradiologist on the picture archiving and communicating system (PACS), regarding sites (intracranial/ extracranial ICA, MCA, ACA, PCA and basilar artery), severity of occlusion (angiographic staging according to Suzuki Grading System)[1], types of collateral vessels (basal moyamoya, leptomeningeal, transdural) and associated aneurysm.

Information about gender, age, underlying disease and presenting symptoms of the patients were collected.

Results

Twenty-five patients were identified (Figure1). Nineteen are female (76%), 6 are male (24%), F: M = 3:1, their ages were between 2 - 73 years (median age = 38), 19 were adults (above 15 years), 6 were children (15 years or under).

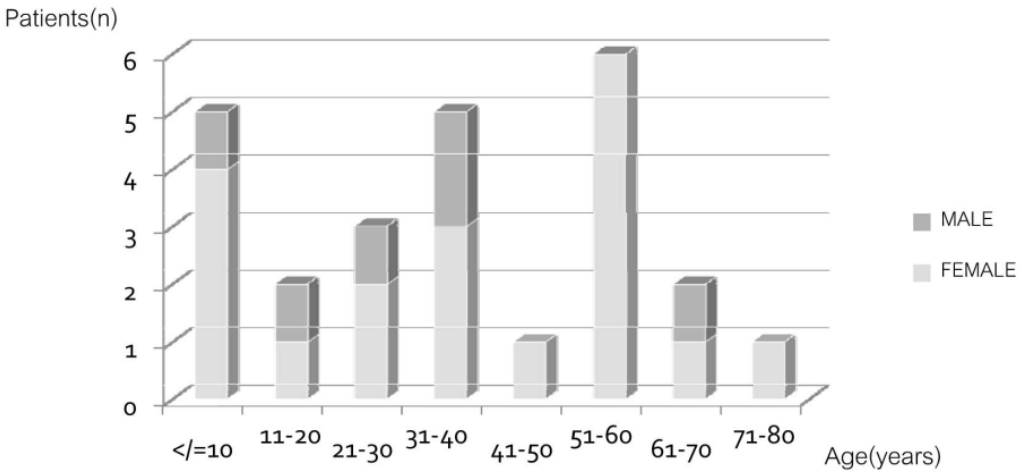

Figure 1. Age distribution of the patients with moyamoya disease by sex.

Five of 6 pediatric patients (83%) presented with ischemic stroke, only one patient (17%) presented with hemorrhagic stroke. Thirteen of 19 (68%) adult patients presented with hemorrhagic stroke, whereas 6/19 (32%) presented with ischemic stroke.

Twenty- three cases are bilateral involvement (definite cases), whereas 2 cases were unilateral involvement (probable cases), total number of involved hemispheres was 48 hemispheres. Extracranial ICA involvement was seen in 35/48 hemispheres (73%). In total of 15/50 PCAs (30%) manifested steno-occlusive lesion. Only one patient (4%) who presented with IVH demonstrated associated aneurysm (Figure 5d).

Several associated diseases were found such as thyroid carcinoma, renal artery stenosis (RAS), myasthenia gravis (MG), systemic lupus erythematosus (SLE), dyslipidemia (DLP), diabetes mellitus (DM), hypertension (HT), protein C and S deficiency, hyperthyroidism, neurofibromatosis type 1 (NF1), and mitochondrial myopathy.

Of the 48 examined cerebral hemispheres, none was graded as stage I (0%), 2 (4.2%) stage II, 14 (29.2%) stage III, 19 (39.6%) stage IV, 12 (25%) stage V and 1 (2%) stage VI. Thus, about 94% of the hemispheres were graded as stage III - V (Figure 2-7).

Left ICA angiogram; AP projection (3a) reveals severe stenosis of supraclinoid segment of ICA with minimal lenticulostriated collateral vessels. LVA angiogram; AP (3b) and lateral (3c) projections reveal stenosis of P1 and P2 segments of bilateral PCAs with developed thalamoperforating collaterals. Leptomeningeal collaterals via posterior pericollosal and posterior temporal arteries are also noted.

