

Moyamoya Disease in a Turkish Woman: A Case Illustration

Mehtap Erdogan^{1*}, Keziban Karacan², Alper Karacan³, Ayla Tekin Orha¹, Tuncay Colak¹ and Hamit Macit Selekler⁴

¹Department of Anatomy, Faculty of Medicine, Kocaeli University, Turkey; ²Department of Anatomy, Faculty of Medicine, Sakarya University, Turkey; ³Department of Radiology, Faculty of Medicine, Sakarya University, Turkey; ⁴Department of Neurology, Faculty of Medicine, Kocaeli University, Turkey

Corresponding author:
Mehtap Erdogan, Department of
Anatomy, Faculty of Medicine,
Kocaeli University, Turkey,
Tel: +90(553)1124746; E-mail:
mehtaperdogan@sakarya.edu.tr

Abstract

The etiology of Moyamoya Disease (MMD) is unknown. It is a cerebrovascular disease which is caused by occlusion or stenosis and can be diagnosed by angiographic methods. Digital Subtraction Angiography (DSA) is the gold standard for diagnosis, especially in cases with mild stenosis and few collateral lesions. The causes of this disease are unknown; it is usually associated with congenital or tumoral defects. This disease is more common in the yellow race, especially woman in Japan. Its prevalence is low in countries other than Japan. The high prevalence of the disease in Japan has given it its name from Japanese. Moyamoya; Meaning of Japanese means smoke dispersing in the air.

The veins in the appearance of smoke scattering in the air giving the name of Moyamoya are, in fact, perforating arteries which are enlarged and angiographically visible to form collateral pathways due to stenosis. The first symptoms in Moyamoya may occur in various forms, including recurrent headache, epilepsy, acute hemiplegia. The clinical picture varies between children and adults. Ischemia in children and early hemorrhage in adults. Angiography is accepted as the gold standard for diagnosis of MMD. The criteria for angiographic diagnosis of MMD have been determined by the Research Committee for Progressive Obstructive Diseases of Circle of Willis of the Ministry of Health of Japan.

In this article, a 38-year-old female patient, who was admitted to the emergency department with complaints of severe headache and vomiting two years ago and whose hemorrhage areas were detected and discharged with conservative treatment, was admitted to the emergency department with sudden onset hemiparesis, nausea, vomiting and left hemiparesis. We present a case of MMD diagnosed in a 38-year-old female patient presenting with right capsula interna crus posterior and basal core hemorrhage on Magnetic Resonance Angiography (MRA) images.

Keywords: Angiography; Hemorrhage; Hemiparasi; Moyamoya; Digital Subtraction Angiography (DSA)

Introduction

MMD is a chronic cerebrovascular disease that characterized by progressive stenosis or occlusion at the distal ends of internal carotis artery bilaterally. [1-3]

For the first time in history, the case thought to be MMD was described by Kudo in 1956. [4] In 1957, Takeuchi and Shimizu talked about the structural change in the internal carotid arteries in a male case. [5] In 1964, Nishimoto described the disease in detail, and from that date until 1969 it was called Nishimoto's disease. [6] The name moyamoya", which means cigarette smoke spreading in the air, was given by Suzuki in 1969 and this name was generally accepted. [5]

Collateral vessels develop slowly from the distal region of the occlusion to compensate for the resulting occlusion. This abnormal form of revascularization creates an appearance similar to cigarette smoke dispersed in the air on angiography. The disease takes its name from this image of cigarette smoke because moyamoya in Japanese means cigarette smoke. [7]

Anterior cerebral circulation is affected in MMD. [8] Moyamoya syndrome, which is accompanied by some tumoral and genetic diseases, is most common in Japan, especially in women. [9]

While more hemorrhagic findings are seen in the adult type, ischemic symptoms are at the forefront in pediatric cases. [10,11]

Magnetic Resonance Imaging (MRI), Magnetic Resonance Angiography (MRA) and Diffusion Cerebral Angiography (DSA) are used for diagnosis. [12]

The treatment of this disease, which generally holds the brain areas bilaterally, is surgery in the acute stage and conservative in the chronic stage. The goal of treatment is to minimize the

This is an open access article distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 3.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms.

How to Cite this Article: Erdogan M, et al. Moyamoya Disease in a Turkish Woman: A Case Illustration. Ann Med Health Sci Res. 2021;11:1351-1354.

