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Histopathologic changes of The Circle of Willis in a Thai patient with Moyamoya disease: A case report

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Moyamoya disease is a vaso-occlusive disease involving the circle of Willis with an increase in collateral blood vessels. The sequelae of the disease are cerebral ischemia in children and intracranial hemorrhage in adults.

A histopathologic study of a 25 year-old Thai woman who died from moyamoya disease is reported. Blood vessels at the circle of Willis displayed eccentric fibrous thickening with prominent internal elastic laminae and an abnormal vascular network.

Key word: *Moyamoya disease.*

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วิเชียร เขาว์ศรีกุล, สุชาติ พุทธิเจริญรัตน์. การเปลี่ยนแปลงทางพยาธิวิทยาของ The Circle of Willis ในผู้ป่วยโรค Moyamoya: รายงานผู้ป่วย 1 ราย. จุฬาลงกรณ์เวชสาร 2544 เม.ย; 45(4): 331 – 6

โรค Moyamoya เป็นโรคที่มีการอุดตันของเส้นเลือดบริเวณ The Circle of Willis พร้อมกับมีการแตกแขนงของเส้นเลือดเพิ่มขึ้น ผลตามมาของโรคนี้ทำให้เกิดสมองขาดเลือดในเด็ก และมีเลือดออกภายในกะโหลกศีรษะในผู้ใหญ่

การศึกษาทางพยาธิวิทยาของผู้หญิงไทย อายุ 25 ปี ซึ่งเสียชีวิตด้วยโรค Moyamoya พบว่าเส้นเลือดบริเวณ The Circle of Willis มีการหนาตัวแบบ eccentric fibrous thickening ร่วมกับ prominent internal laminae และมีเครือข่ายเส้นเลือดที่ผิดปกติ

คำสำคัญ : โรค Moyamoya

Moyamoya disease is a vaso-occlusive disease of the circle of Willis. The most affected part is the distal end of the internal carotid artery. Cerebral angiography displays steno-occlusive changes at the inner portion of the internal carotid artery and a coexisting abnormal vascular network (moyamoya vessels) in the base of the brain. The name moyamoya came from the Japanese language meaning 'misty, foggy, hazy' describing the appearance seen in angiographic studies.⁽¹⁾ The age distribution has two peaks, with a higher peak at five years of age and a lower peak from 30-49 years of age.⁽²⁾ The former is termed juvenile type and usually presents with cerebral ischemia due to steno-occlusive changes of the vessels. The latter is termed adult type, in which the common clinical presentation is cerebral hemorrhage caused by a breakdown of collateral vessels formed at a young age. The main cause of death in both types is cerebral hemorrhage.^(2,3) However, the incidence of intracranial hemorrhage is less frequent in childhood.⁽³⁾

There are few clinical reports about this disease occurring in Thai children and adults.^(4,5) In this study, we describe the histopathologic features of the cerebral vessels in moyamoya disease occurring in a Thai woman.

Case Report

A 25 year - old Thai woman who had been in good health developed low grade fever, cough, and myalgia about two weeks before admission. She also had a dull headache, and occasional amnesia with slow speech. She presented to King Chulalongkorn Memorial Hospital. Physical examination revealed neck stiffness and a positive Kernig's sign. A lumbar

puncture revealed yellowish cerebrospinal fluid with 240 cells/mm³ of white blood cells (lymphocytes 93 %, monocytes 7 %), 58 mg/dl of glucose, and 0.24 g/dl of protein was found. Computed tomography (CT) of the brain displayed left basal ganglia hemorrhage with brain edema, but no midline shift was noted.

After admission, she was drowsy and had impaired cognition, in calculation as well as short term memory. Repeated CT of the brain demonstrated the same result, as previously. The findings on four vessel angiography were compatible with moyamoya disease. The last CT of the brain illustrated infarction of both parieto-occipital regions with brain edema. She was declared brain dead after eight days of admission.

