Imaging of Moya Moya Disease

Pages with reference to book, From 181 To 185

Rashid Ahmed, Hurnera Ahsan (Liaquat National Hospital, Karachi.)

Abstract

Moya Moya disease is a rare disease causing occlusion of the supraclinoid portions of the internal carotid arteries. The CT, MRI and Angiography findings of four patients of Moya Moya disease are presented. CT revealed presence of infarction in the bilateral cerebral hemispheres and atrophy in all patients who had CT. CT was also able to detect abnormal vessels at basal ganglia in one patient. MRI was more informative and besides showing the infarctions and atrophy it also, revealed abnormal Moya Moya vessels in all patients. Cerebral angiography is the most definitive method of diagnosis. It showed occlusion of supra clinoid portion of internal carotid arteries on both sides in three patients and on one side in one patient. It also showed pressure of Moya Moya vessels in all cases. It also showed collaterals from meningeal and ophthalmic arteries in all cases. Imaging findings of Moya Moya disease are very specific and provide early diagnosis (JPMA 47:181,1997).

Introduction

Moya Moya disease is a rare cerebrovascular occlusive disease of unknown origin[1,2]. Although it is morn commonly seen in Japan, cases have also been reported elsewhere[3,4]. We present CT, MRI and Angiographic finding in four patients with Moya Moya disease.

Patients and Methods

Four patients of Moya Moya disease were studied from 1992 to 1996. Three were females and one was male. Age ranged from 3 years to 8 years. In three patients CT, MRI and Angiography was performed while in one patient only angiography was done. CT scans were obtained on Shimadzu Scanner SCT-5000T. Ten mm axial images were obtained with and without contrast. MRI were obtained on HITACHI 0.2 Tesla units. Both TI and T2 weighted images and proton density images were obtained in axial and coronal planes. Contrast was not used in any of the patients. Angiograms were done on Philips DSA unit. Ionic contrast was used in all patients. A catheter was introduced through transfemoral route and bilateral carotid and right or left vertebral angiograms were obtained.

Results

Results of CT MRI and Angiographic findings are summarized in Table I, II and III.

Table I. Summary of CT findings.

Case No.	Age/Sex	Date of CT	Interval between onset and CT	Focal OW Density area representing infarctions	Atrophy of brain	Ventricular dilatation	Visualization of circle of Willis	Tortuous vessels at basal ganglia MMV'S
1	3 years Male	12/11/94	6 months	Bilateral fronto parietal region more marked on left side	Minimal brain atrophy	Minimal	Not seen	In bilateral basal ganglia and lateral ventricular walls
2	5 years Female	25/6/96	18 months	Infarction in the Bilateral frontal regions hemorrhagic on left side	Moderate Brain atrophy	Moderate	Not seen	Not seen
3	8 years Female	25/12/95	2 Months	Right frontal right parietal and left parietal region	Mild to Moderate	Moderate	Not seen	Not seen
4	8 years Female	CT not done						

Table II. Summary of MRI findings.

Case No.	Age/Sex	Date of MRI	Interval between onset and MRI	Visualization of in for CT low intensity on T1 & 1 high on T2	Atrophy	Ventricle dilatation	Abnormal tortuous vessels at basal ganglia MMV'S
1	3 years Male	15/11/94	6 months	Cortical infarcts in bilateral fronto parietal region	Minimal	Minimal	Present in basal ganglia and periventricular regions of lateral ventricles
2	5 years Female	25/6/96	18 months	Bilateral frontal infarction hemorrhagic on left side	Moderate	Moderate	Minimal in basal ganglia region
3	8 years Female	28/1/96	2-1/2 months	Infarction in right frontal and bilateral parietal region	Mild to Moderate	Moderate	Minimal in basal ganglia region
4	8 years Female	MRI Not done					

Table III. Summary of angiographic findings.

C.No.	Age/Sex	Occlusion of MCA and ACA	Abnormal vessels MMV	Visualization of medullary vessels vessels and external carotid system	Anastomosis between MMV and abnormal	Associated aneurysms	Involvement of posterior circulation
1	3 years Male	Occlusion of supra clinoid portion of on both sides	Present enlarged anterior choroidal vessels	Medullary vessels present	Filling of pericollosal arteries through prominent middle meningeal and ophthalmic arteries	None	Involvement of post circulation filling of MMV from posterior circulation
2	5 years Female	Occlusion of supra clinoid portion of int. carotid artery on both sides	Minimaly dilated anterior choroidal vessels	Not seen	Collaterals between MMV and middle meningeal and opthalmic arteries	Multiple present in posterior circulation	Filling of MCA through vertebral on both sides
3	8 years Female	Occlusion of supra clinoid portion of internal carotid arteries on both sides	Minimally dilated anterior choroidal vessels	Not seen	Meningeal collaterals present opthalmic and transosseous through superficial temporal arteries	Present	Involvement of posterior circulation seen filling of both MCA from vertebral filling of MMV also seen from posterior circulation
4	8 years Female	Complete occlusion of supra clinoid portion of int. carotid art. on one side (left side)	MMV in left basal ganglia	Not seen	Laptomeningeal collaterals	None	Not done

The CT scans and MRI imaging findings were reviewed for signs of infarctions, hemorrhages, atrophy, ventricular size, visualization of circle of Willis and Moya Moya vessels. Angiographic findings were reviewed for Steno-occlusive disease involving the internal carotid, anterior cerebral, middle cerebral and posteriorcerebral vessels, presence of dilated tortuous vessels at basal ganglia (Moya Moya vessels), evidence of medullary arteries, evidence of collaterals through branches of middle meningeal vessels and ophthalmic artery and for presence of aneurysms. Infarctions in bilateral cerebral hemispheres were present in all patients who had CT scan or MRI (Figure 1).