Left ICA angiogram; AP (4a) and lateral (4b) projections reveal irregular narrowing at distal ICA with complete occlusion at supraclinoid segment and complete occlusion of left MCA. Multiple small neovascular collateral vessels are seen at distal ICA and proximal MCA, corresponds with “puff-of-smoke” pattern. Lateral projection of left CCA angiogram (4c) shows a small plaque on the posterior wall at the origin of left ICA. No transdural collateral from ECA is noted.

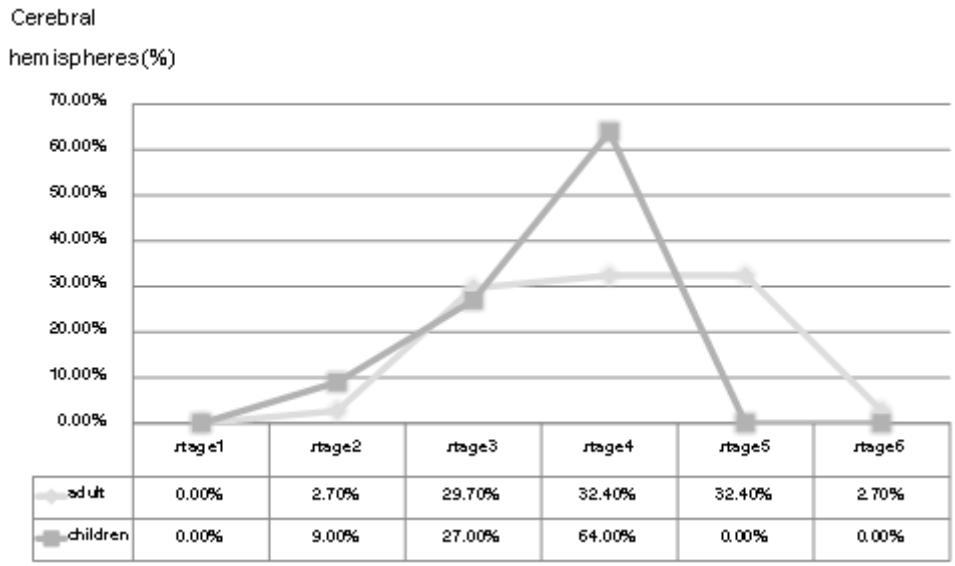

Figure 2. Stage of ICA lesion in adults and children.

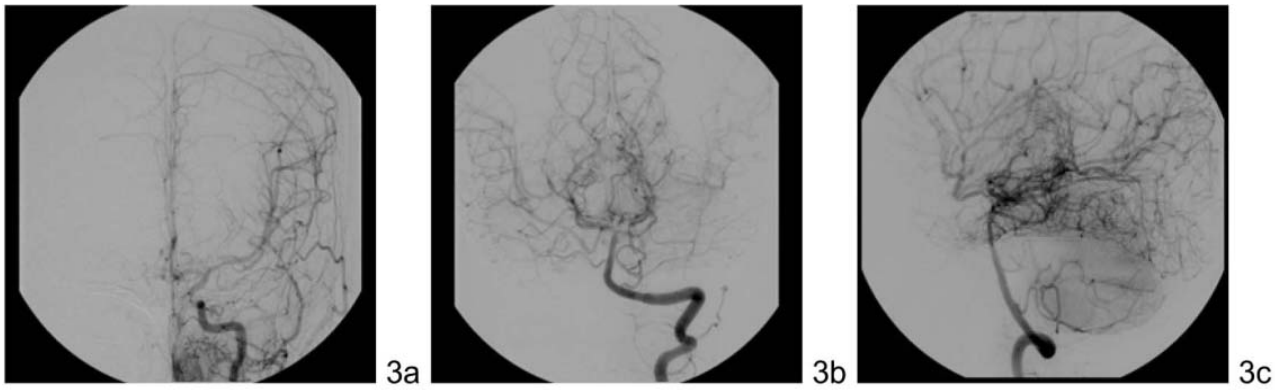

Figure 3. Grade II lesion with PCAs involvement: A 9-year-old female with hypercholesterolemia, presented with recurrent stroke and cerebral infarction was shown from CT scan.