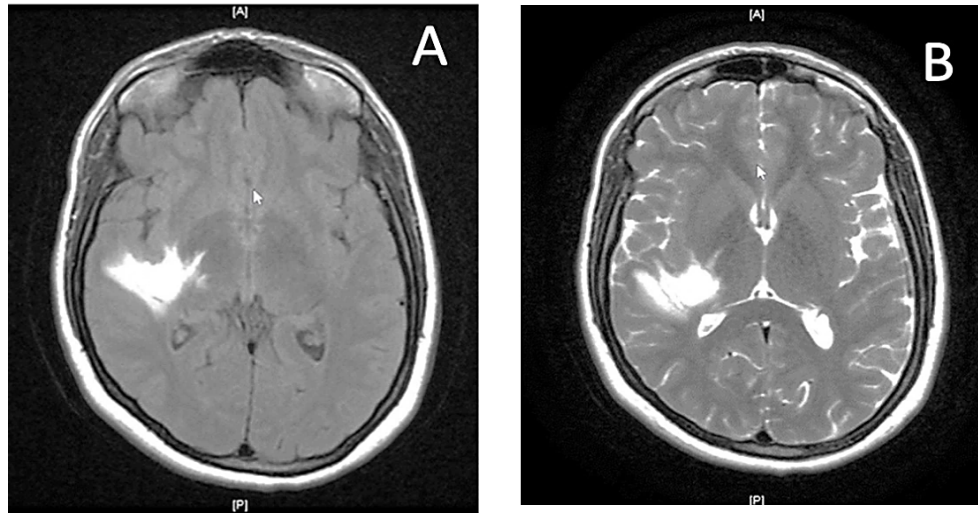


Figure 1: Magnetic resonance angiography (MRA) imaging. A) Right capsular interna crus posterior and B) Hemorrhage into the ventricle at the level of the right putamen.

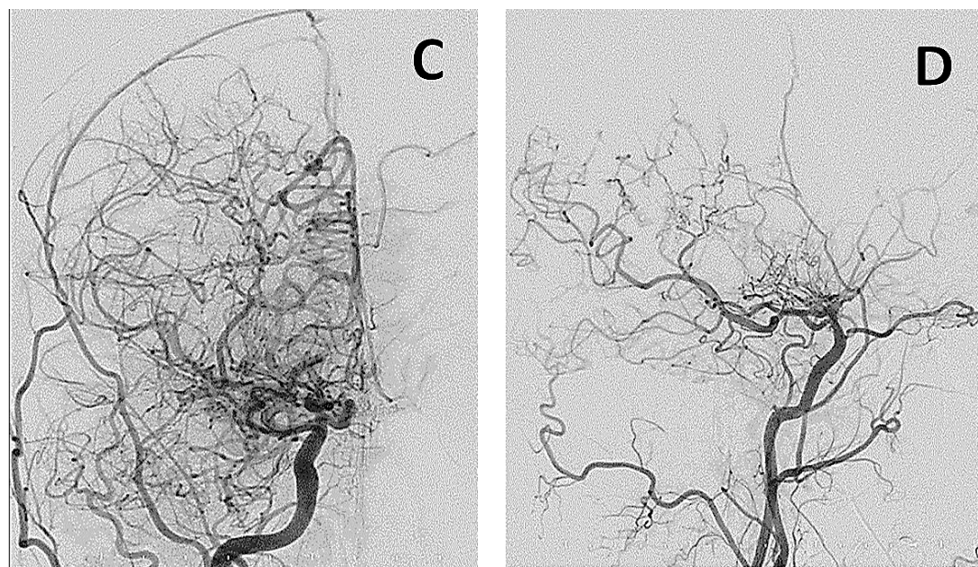


Figure 2: Coronal (C) and sagittal (D) DSA images show collateral vascular structures due to anterior and middle cerebral artery occlusions.

possibility of hemorrhage and prevent ischemic symptoms. However, since the etiopathogenesis of the disease is not clearly known, a complete treatment method to stop pathological progression is not yet available. Morbidity is more than 70% in untreated moyamoya patients.^[13]

Case Report

A 38-year-old woman who applied to the emergency clinic with the complaints of severe headache and vomiting approximately two years ago, hemorrhage areas were detected, and conservative treatment was applied and discharged [Figure 1]. It was learned those two years after this event, hemiparesis started suddenly in the left half of the body. After brain Computed Tomography (CT) and Magnetic Resonance Angiography (MRA) Digital Cerebral Angiography (DSA) was requested from the patient with suspicious bleeding areas at the level of the right capsular interna crus posterior and right basal nuclei. In DSA [Figure 2], bilateral (ICA) pars petrosa was markedly calibrated, with collaterals of left ICA pars supracavernosa and right External Carotis Artery (ECA) and posterior flow through arteria communicans anterior and posterior.