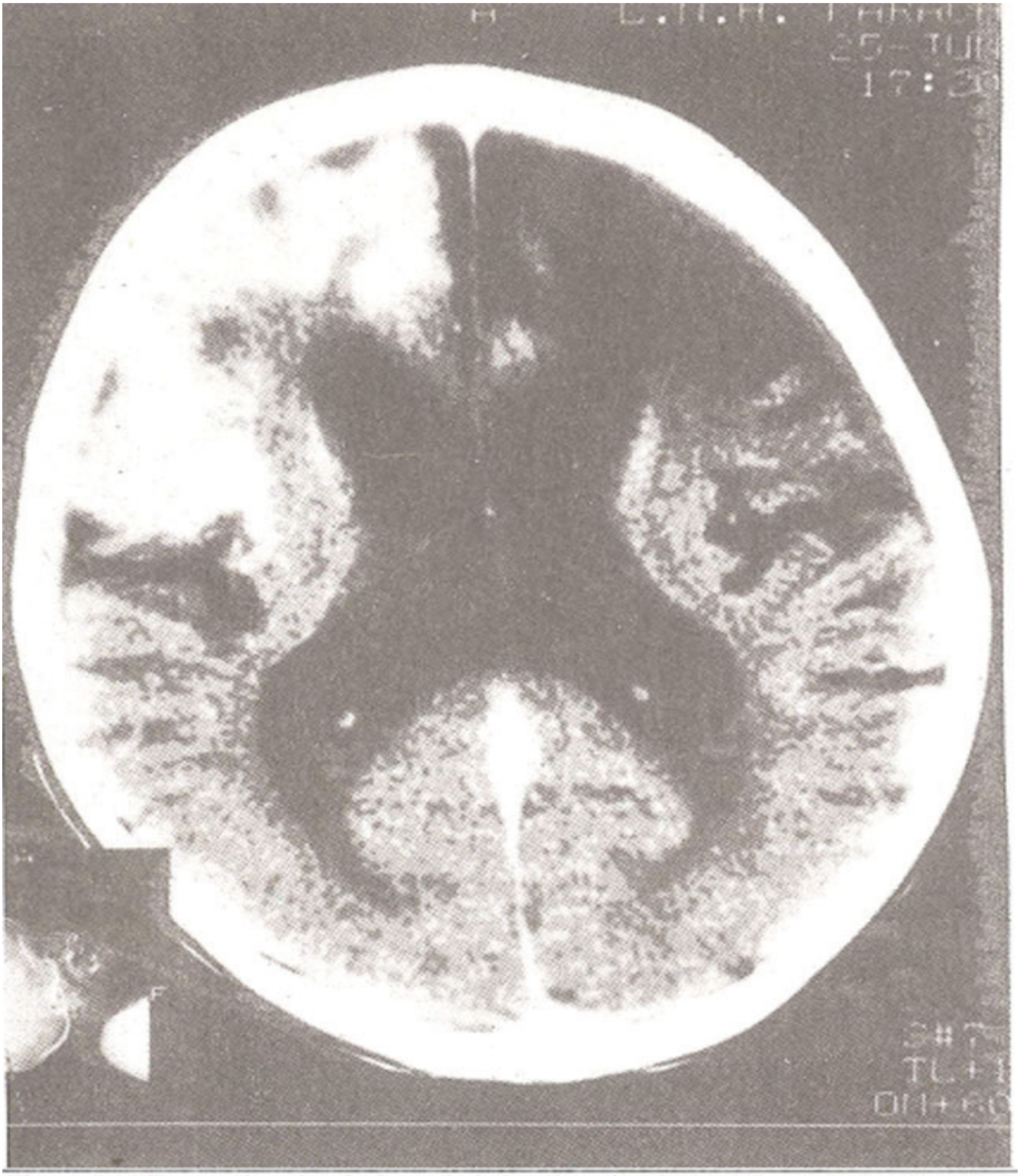

Figure 1. CT scan showing ischaemic infarctions in bilateral fronto parietal regions haemorrhagic on the left side.

Atrophy and ventricular dilatation was moderate in two patients and mild in one patient. Moya Moya vessels were seen on CT scan only in one patient (Figure 2).