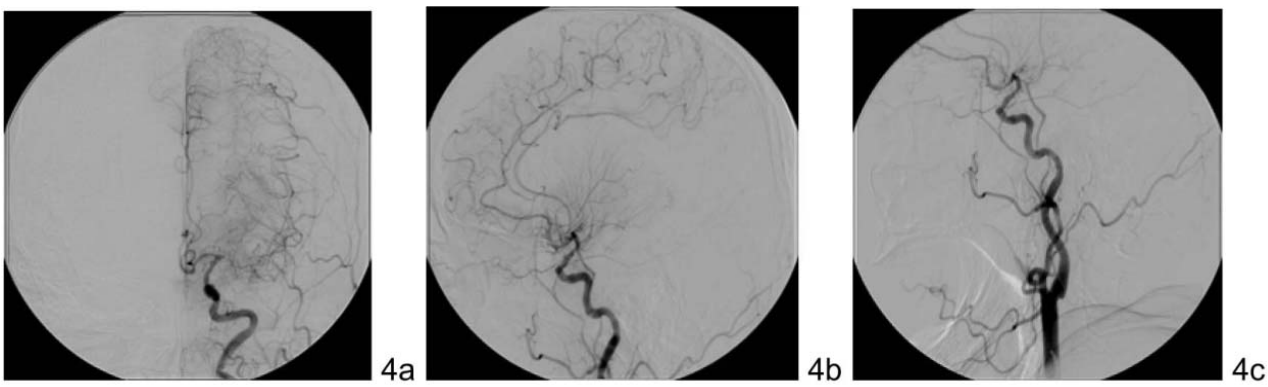

Figure 4. Grade III lesion: A 59-year-old female with history of hypertension, presented with subarachnoid hemorrhage.

