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Moyamoya Disease Presenting with Intracranial Haemorrhage

Pages with reference to book, From 349 To 351

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Moyamoya disease is a spontaneous occlusion of the circle of Willis with abnormal compensatory anastomotic vascular networks at the base of the brain. Early reports characterised the condition as a cause of cerebral ischaemia with progressive neurological deficits in infants and children or intracranial haemorrhage in adolescents and adults of Japanese Community^{1,2}. However, it is now well known to occur in the other parts of the world as well - though being contingent on angiographic findings, the diagnosis depends on the level of the nation's health care provision. Two cases of moyamoya diseases admitted to neurosurgical unit with intracranial haemorrhage are being presented here.

Case Reports

Case 1

This previously healthy 38 year old male suffered an acute episode of headache with vomiting and within few minutes became unresponsive and was deeply unconscious. He was admitted to the district hospital where on conservative management he regained consciousness after 7 days without focal signs. However, he again suffered a sudden attack of headache with vomiting resulting in unconsciousness. Thirty-six hours after improvement from the initial episode, the patient was then shifted to our hospital.

On admission his vital signs were normal and general physical examination was unremarkable. He was unconscious but responding to painful stimuli (Glasgow Coma Score 10/15). Computerized Tomography (CT) Scan showed an intraventricular haemorrhage. Carotid angiography revealed the occlusion of the supraclinoid portion of the internal carotid arteries on both sides. There were foetal type posterior cerebral arteries and fine anastomotic vessels in the form of rete, irrigating the territory of anterior Circle of Willis characteristic of moyamoya disease (Figure 1A, B, C).