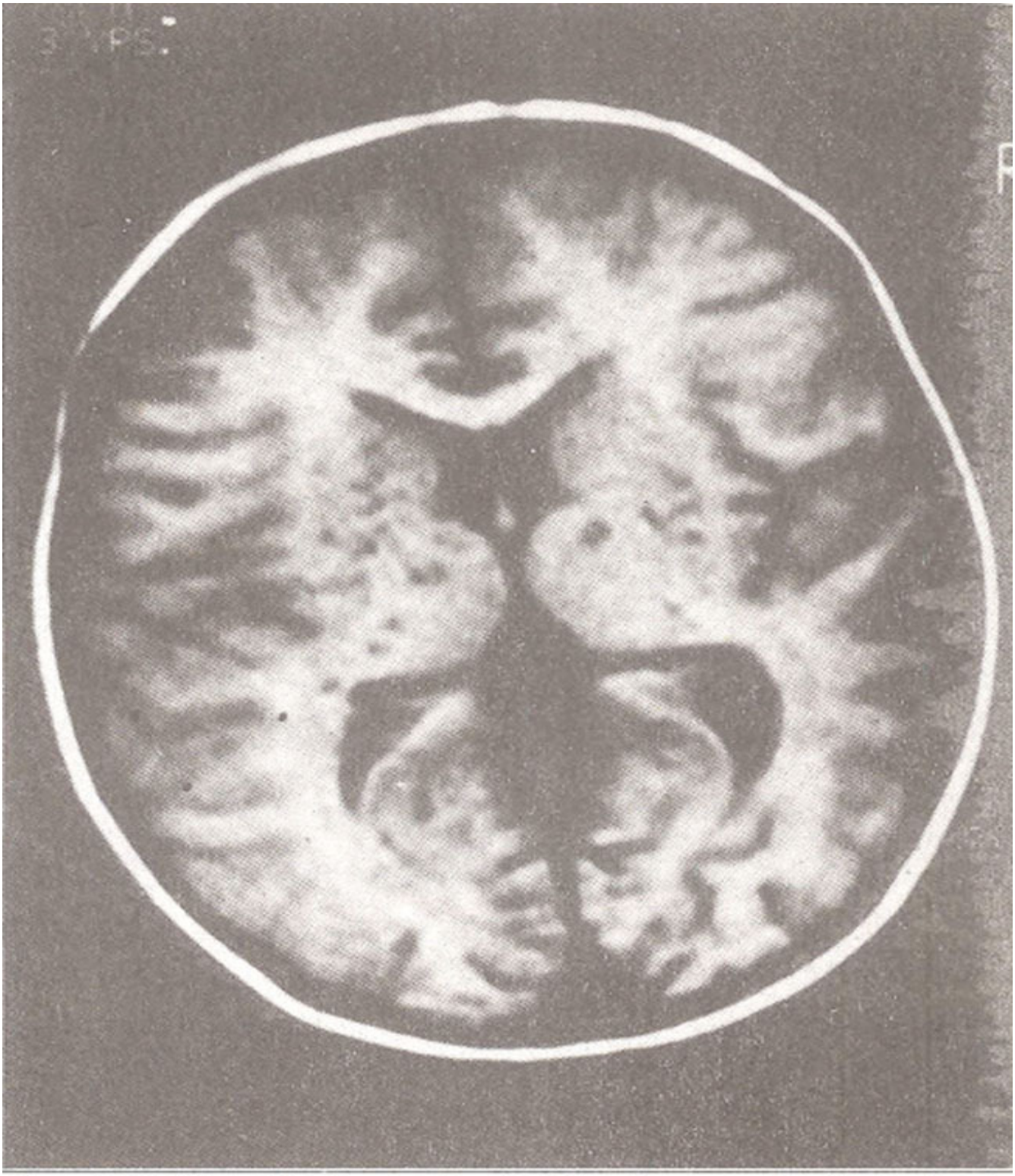

Figure 3. MRI T1 weighted images showing Moya Moya vessels in bilateral basal ganglia region.

Circle of Willis was not visualized in all three patients who had CT scan. MRJ was able to detect small dilated tortuous signal void areas in the basal ganglia region in three patients (Figure 3).