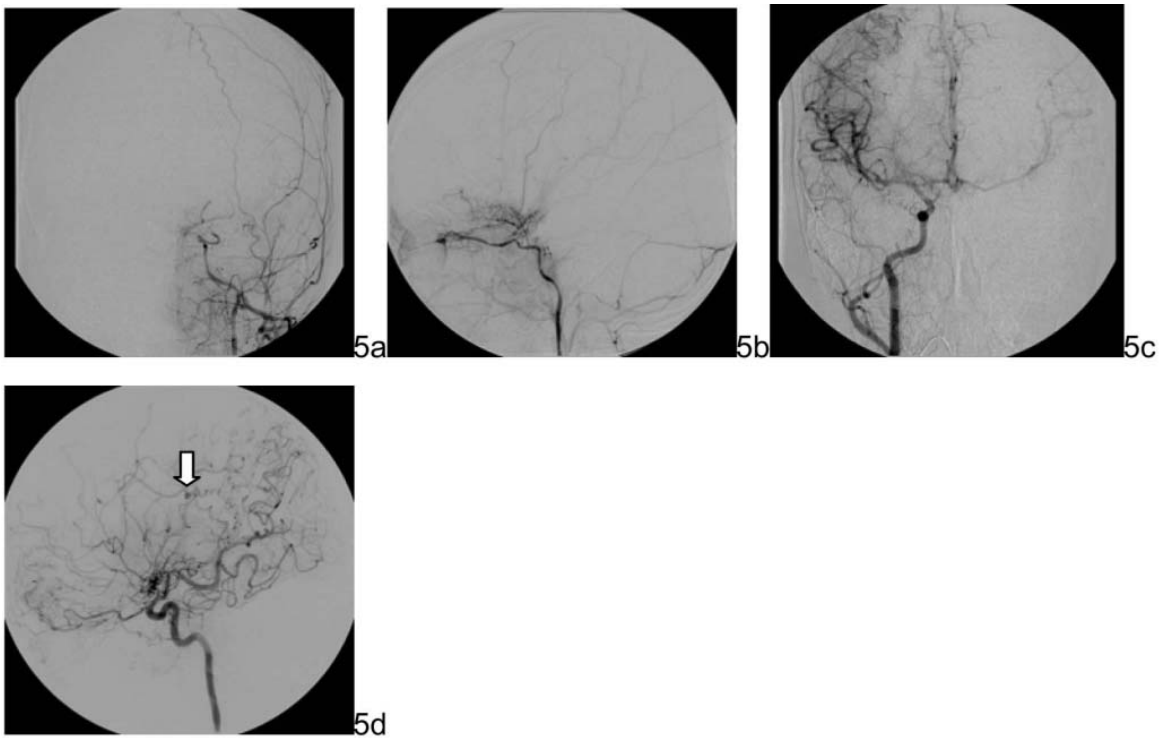

Figure 5. Grade IV lesion: A 6-year-old female presented with progressive right hemiparesis.

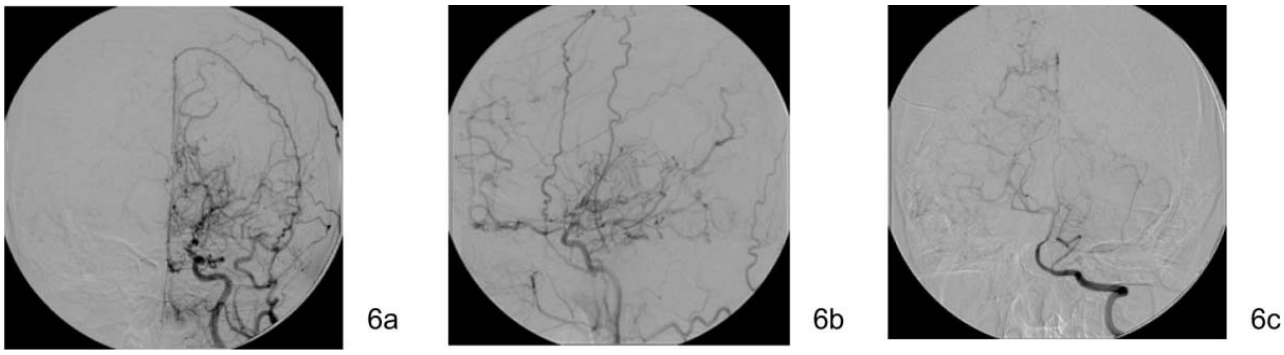

Figure 6. Grade V lesion: A 42-year-old female presented with intracerebral hemorrhage

