

## Case Report

# A Case of Moyamoya-like Vessels with History of Brain Radiation

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### Abstract

A 13 years old boy underwent surgical operation of craniopharyngioma which followed by postoperative radiation therapy. Eighteen months later he was admitted to the neurological ward due to sudden sensory aphasia. Brain CT showed infarction of left hemisphere. Since no other predisposing factor was present, the cause of stroke in this patient can be related to Moyamoya - like disease after radiation.

**Keywords:** Moyamoya, Stroke, Aphasia, Craniopharyngioma, Radiation Therapy

**M**oyamoya diseases is an unusual form of chronic cerebrovascular occlusive disease that is characterized by angiographic findings of bilateral stenosis or occlusion at the terminal portion of the internal carotid artery together with the abnormal vascular network at the base of the brain<sup>1, 2, 3</sup>. The term Moyamoya means 'puff of smoke' in Japanese that used to describe the appearance of multiple collateral vessels, usually lenticulostriate and thalamostriate vessels, seen in cerebral angiography that is hallmark of this condition. This disease was first diagnosed by Jiro Suzuki in the 1963 and Takeuchi and Suzuki described this condition affecting Japanese families in 1969<sup>3</sup>. Extensive investigations on patients with these characteristic angiographic findings have been conducted<sup>4</sup>. As a result, the clinical entity of this disease and its concept has now been established. It is well-known for example that progression of stenosis or occlusion of the intracranial major arteries including distal ends of the internal carotid arteries is the primary lesions of this disease, and that the abnormal vascular network (Moyamoya) at the base of the brain is their collateral secondary to brain ischemia<sup>4</sup>.

### Case History

A 13 years old male patient was admitted with sudden speech problem. He was evaluated because of short stature. The cause of short stature was craniopharyngioma (figure 1) and he had undergone postoperative radiotherapy after craniopharyngioma surgery about 18 months before admission. Neurological examination revealed sensory aphasia in addition to short stature but otherwise was normal. All routine and specific laboratory examinations including ECG, echocardiography, homocystin level, vasculitis tests, peripheral blood smear and antiphospholipid anticardiolipin titer were normal. Brain CT Scan and MRI revealed infarction of left hemisphere (figure 2). Cerebral angiography revealed obstruction in the distal part of both siphones and multiple stenoses in the circle of Willis with large ischemic process in the left hemisphere (figure 3 and 4).

### Discussion

Moyamoya disease is a rare cause of stroke in children and adults. The onset of the disease is about five years olds at the maximum, and next in the fourth decade. Females are more commonly affected than males. The etiology of this disease is not known but it sometimes occurs in connection

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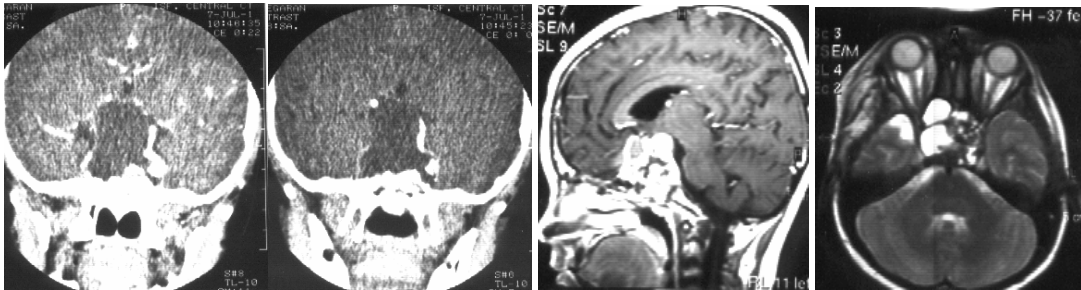
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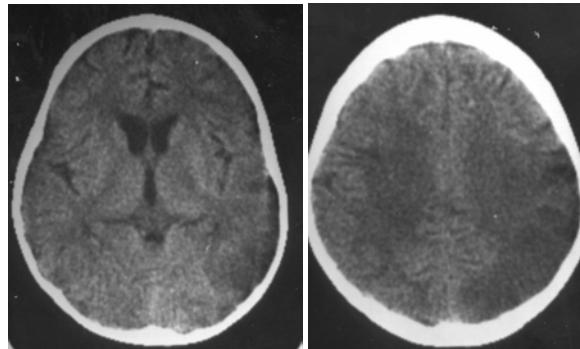
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with underlying conditions including arteritis, sickle cell disease, Down's syndrome<sup>5</sup>, neurofibromatosis, Marfan's syndrome, Bacterial infections particularly nasopharyngeal infections, and radiation<sup>6, 7</sup>. In this case there is a positive history of postoperative radiation. The cause of speech problem is sensory aphasia due to obstruction of inferior branch of middle cerebral artery and Wernike area involvement. In this case all causes of stroke were ruled out. Therefore stroke can be related to vasculopathy. The supraclinoid region of the internal carotid seems especially vulnerable in children. Occlusive vascular disease, similar in angiographic appearance to Moyamoya disease, and presenting with ischemic events and stroke, is not uncommon after radiotherapy<sup>8</sup>. The angiography revealed obstruction in the distal part of both