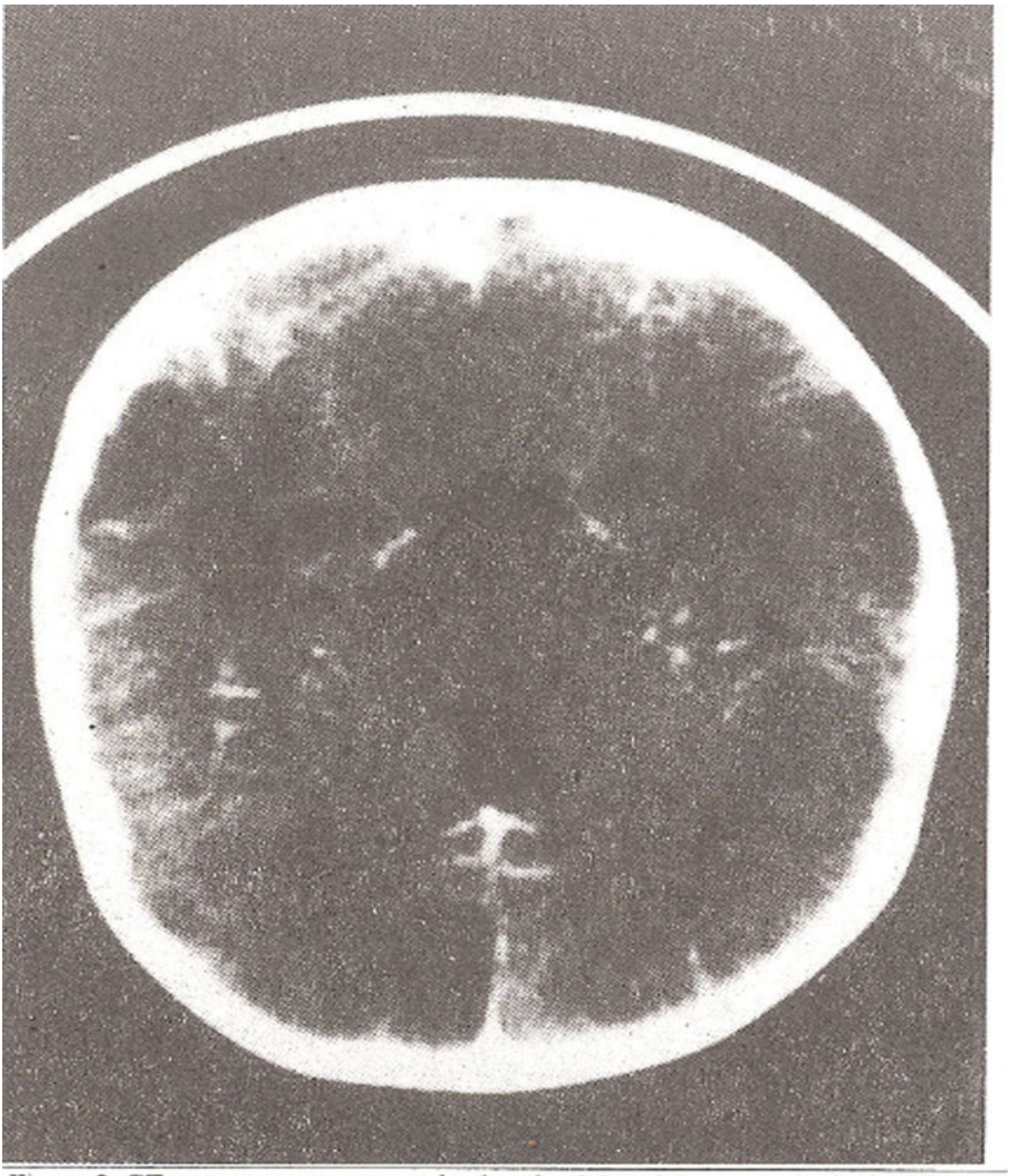

Figure 2. CT scan post-contrast study showing Moya Moya vessels in the bilateral basal ganglia region.

Among four patients studied, three patients showed complete occlusion of supraclinoid portion of internal carotid artery on both sides (Figure 4).