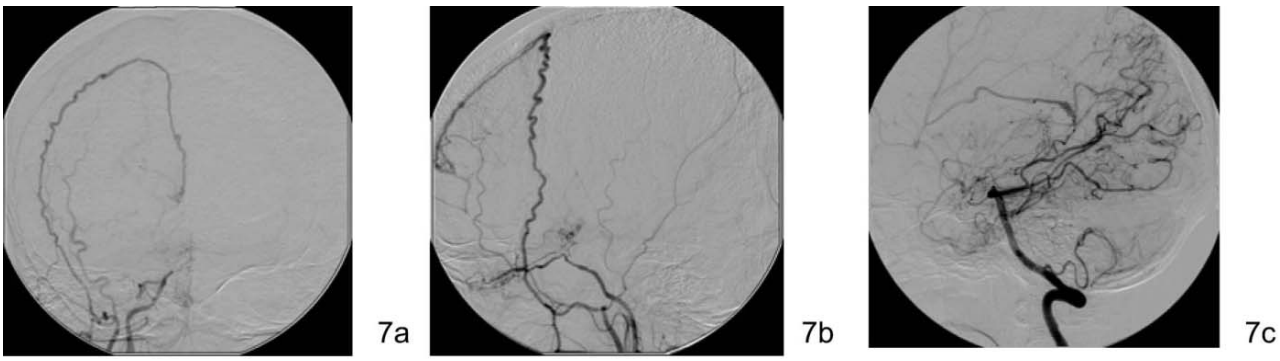

Figure 7. Grade VI lesion: A 63-year-old female with underlying hypertension and dyslipidemia presented with intracerebral hemorrhage.

Left CCA angiogram; AP (5a) and lateral (5b) projection shows stenosis of ICA from its origin (not shown) with total occlusion at supraclinoid segment. Transdural collaterals from middle meningeal artery and opthalmic artery are noted. AP projection of right ICA (5c) shows normal caliber of right ICA, ACA and MCA. Left MCA and ACA received blood flow from contralateral side via anterior communicating artery (ACoA).

Another case (5d), a 39-year-old woman with history of CA thyroid who presented with intraventricular hemorrhage. A small saccular aneurysm is detected, originated from dilated posterior choroidal artery (arrow).

Left ICA angiogram; AP(6a)and lateral(6b) projections show occlusion of supraclinoid segment of ICA, A1 segment of ACA and M1 segment of MCA. Multiple collateral vessels via basal perforators "Moyamoya vessels" are seen with extensive transdural collaterals from ECA via middle meningeal artery and ophthalmic artery. Left VA angiogram reveals severe stenosis of basilar artery and left PCA (6c).

Right ICA angiogram; AP (7a) and lateral (7b) projections and LVA angiogram; lateral projection (7c) show severe stenosis of ICA from its origin (not shown) with occlusion of supraclinoid segment of right ICA, total occlusion of ACA and MCA. Decreased Moyamoya vessels with extensive transdural and leptomeningeal collaterals accompanying with decreased cerebral blood flow are observed.

Discussion

Clinical presentation and radiographic findings of moyamoya in this study is consistent with prior observations of Suzuki *et al*.[1] and Scott *et al*.[2], who reported that adults have much higher rate of hemorrhage as a presenting symptom than children. However, clinical presentations in this study differs from studies from those of Chiu *et al*.[7] and Hallemeier *et al*.[8] who reported that ischemic stroke is predominant symptom in both adults and children. This may be attributed to genetic and environmental factors.

Our study had slightly older population than others, possibly due to population of solely symptomatic patients. Atherosclerosis may involve in the development of the disease in elderly patients.[10] In our institution, cerebral angiography is the standard diagnostic work up in subarachnoid hemorrhage. This may contribute to high percentage of adult moyamoya patient presented with hemorrhage in this study, therefore moyamoya associated ischemic stroke was under-recognized.

Several associated diseases in our study are similar to the previous reports, such as, RAS, SLE, DLP, DM, HT, hyperthyroidism and NF1.[1,11,12] However, some diseases have not been previously reported such as thyroid carcinoma, protein C and protein S deficiency, and mitochondrial myopathy.

Other than indicating performed for diagnosis and evaluate disease severity, cerebral angiography also play an important role in evaluating donor vessels before determination of revascularization surgery. Therefore, the data from this study could suggesting the more careful consideration of this disease among the patients who presented with stoke-like symptoms, as well as provides information for further surgical treatment planning.

Our study has some limitations. First, the number of patients recruited in the study was small. Second, because of the retrospective nature, a possible bias in patient selection could not be completely excluded. We recruited only patients who had cerebral angiography, patients who had been diagnosed on MRA or CTA alone were excluded. Therefore, some bias might appear in the more advanced stages.

Conclusion

Clinical presentation and angiographic findings of moyamoya in this study is consistent with prior observations, especially those in Japan. However, the pattern of the results are different from the studies in the United States, particularly in clinical manifestation.

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