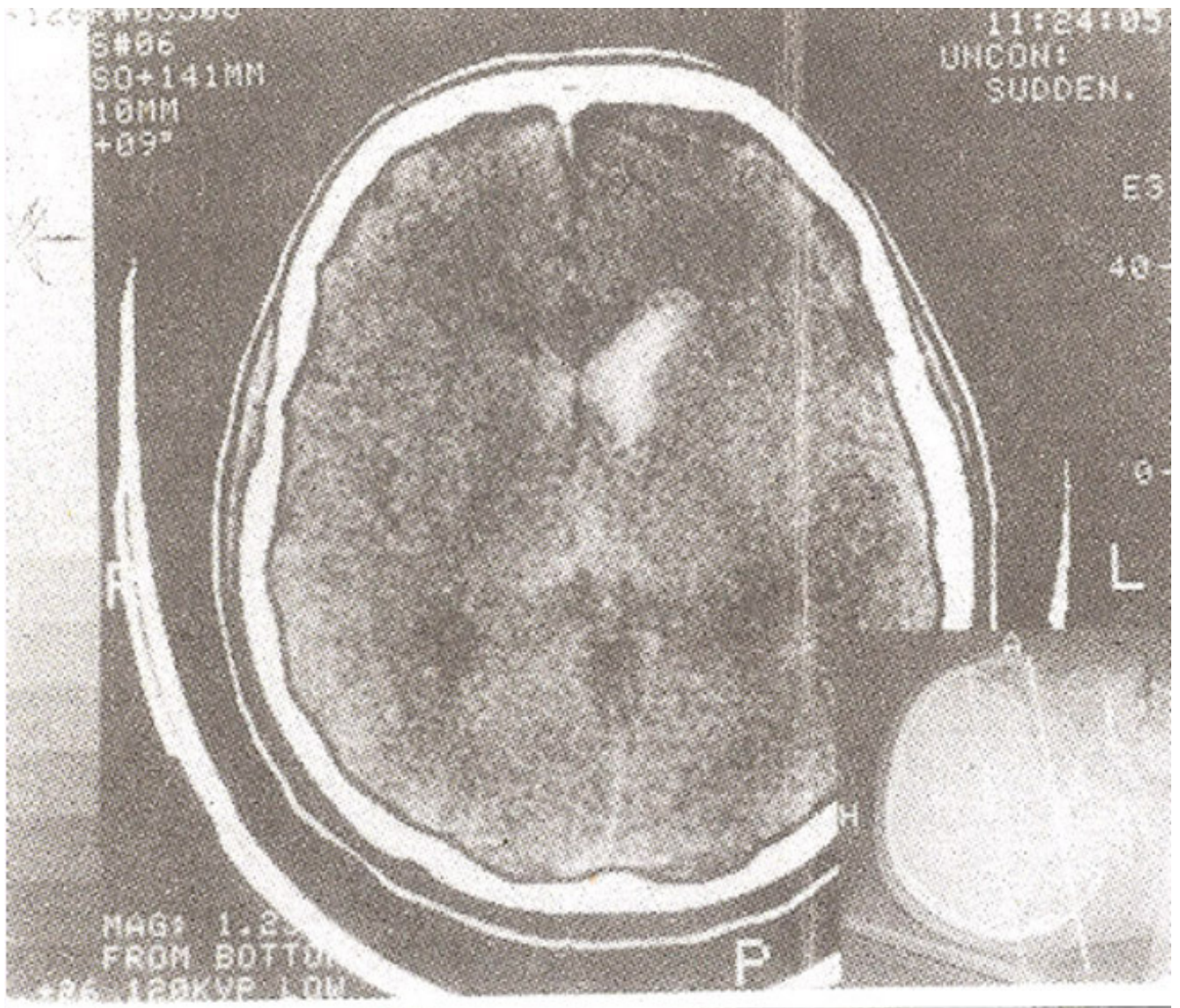


Figure 1A. CT scan showing intra ventricular haemorrhage.

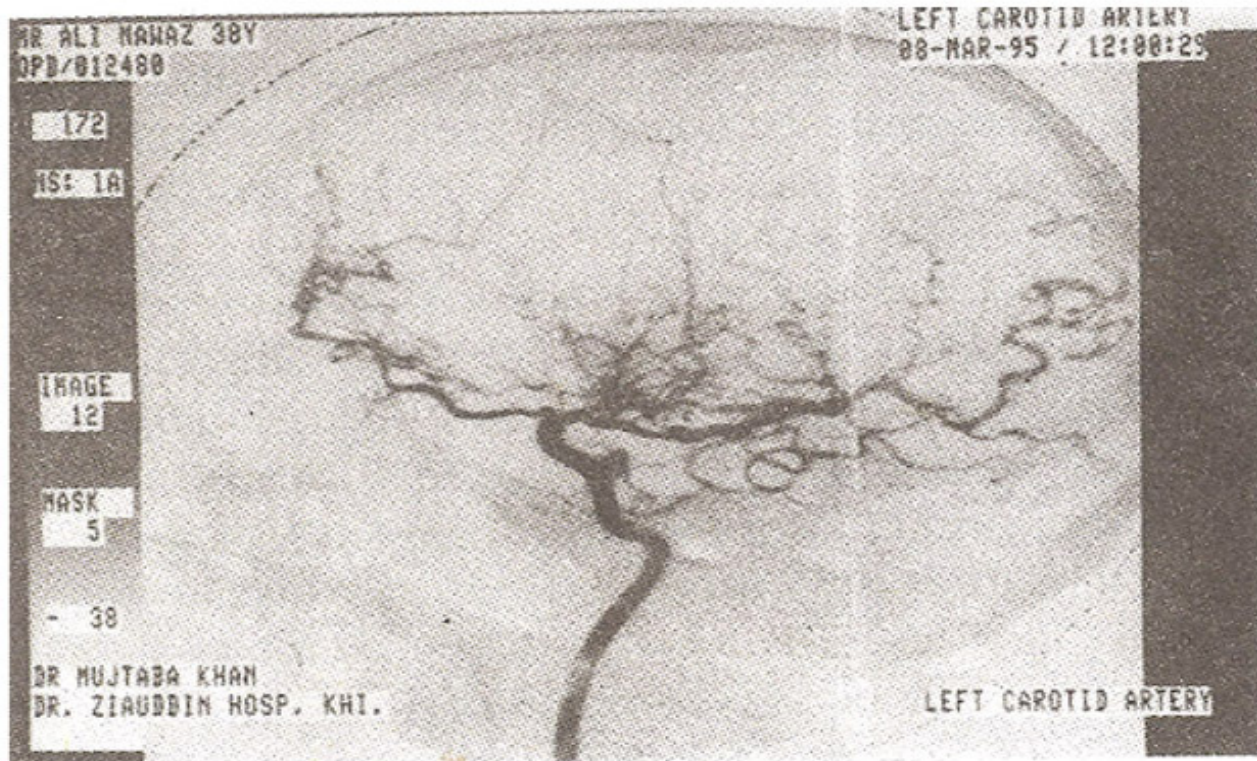


Figure 1 B & C. Angiogram revealing fine anastomotic vessels along with foetal type posterior cerebral artery typical of moyamoya disease.

The patient's condition gradually improved with conservative management and he was discharged 15 days after being fully conscious and mobile.

Case 2

This previously healthy 25 years old man experienced a sudden attack of severe headache, with nausea

and vomiting. Twelve hours later he developed neck stiffness and became drowsy. He was taken to a near by hospital where spinal tap yielded bloody cerebrospinal fluid. He was immediately referred to our department. On admission his vital signs were normal and his general physical status was good. He was drowsy (Glasgow Coma Score 13/1 5) had stiff neck and right sided haemiparesis. CT Scan showed a small left intracerebral haematoma along with subarachnoid haemorrhage. Carotid angiogram demonstrated occlusion of the supra clinoid carotid artery with profuse anastomosis between the ophthalmic artery and intracranial vessels, in the manner of rete typical of moyamoya disease (Figure 2A, B, C).

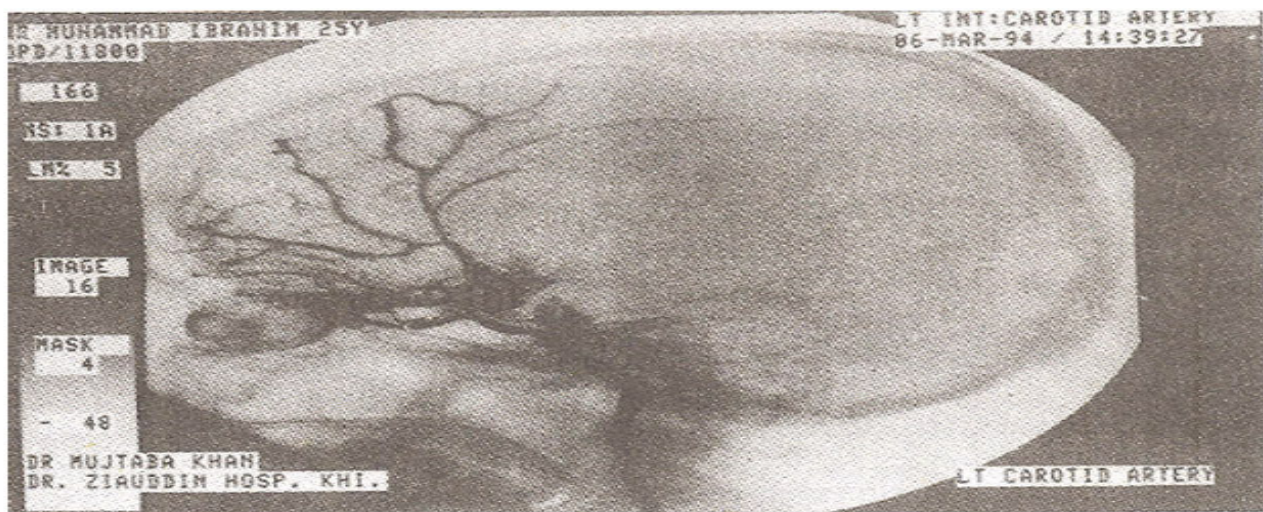
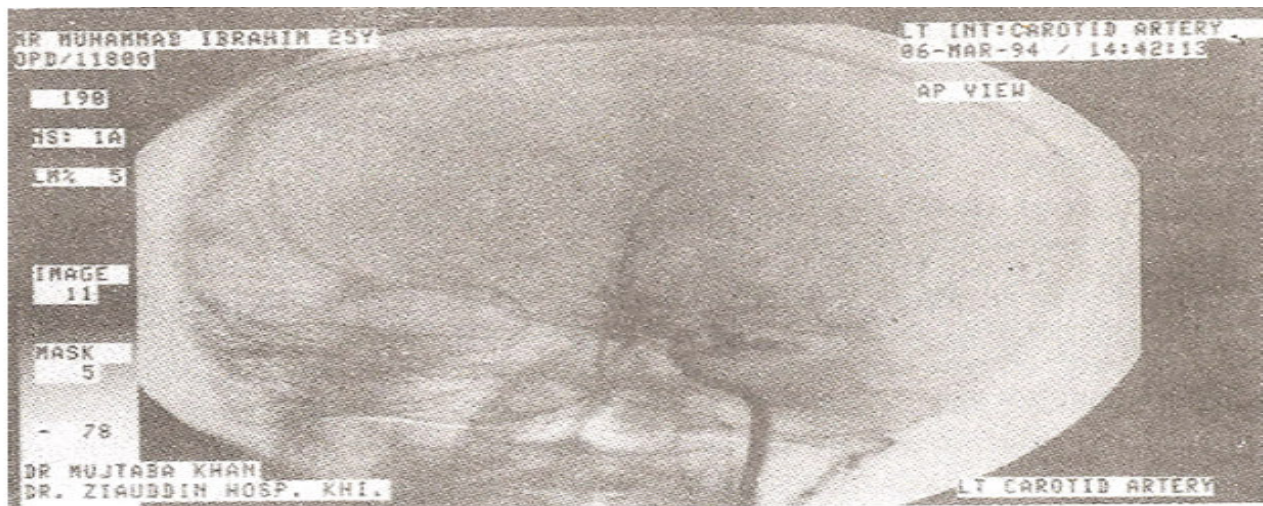


Figure 2 ABC, Anteroposterior and lateral views of left internal carotid artery angiogram revealing occlusion of supra clinoid artery with profuse anastomosis between the ophthalmic artery and intracranial vessels typical of moyamoya disease.

Non-surgical management resulted in return of consciousness over 20 days and when seen at six month follow-up he was without deficit though the family reported aggressive behaviour.

Discussion

The term moyomoya means “puff of smoke” in Japanese and the term was first used by Suzuki et al³, for a disease involving occlusion of moderate sized arteries at or near the Circle of Willis. Such occlusive basal vasculopathies are chronic and are accompanied by a pathologic attempt at collaterization, involving proliferation of arterioles and small penetrating arteries near the perforating substance. Such proliferation of deep collaterals provides the typical angiographic appearance of a “puff of smoke”. There may be other attempts at collaterization also via transdural collateral channels. The aetiology of the disease is still not well established. It has been hypothesized that the carotid stenosis to be the primary condition and moyamoya vessels to be the result of it^{1,4,5}, while others believe that the disease results from progressive occlusion of vessels with stenosis initially involving internal carotid artery then reaching the anterior and middle cerebral arteries and at last involving the communicating and posterior cerebral arteries along with an abnormal vascular network of moyamoya vessels^{1,3}.

This disease manifests in two forms, Juvenile form characterised clinically by seizures or progressive neurological deficits due to ischemia and infarctions and the adults form, usually made manifest by intracranial haemorrhage, which may be subarachnoid, intracerebral or intraventricular. Subarachnoid haemorrhage may result from rupture of basal pathologic perforating vessels which are highly friable or ruptured aneurysms or pseudoaneurysms⁶⁻⁸. Moyamoya is a cause of 4-5% ischaemic strokes in paediatric age groups⁹. Many children who have recurrent ischaemic strokes develop progressive mental deterioration¹⁰.

The purpose of reporting these two cases is that this disease does exist in our community but due to lack of diagnostic facilities, the paediatric cases are not being diagnosed, only adult patients who present with strokes are being referred to tertiary care hospitals for investigations and management. Hence children who present with neurologic deficits and strokes should be accurately evaluated and referred to neurosurgical centres for revascularization procedures.

References

1. Handa J, Handa H. Progressive cerebral arterial occlusive disease, an analysis of 27 cases. *Neuroradiology*, 1972;3:119-33.
2. Carlson CB, Harvey FH, Loop J. Progressive alternating hemiplegia in early childhood with basal arterial anastomosis and telangiectasia (moyamoya syndrome). *Neurology*, 1973;23:734-44.
3. Suzuki J, Takaku A. Cerebrovascular ‘moyamoya’ disease. Disease showing abnormal net like vessels in the base of the brain. *Arch. Neurol.*, 1969;20:288-99.
4. Kudo T. Spontaneous occlusion of the circle of Willis. *Neurol. Surg.*, 1975;3:711-24.
5. Vulia O, Alexianu M, Gabor S. Hypoplasia and obstruction of the circle of Willis in a case of atypical cerebral haemorrhage and its relationship to the Nishimoto’s disease. *Neurology*, 1970;20:361-67.
6. Oka K, Yamashita M, Sadoshima S, et al. Cerebral haemorrhage in moyamoya disease at autopsy. *Virchows Arch.*, 1981;392:247-51.
7. Jun-Ichiro H, Nobuo H, Tetsuya T. Moyamoya disease with repeated intraventricular haemorrhage due to aneurysm rupture. Report of two cases: *J. Neurosurg.*, 1994;80:328-31.
8. Hiroshi Y, Sumitak T. Cerebrovascular moyamoya disease associated with an intracranial pseudoaneurysm. *J. Neurosurg.*, 1982;56:131-34.
9. Schoenberg B, Mellinger J, Schoenberg DG. Cerebrovascular disease in infants and children. A study of incidence. Clinical features and survival. *Neurology*, 1978;28:763-68.
10. Ishii R, Takeuchi S, Ibayashi K, et al. Intelligence in children with moyamoya disease. Evaluation after surgical treatment with special reference to changes in cerebral blood flow. *Stroke*, 1984; 15:873-77.