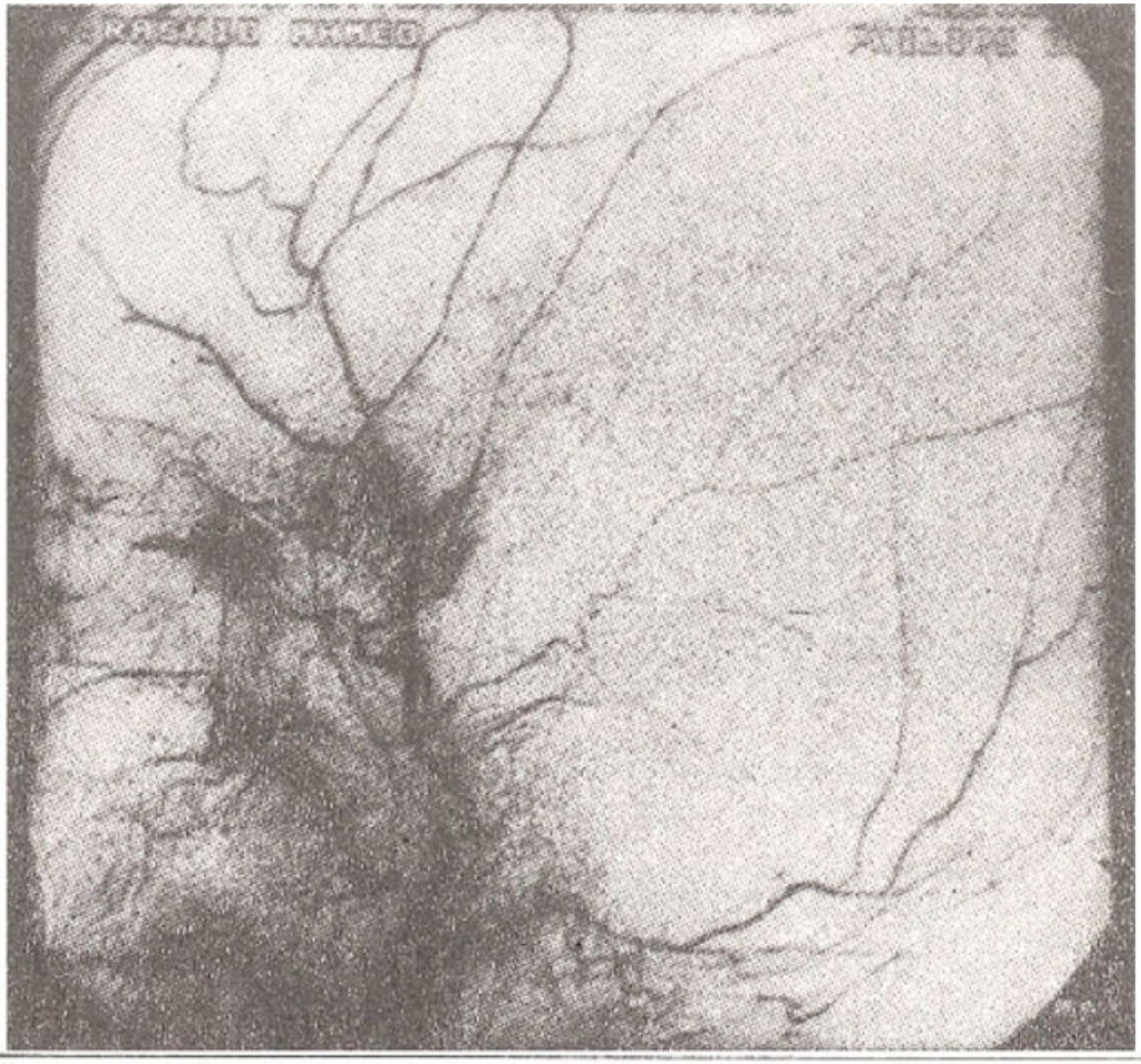

Figure 4. Carotid angiogram lateral view showing complete occlusion of supraclinoid portion of both internal carotid arteries, only the branches of external carotid are visualized.

One patient showed occlusion of supra clinoid portion of internal carotid artery only on one side (left side). On angiography Moya Moya vessels were seen profusely in one case (Figures 5 and 6).

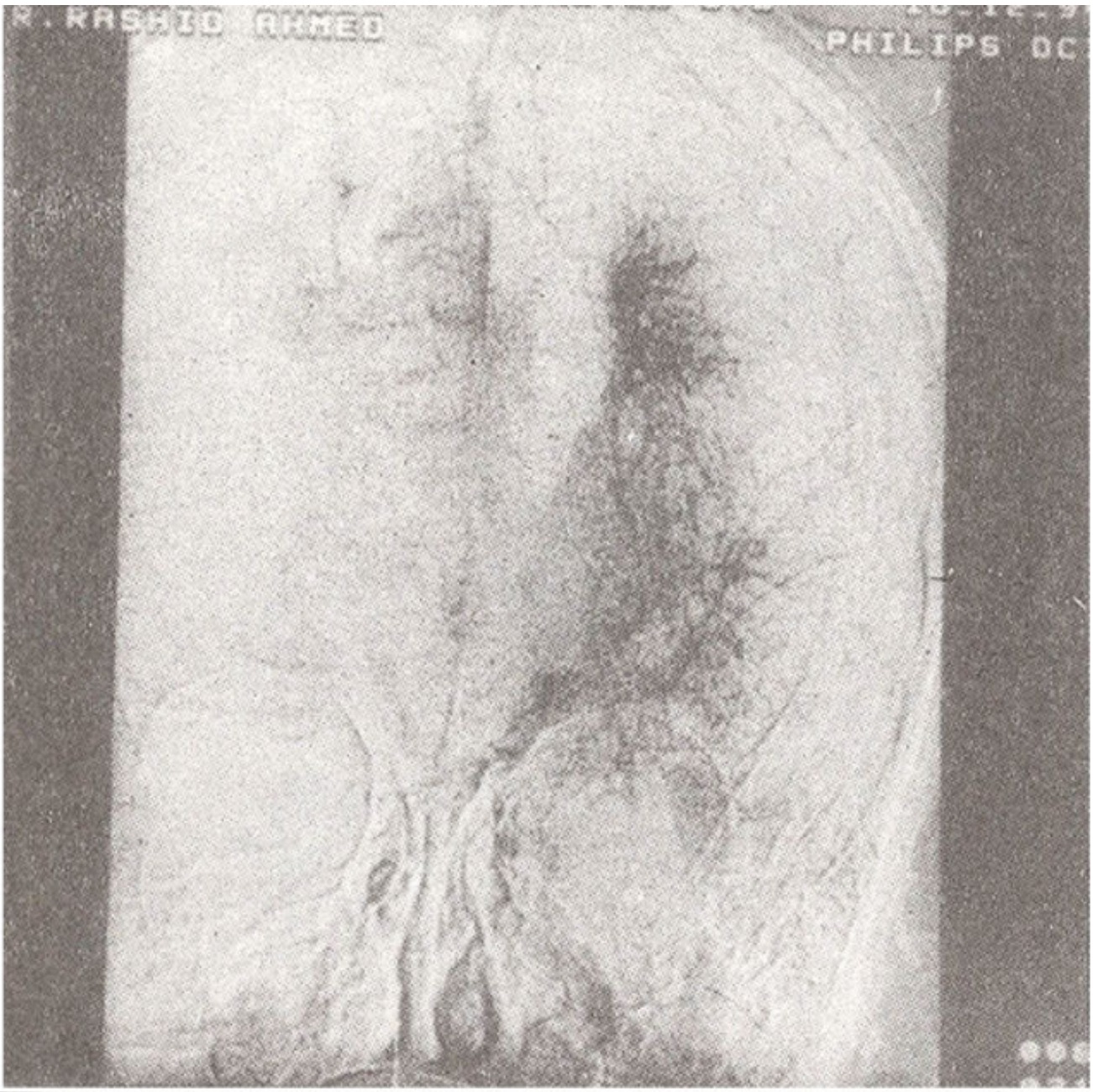

Figure 5. Left carotid angiogram AP view showing Moya Moya vessels on the left side.