An autopsy was performed. The brain weighed 1,350 grams and exhibited severe bilateral uncal and cerebellar tonsillar herniations. The circle of Willis showed an unusual vascular network branching off from the cerebral blood vessels. Serial sections of the brain revealed intraventricular hemorrhage with mild subarachnoid hemorrhage, softening of bilateral parieto-occipital areas and secondary (Duret's) hemorrhage.

Blood vessels for light microscopy were fixed in 10 % neutral formalin. Sections were cut at five micron thick and stained with hematoxylin and eosin, Masson's trichrome, and elastic van Gieson (EVG) stains.

Eccentric fibrous intimal thickening (Figure 1.) with duplicated internal elastic laminae (Figure 2.) of the cerebral arteries and the distal parts of the internal carotid arteries were observed. Near these vessels, proliferation of an abnormal collateral vascular network was demonstrated. Neither inflammation nor vasculitis was detected. The ruptured site causing the hemorrhage could not be found.



Figure 1. Histopathology of the left anterior cerebral artery. Eccentric fibrous intimal thickening is demonstrated (EVG stain, original magnification x 40).



Figure 2. The left internal carotid artery shows duplicated elastic laminae (EVG stain, original magnification x 100).

According to the guidelines for diagnosis of moyamoya disease⁽²⁾ modified and established in 1996 by the Research Committee on Spontaneous Occlusion of the Circle of Willis (moyamoya disease), the Ministry of Health and Welfare, Japan, the findings in this reported case were moyamoya blood vessels.

Discussion

Our study of the histopathologic vascular changes showed the same results as other previous findings.⁽⁶⁻⁸⁾ The affected vessels displayed eccentric intimal fibrous thickening and multilayered elastic laminae with the formation of an abnormal vascular network. Neither inflammation nor underlying vascular disease was present. Studying the abnormal vascular network in moyamoya disease, Kono and co-workers revealed no difference in vascular density and arterial/venous ratio between normal leptomeningeal vessels and moyamoya vessels.⁽⁹⁾ Kono et al. concluded that the changes were probably due to dilated preexisting

vessels and not newly formed blood vessels.⁽⁸⁾

The etiology of moyamoya disease is still unknown. Yashiro et al. performed 31 cases and demonstrated thrombotic lesions in 17 cases. They believed that thrombi play an important role at least in progression of the vascular obstruction.⁽⁶⁾ Some investigators have postulated that inflammatory and autoimmune processes are responsible for the pathogenesis.⁽¹⁰⁾ Tanigawara et al. showed a relationship between viral infection and the disease. Serum antibody to the Epstein-Barr virus (EBV) was detected more frequently in patients with moyamoya disease than in normal controls.⁽¹⁰⁾ Yamada and colleagues investigated the effects of *Propionibacterium acne* infection on intracranial internal carotid arteries in rats and found coarse, disrupted duplicated internal elastic lamina.⁽¹¹⁾ Because of the higher incidence of the disease in Japanese, the very low incidence in Caucasians, and a familial occurrence including identical twins, a genetic study on moyamoya disease

is now being undertaken in Japan.⁽³⁾ Inoue et al. exhibited several alleles of class II genes of human leukocyte antigen (HLA) associated with moyamoya disease.⁽¹²⁾

Intracranial hemorrhage in adults is much more frequent than in children and may be a lethal condition.^(2,3,7,13) Medial fibrosis and attenuation of the wall are probably primary and closely related to the cause of ruptured moyamoya vessels.⁽⁷⁾ Mauro et al. proposed a role for lipohyalinosis and miliary aneurysms in intracranial hemorrhage.⁽¹³⁾ Kodama and Suzuki speculated that microaneurysms resulting from fragility and localized disruption of the vascular wall due to continuous and insidious ischemia could cause intracerebral bleeding.⁽¹⁴⁾ Iwama et al. found that a ruptured aneurysm was the cause of rebleeding at the same site in patients with moyamoya disease. However, neither ruptured aneurysm nor vascular abnormalities was identified in those patients who had rebleeding at a different site.⁽¹⁵⁾

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