Right ICA pars supracavernosa and arteria communicans posterior dextra were not observed, arteria cerebri media dextra (MCA) was poorly observed through the arteria communicans anterior. Bilateral ICA pars supraclinoidea compatible with MMD, late filling in MCA, and smoky appearance in lenticulostriate artery localization was observed. Bilateral arteria cerebri anterior was not observed. It was observed that arteria ophtalmica sinistra and arteria orbitofrontalis and arteria pericallosae, both of the arteria cerebri posterior and arteria cerebellaris superiores, were filled.

Discussion

Moyamoya disease is a disease that causes multiple perforating anastomoses around Circle of Willis, the etiology of which is not fully known.^[14,15] The annual prevalence of MMD in Japan, whose 90% of its incidence is made up of Japanese patients worldwide, is 3.16/100,000.^[16] However, in recent years, the prevalence of MMD in different countries has also increased in non-Asian populations.^[17]

The disease is predominantly seen in women. MMD; It is

named according to the diagnosis period, as early (5-15 years old) and late (35-50).^[18] Our case of women presented in this study; Being diagnosed at age 38, she is in the late MMD class.

Stenosis or complete occlusion in the terminal part of the ICA in MRA (Magnetic Resonance Angiography) in an idiopathic moyamoya case; there are narrowing or complete occlusions in the proximal parts of the anterior cerebral artery and middle cerebral artery and abnormal vascular networks at the level of the basal ganglia, as in our case.^[19]

In Positron Emission Tomography (PET) imaging in MMD; Oxygen Extraction Fraction (OEF) increased and cerebral blood volume decreased.^[20] In parallel with this information, in our moyamoya case we presented, a decrease in cerebral volume and an increase in OEF was found in PET radiography taken before DSA.

Diagnostic standard in MMD is DSA. In angiography; stenosis or complete occlusion of distal ICA vessels, indistinct proximal parts of the anterior and middle cerebral arteries, well-developed moyamoya vessels in the basal parts of the brain, collaterals between cortical arteries or between cortical arteries and leptomeningeal arteries can be detected.^[21] In our case, in all radiological imaging performed before DSA, proximal parts of the anterior and middle cerebral arteries were indistinct and hematoma areas at the level of the basal ganglia were present. ICA occlusions and a large number of small collateral vascular network images were detected in DSA imaging. Definitive diagnosis made after DSA. Moyamoya should definitely be considered in the differential diagnosis if there is bilateral internal carotid artery distal anterior and middle cerebral artery stenosis in cerebral angiography imaging. Since collateral vascular structures called Moyamoya vessels cannot be demonstrated in CT and MR angiography, DSA must be performed in the diagnosis of Moyamoya patients. Our case was diagnosed with DSA, similar to the literature.

The primary goal of MMD treatment is to increase cerebral blood flow and decrease intracranial pressure. Intervention may be required for hematoma evacuation and ventricular drainage. Thrombolytic therapy is not recommended in MMD cases with ischemic stroke to prevent rupture due to the tendency of collaterals to bleed.^[22] In our case; Left ischemic hemiparesis developed due to impaired cerebral blood flow. Therefore, to our patient; First applied in 1972 even though the application of superficial temporal artery and middle cerebral artery anastomosis technique;^[23] direct intervention was abandoned due to the reduction of ischemic symptoms and Intracranial Pressure (ICP).

Conclusion

In our case report study, although it is very rare in our country, it is necessary to be careful about MMD in addition to conservative treatment in resistant headaches, young age haemorrhage cases, MMD can be seen in our country and after conservative treatment, and we wanted to remind him to be warned about coming to the controls. If there is bilateral internal carotid artery distal and anterior cerebral artery and media involvement in cerebral angiography images, Moyamoya should definitely be

considered in the differential diagnosis and should be confirmed by DSA.

Competing Interests

The authors report no competing (commercial/academic) interests.

References

1. Kudo T. Spontaneous occlusion of the circle of Willis: a disease apparently confined to Japanese. *Neurol.* 1968;20:485-496.
2. Nishimoto A, Takeuchi S. Abnormal cerebrovascular network related to the internal carotid arteries. *J Neurosurg.* 1968;29:255-260.
3. Benbir G, Bozluolcay M, İnce B. Moya Moya Disease, Morning Glory Disc Anomaly, and Neurosensorial Hearing Loss. *J Neurol Sci.* 2005;22:409-413.
4. Eguchi T, Ugajin K. Surgical management of moyamoya disease. Schmideck HH ed. *Operative Neurosurgical Techniques.* Philadelphia: WB Saunders, 1988;797-806.
5. Maki Y, Enomoto T. Moyamoya disease. *Childs Nerv Syst.* 1988;4:204-212.
6. Vuia O, Alexianu M, Gabor S. Hypoplasia and obstruction of the circle of Willis in a case of atypical cerebral hemorrhage and its relationship to Nishimoto's disease. *Neurol.* 1970;20:361-367.
7. Yamamoto M, Aoyagi M, Tijima S. Increase in elastin gene expression and protein synthesis in arterial smooth muscle cell derived from patients with moyamoya disease. *Stroke.* 1998;28:1733-1738.
8. Cerrato P, Grasso M, Lentini A, Destefanis E, Bosco G, Caprioli M, et al. Atherosclerotic adult Moya-Moya disease in a patient with hyperhomocysteinaemia. *Neurol Sci.* 2007;28:45-47.
9. Tripathi P, Tripathi V, Naik RJ, Patel JM. Moya Moya Cases Treated with Encephaloduroarteriosynangiosis. *Indian Pediatrics.* 2007;44:123-127.
10. Jeong HC, Kim YJ, Yoon W, Joo SP, Lee SS, Park YW. Moyamoya syndrome associated with systemic lupus erythematosus. *Lupus.* 2008;17:679-682.
11. Peltier J, Vinchon M, Soto-Ares G, Dhellemmes P. Disappearance of a middle cerebral artery aneurysm associated with Moyamoya syndrome after revascularization in a child: case report. *Childs Nerv Syst.* 2008;24:1483-1487.
12. Kuroda S, Hashimoto N, Yoshimoto T, Iwasaki Y. Radiological findings, clinical course, and outcome in asymptomatic moyamoya disease: results of multicenter survey in Japan. *Stroke.* 2007;38:1430-1435.
13. Baaj AA, Agazzi S, Sayed ZA, Toledo M, Spetzler RF, van Loveren H. Surgical management of moyamoya disease: A review. *Neurosurg Focus.* 2009;26:E7.
14. Kuroda S, Houkin K. Moyamoya disease: Current concepts and future perspectives. *Lancet Neurol.* 2008;1056-1066.

15. Suzuki J, Kodowa N. Moyamoya disease. *Stroke*. 1983;14:104-109
16. Burke GM, Burke AM, Sherma AK, Hurley MC, Batjer HH, Bendok BR. Moyamoya disease: a summary. *Neurosurgical focus*. 2009;26:E11.
17. Kainth D, Chaudhry SA, Kainth H. Epidemiological and clinical features of moyamoya disease in the USA. *Neuroepidemiology*. 2013;40:282-287.
18. Duan L, Bao XY, Yang WZ. Moyamoya disease in China: its clinical features and outcomes. *Stroke*. 2012;43:56.
19. Fukui M. Guidelines for the diagnosis and treatment of spontaneous occlusion of the circle of Willis ('moyamoya' disease). Research Committee on Spontaneous Occlusion of the Circle of Willis (Moyamoya Disease) of the Ministry of Health and Welfare, Japan. *Clin Neurol Neurosurg*. 1997;99:S238.
20. Karasawa J, Touho H, Ohnishi H, Mujamoto S, Kikuchi H. Cerebral revascularization using omental transplantation for adulthood moyamoya disease. *Neurosurg*. 1993;79:192-196.
21. Kinugasa K, Mandai S, Kamata I, Sugiu K, Ohmoto T. Surgical treatment of moyamoya disease: Operative technique for encephaloduro-arterio-myo-synangiosis, it's follow-up, clinical results and angiograms. *Neurosurgery*. 1993;32:527-531.
22. Miyawaki S, Imai H, Takayanagi S. Identification of a genetic variant common to moyamoya disease and intracranial major artery stenosis/occlusion. *Stroke*. 2012;43:3371.
23. Reis CVC, Safavi-Abbasi S, Zabramski JM, Gusmao SNS, Spetzler RF, Preul MC. The history of neurosurgical procedures for moyamoya disease. *Neurosurg Focus*. 2006;20:E7.