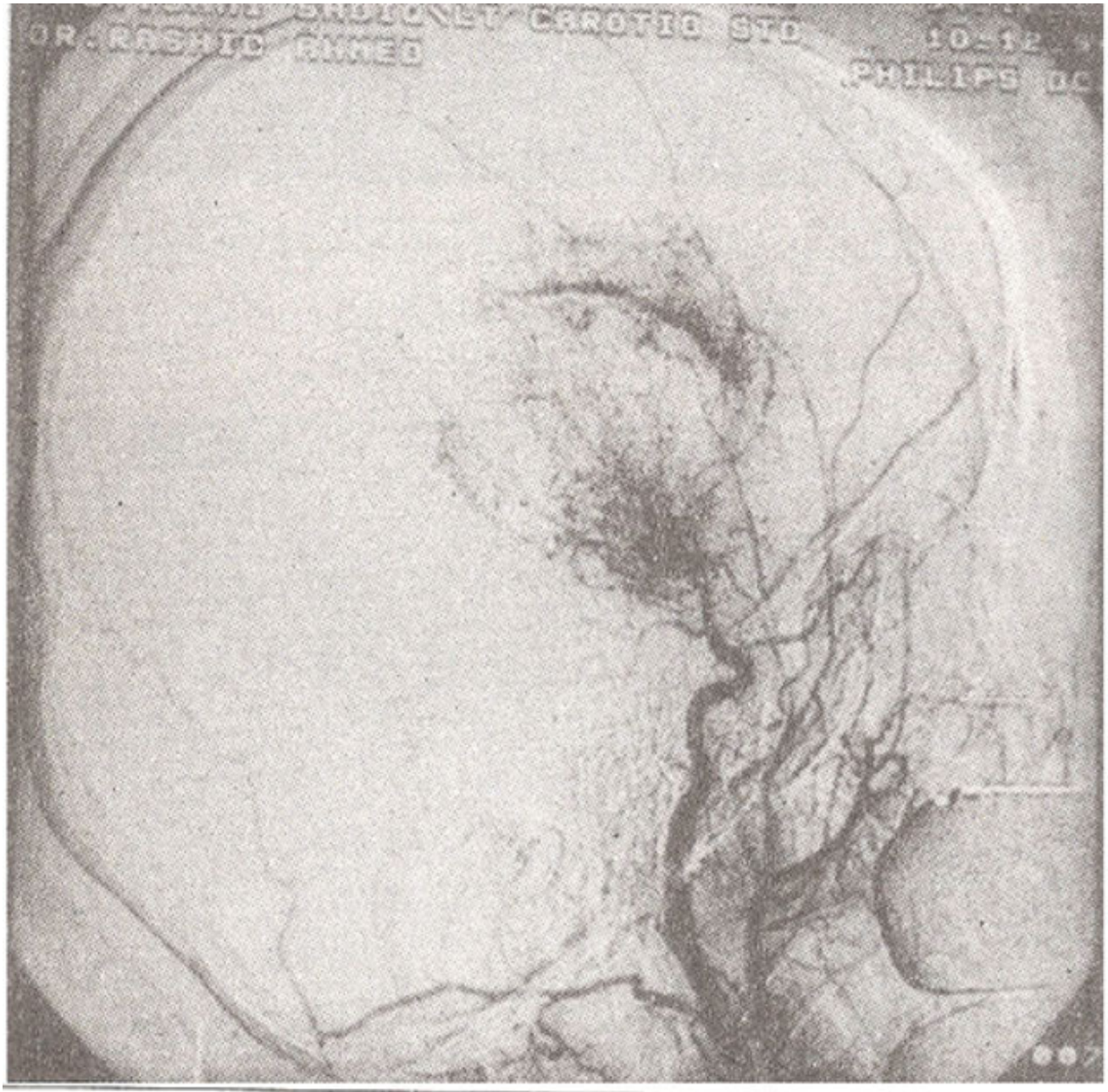

Figure 6. Left carotid angiogram lateral view showing occlusion of supraclinoid portion of internal carotid arteries with Moya Moya vessels.

Moderate visualization was evident in two cases. In one case Moya Moya vessels were only present on one side. Collateral from meningeal and ophthalmic artery was seen in all cases. Filling ofMoyaMoya vessels from posterior circulation was seen in all four cases (Figure 7).

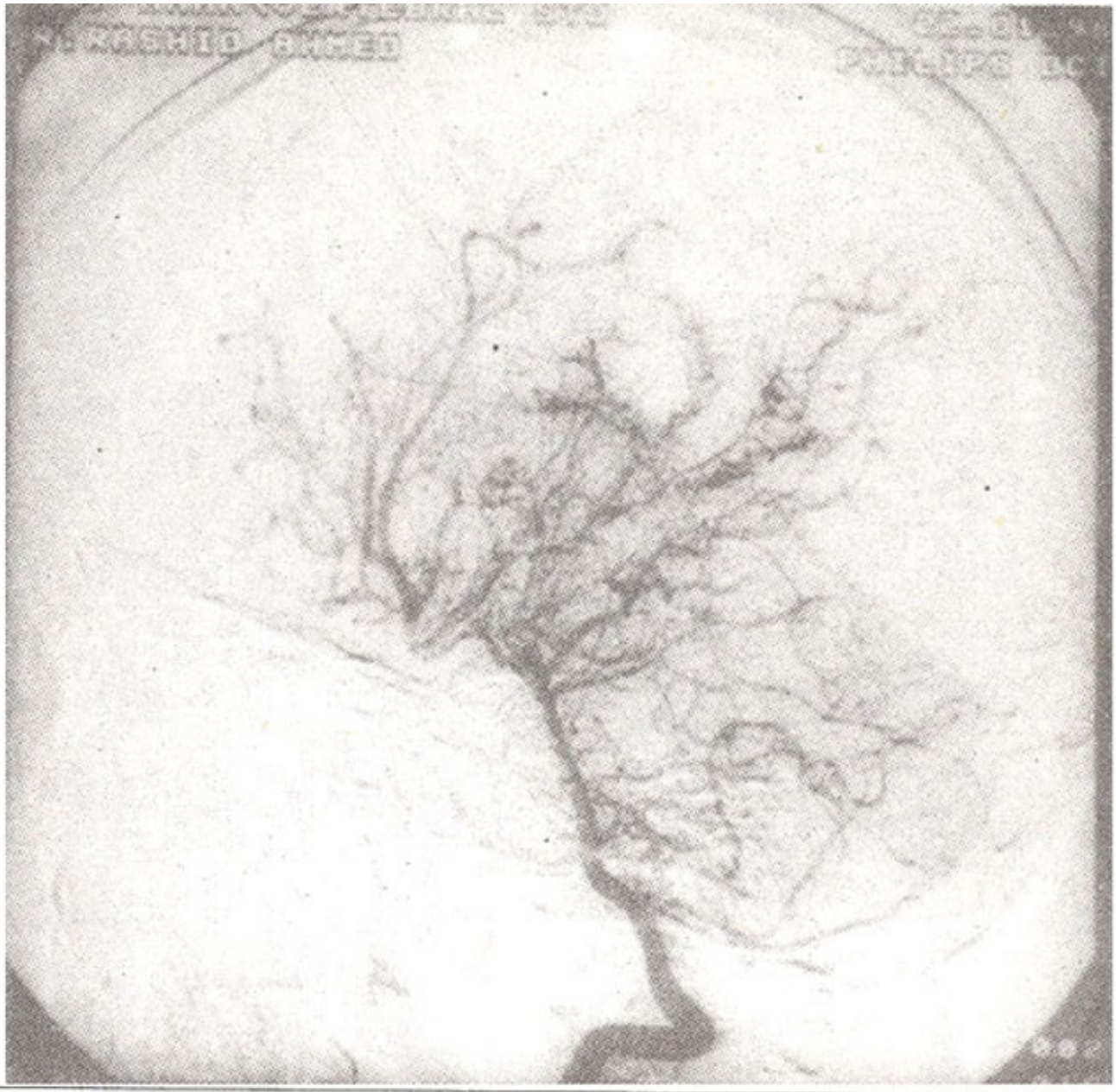

Figure 7. Vertebral angiogram showing filling of branches of middle cerebral arteries from posterior circulation.

Involvement of vertebral artery was seen in three cases, while it was spared in one case. Small aneurysms were seen within the posterior circulation and collateral vessels in two cases (Figure 8).

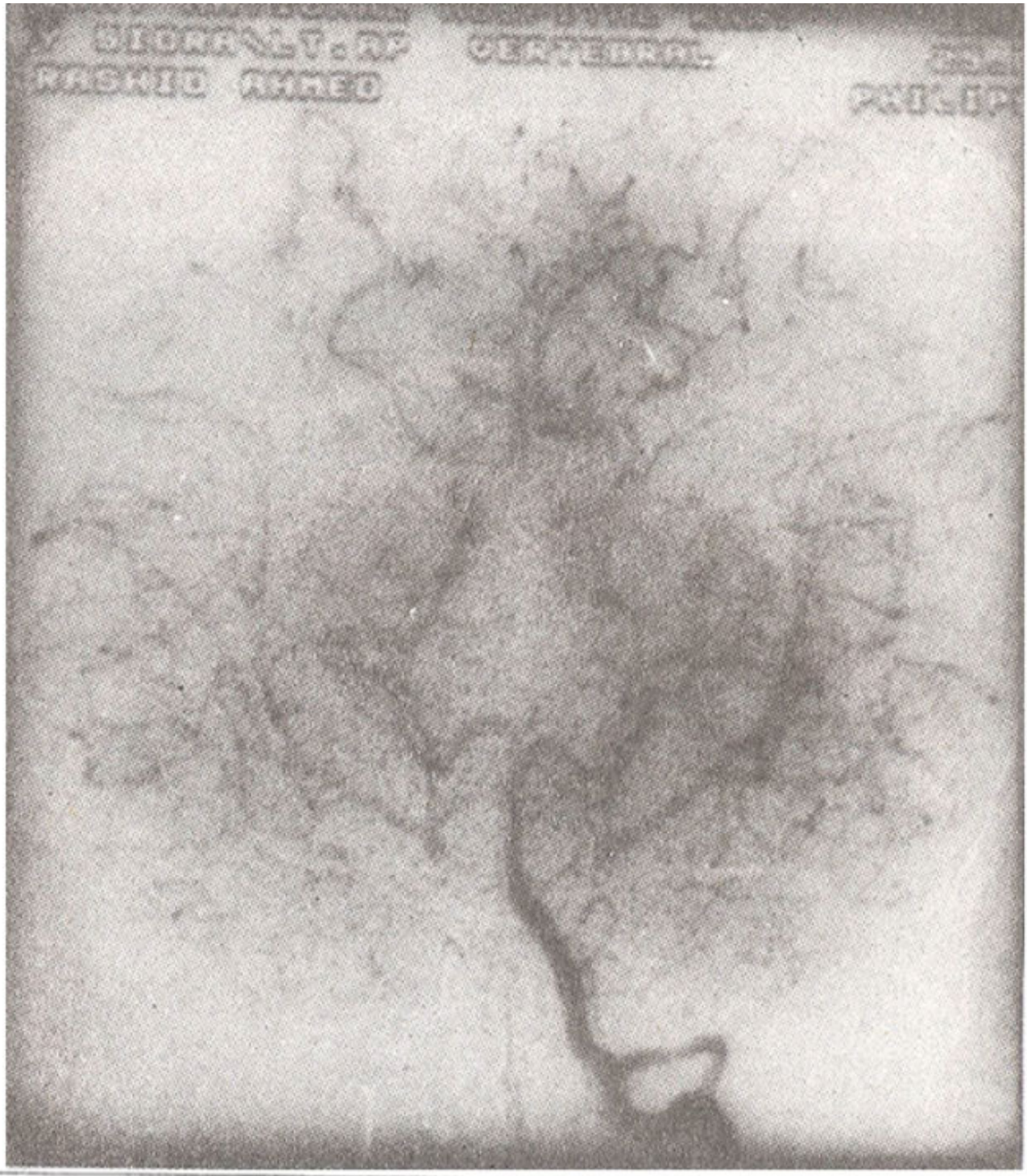

Figure 8. Vertebral angiogram AP view showing small aneurysm in the posterior circulation.

Discussion

Moya Moya disease is characterized by progressive narrowing of the bilateral internal carotid bifurcation, followed by development of extensive parenchymal vessels known as MoyaMoyavessels (MMV'S) from the supra-seller cistern to the basal ganglia[5]. The cause of MoyaMoya disease is still

unknown and it occurs most frequently in young patients. Stroke in children occurs either as a known complication of an already established disease such as cardiopathy, neurofibromatosis type I, homocystinurea, meningitis or sickle cell anaemia or occurs in isolation. One of the cases (case No. 3) was a diagnosed case of sickle cell disease. In all other cases no cause for cerebral occlusion was found. In sickle cell disease stroke occurs in 6-9% of the cases. Both small and large vessel occlusion occurs and multiple intracranial anewysm have been reported. The pathogenesis of sickle cell disease causing stroke is controversial. Recently it is thought that degenerative insult initiated by endothelial cell injury from adhesions of sickled RBC is responsible for occlusion. In children the principle clinical symptoms result from brain ischaemia and consist of hemiplegia, monoplegia, paresthesia involuntary movements, headache and convulsions. In adults most common symptom is intracranial hemorrhage either intracerebral or subarachnoid[6]. The application of CT to the study of this condition has revealed areas of low density in the basal ganglia and cortices with cerebral atrophy, presence of abnonnal vessels and poor visualization of the proximal portions of the anterior and middle cerebral arteries, The demonstration of these findings by CT is important in making a diagnosis[5]. All cases studied by CT in our series showed presence of multiple low density areas in bilateral cerebral hemispheres representing infarctions. MRI was also able to show the Moya Moya vessels in the basal ganglia region in addition to the infarcts and brain atrophy in 3 cases. Cerebral angiography is the only definitive method of diagnosis. The main angiographic findings consist of narrowing or occlusion of the supm clinoid portion of the internal carotid artery with involvement of the anterior and middle cerebral arteries. This is usually. bilateral.

In later stages of the disease the posterior communicating, posterior cerebral and basilar arteries become involved and extensive abnormal vessels (Moya Moya vessels) develop at the base of the brain. The individual vessels of Moya Moya vessels cannot be clearly discerned and its extent is vaguely outlined on routine cerebral angiograms, explaining the name “MoyaMoya” which is Japanese forpuff of smoke or”Hazy”[7]. Collateral circulation develops between external and internal carotid system through branches of middle meningeal and ophthalmic arteries and anterior and middle cerebral arteries. Medullary arteries provide communication between the branches of middle cerebral arteries and MMV (dilated tlialamostriate vessels). MR angiography is a new technology inwhichthevessels canbe imaged with MR signal without the use of contrast agents. Recently there are reports on the role of MR angiography for evaluation of MoyaMoya disease which are vety promising. Diagnosis of Moya Moya disease is most thorough with MRI angiography and MRI imaging.

In patients with Moya Moya disease the CT, MRI and Angiographic findings are very specific and they can readily diagnose and evaluate the extent of the disease. Due to its high resolution and contrast MRI was found to be more informative than CT in detection of abnormal vessels (MMV). Angiography has been the most accurate method for detection of Moya disease but as it is an invasive procedure, it has inherent risks. Due to its non- invasive quality, MRI and MR angiograhy are now preferred methods of investigation for MoyaMoya disease.

References

1. Kudo, T. Spontaneous occlusion of the circle of Willis. Neurology, 1968 ; 18 :485.
2. Suzuki, T. and Takaku, A. Cerebrovascular Moya Moya disease showing abnormal net like vessels in base ofbrain. ARCH. Neurol., 1969,20:288-299.
3. Traveras, J.M. Multiple progressive intracranial arterial occlusion: A syndrome of children andyoung adults. Am.J,Radiol,, 1969;106:235-268.
4. