Moyamoya Disease Presenting with Intracranial Haemorrhàge

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Ali Akbar,Aftab Qureshi,Rashid Jooma (Department of Neurosurgery, Jinnah Postgraduate Medical Centre, Karachi.)

Moyamoya disease is a spontaneous occlusion of the circle of Willis with abnonnal compensatory anastamotic 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 depend 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 nonnal and general physical examinationwas 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 supra clinoid portion of the internal carotid arteries onboth sides. There were foetal type posterior cerebral arteries and fme anastamotic vessels in the form rete, irngating the territory of anterior Circle of Wills characteristic
of moyamoya disease (Figure 1A, B, C).

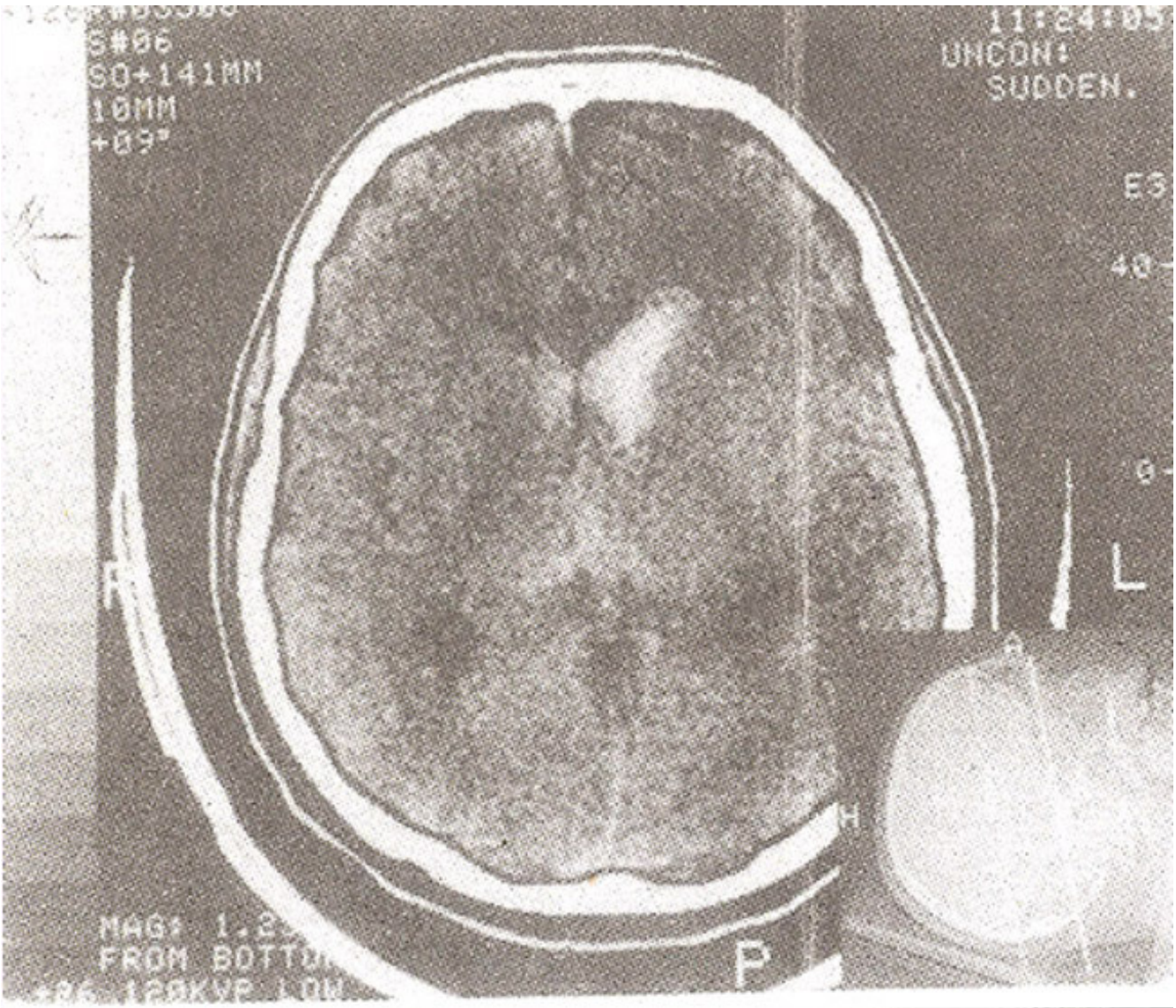

Figure 1A. CT scan showing intra ventricular haemorrhage.

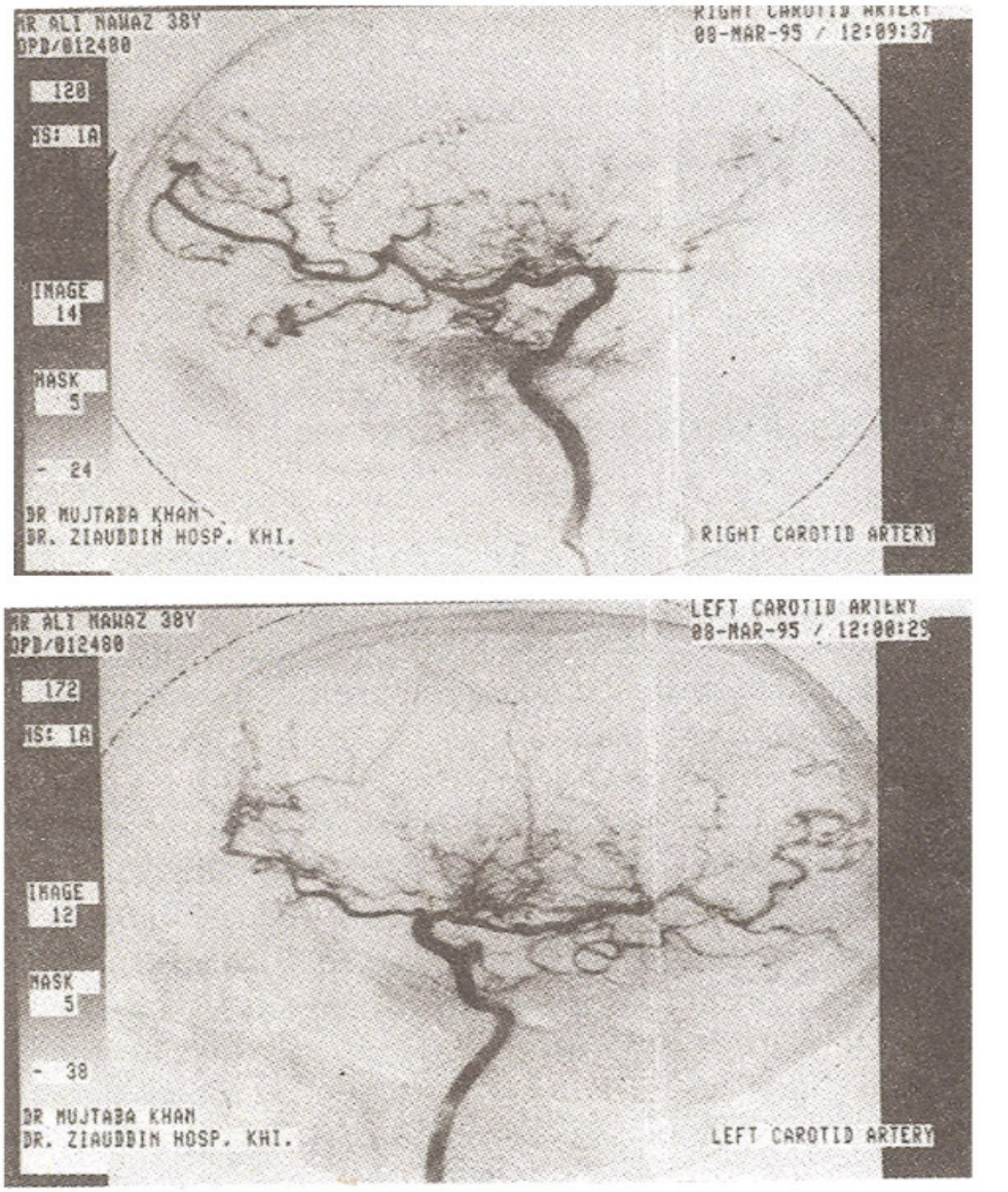

Figure 1 B & C. Angiogram revealing fine anastamotic vessels along with foetal type posterior cerebral artery typical of moyamoya disease.

The patient's condition gradually improved withconseivative management and he was discharged 15 days afterbeing 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 stuffiness and became drowsy. He was taken to a near by hospital where spinal tap yielded bloody cerebrospinal fluid. He was mimediately referred to our department. On admission his vital signs were nonnal 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).

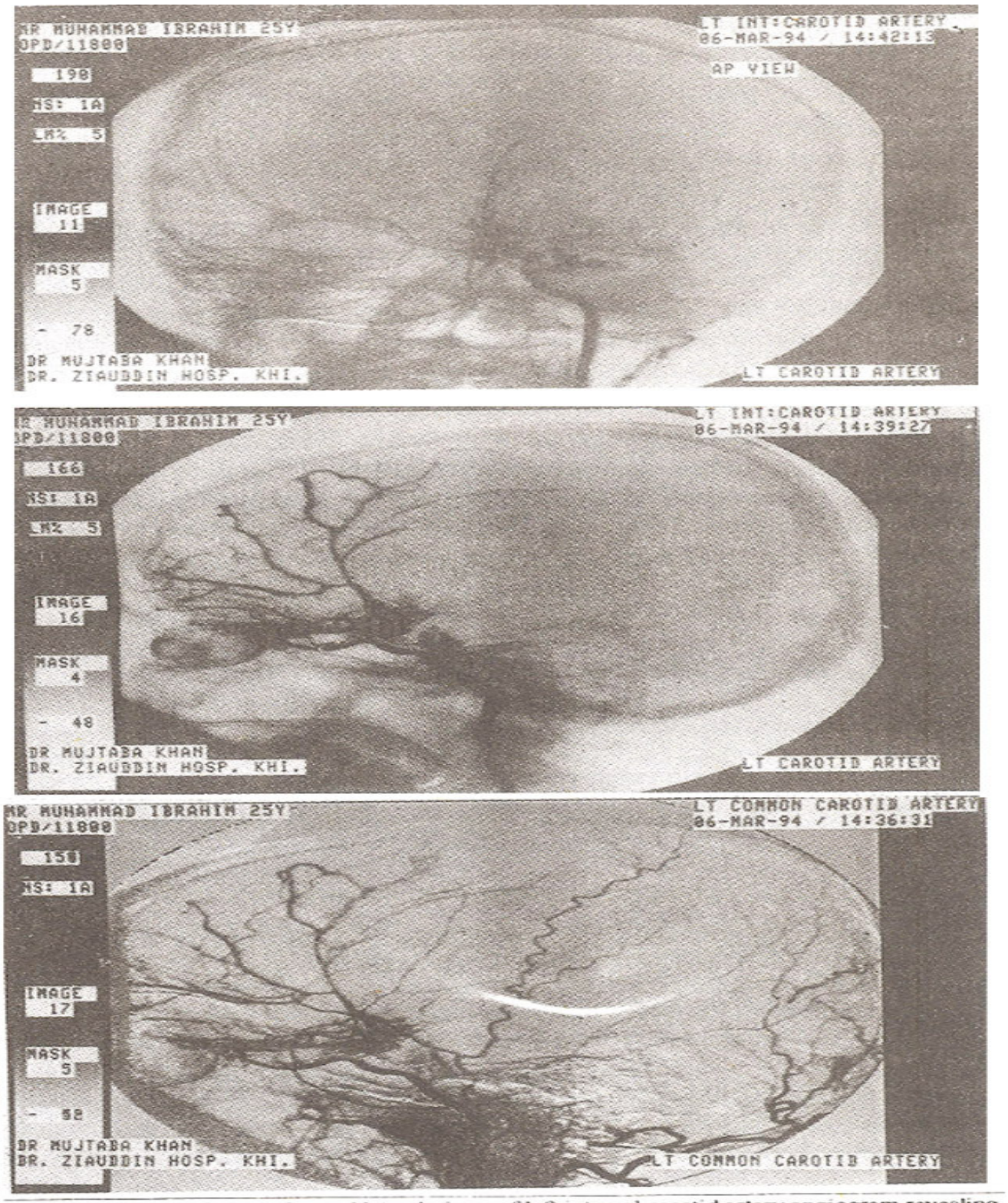

Figure 2 ABC, Anterioposterior and lateral views of left internal carotid artery angiogram revealing occlusion of supra clinoid artery with profuse anastamosis between the ophthalmic artery and intracranial vessels typical of moyamoya disease.

Non-surgical management resulted in return of consciousness over 20 days and when seen at six month follow-up be 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 al[3], 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 collatemlization, involving proliferation of arterioles and small penetrating artenes nearthe perforating substance. Such proliferation of deep collaterals provides the typical angiographic appearance of a “puff of smoke”. There may be other attempts atcollateralization 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 primaiy 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 antenor and middle cerebral arteries and at last involving the communicating and posterior cerebral arteries along with an abnormal vescular net work 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 pseudoaneurysms[6-8]. Moyamoya is a cause of 4-5% ischaemic strokes inpaediatnc age groups[9]. Many children who have recurrent ischaemic strokes develop progressive mental deterioration[10].

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.

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