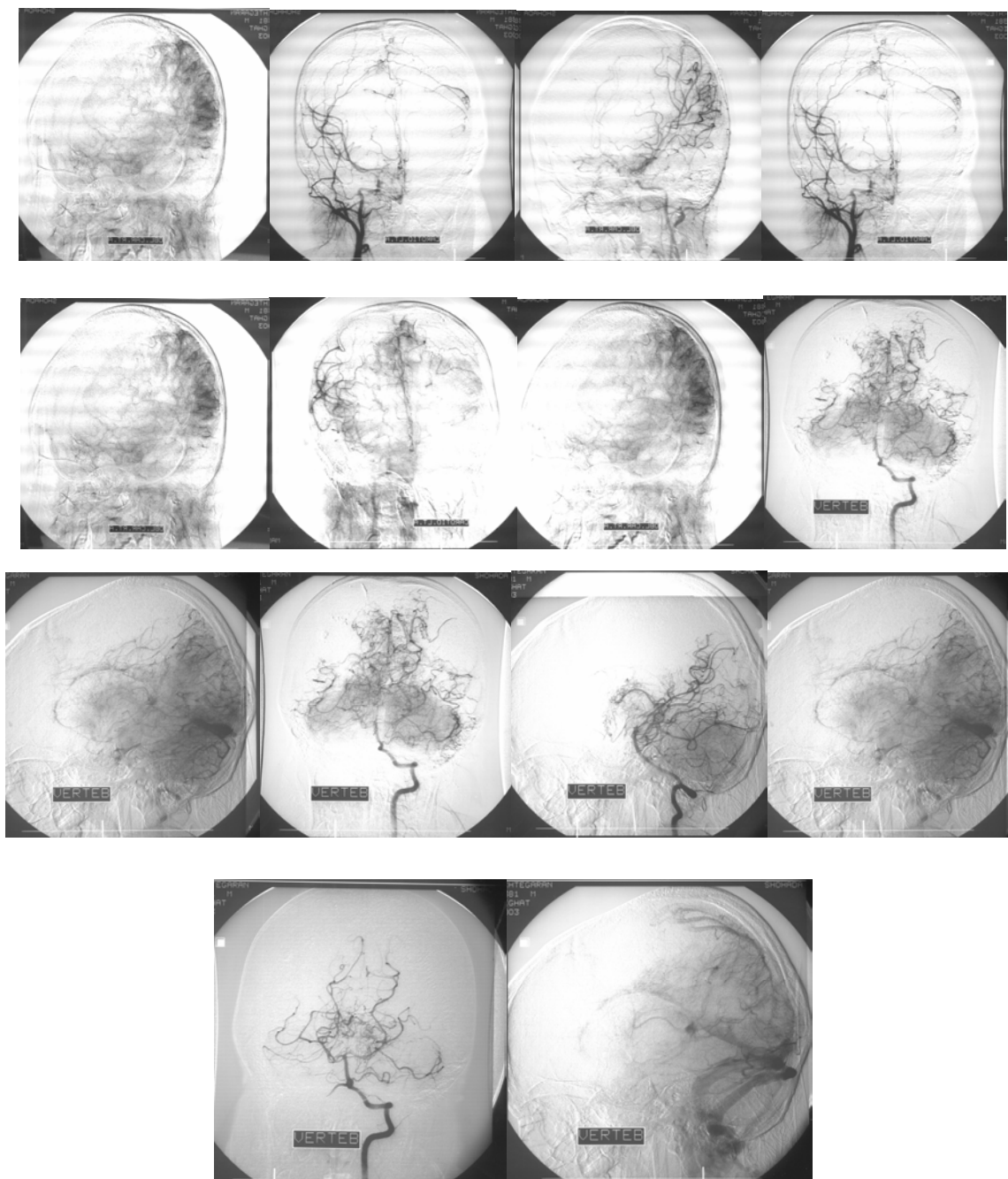
siphons and multiple stenosis in the circle of Willis with large ischemic process in the left hemisphere that suggests Moyamoya-like arteropathy. Because there is no clear factor related to Moyamoya disease therefore the cause of stroke in this patient can be related to Moyamoya-like disease due to radiation. Extensive similarities between irradiation-induced cerebral vasculopathy and primary Moyamoya syndrome support the notion that both disorders share common pathophysiological mechanisms. The occurrence of Moyamoya-like vascular changes may not depend on specific trigger mechanisms but may rather represent a nonspecific response of the developing vascular system to a number of various noxious events such as radiation<sup>9</sup>.



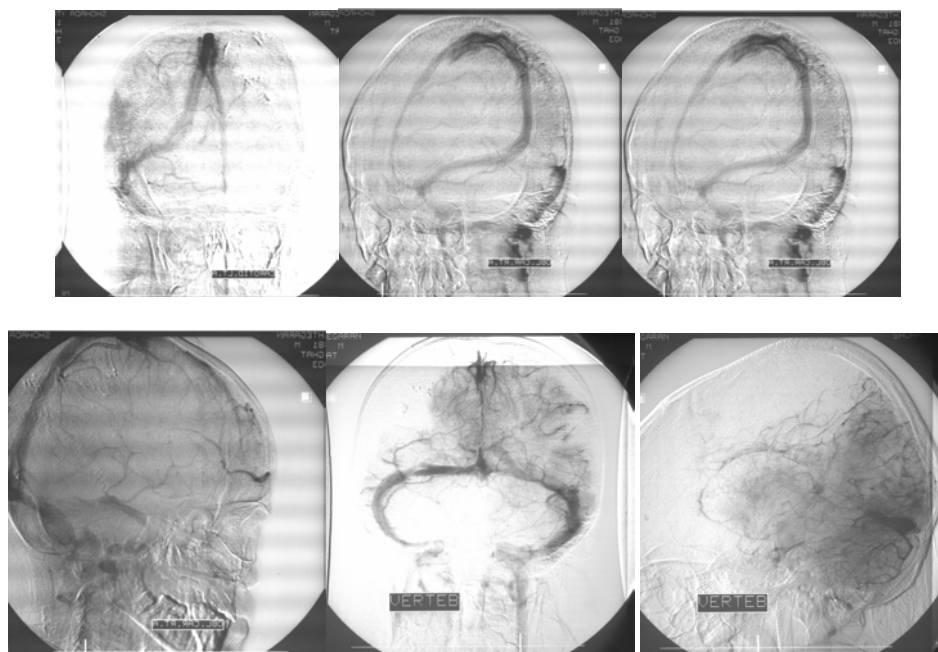
**Figure 1.** CT-scan and MRI of the patient in this report which shows craniopharyngioma. From left to right: CT with contrast; CT without contrast; MRI sagittal view; MRI axial view.



**Figure 2.** CT-scan of the patient in this report showing infarction of the left parieto-temporal lobe.



**Figure 3.** Arterial phase of Angiography of the patient revealed obstruction in distal part of both siphones and multiple stenosis in the circle of Willis with large ischemic process in the left hemisphere. Note the puff-smoke pattern particularly in the upper-left corner image.



**Figure 4.** Venous phase of angiography.

## References

1. Mingli RAO , et al . Clinical and experimental pathology of Moyamoya disease. *Chin Med J* 2003;116(12):1845-1849.
2. Khan N, Schuknecht B, Boltshauser E, Capone A, Buck A, Imhof HG, Yonekawa Y. Moyamoya disease and Moyamoya syndrome: experience in Europe; choice of revascularisation procedures. *Acta Neurochir (Wien)* 2003;145(12):1061-71.
3. Biller J, Betsy BL. Ischemic cerebrovascular disease . In:Walter G. Bradly , Robert B Daroff, Gerald M. Fenichel, Joseph Jankovic, editors. *Neurology in clinical practice, vollum II, 4nd ed.*Boston. Butterworth Heinemann. 2004:1217-1218.
- 4..Suzuki J, Takaku A. Cerebrovascular Moyamoya disease : a disease showing abnormal net – like vessels in base of brain. *Arch Neurol* 1969;20(3):288-99.
5. Seeler RA ,Royal JE , Powe L et al : Moyamoya disease in children with sickle cell anemia and cerebrovascular occlusion. *J Pediatr* 1978; 93(5):808-10.
6. Victor M, Ropper AH , Adams RD ,moyamoya disease. In : Adams and Victor,editors. *Principles of Neurology , Vol 2. 7<sup>nd</sup> ed.* New York.McGraw –Hill. 2001:877-878.
- 7.Suzuki J, Kodama N . Moyamoya disease : a review. *Stroke* 1983;14(1):104-9.
8. Arnold P. Gold, ABBA L. Cargan . stroke in children. In:Lewis P.Rowland , editor. *Merritt's Neurology.*10<sup>nd</sup> ed. New York .Lippincott Williams & Wilkins.2000: 254 .
9. .Kestle JR, Hoffman HJ, Mock AR. Moyamoya phenomenon after radiation for optic glioma. *J Neurosurg* 1993;79(1):32-5.
10. Alfredo D . Voloschin , Tracy T , Batchelor , and Jeffrey C .Allen . Management of primary nervous system tumors in infants and children. In: Walter G. Bradly , Robert B Daroff, Gerald M. Fenichel, Joseph Jankovic, editors. *Neurology in clinical practice, vollum II. 4<sup>nd</sup> ed.*Boston. Butterworth Heinemann.2004:1435.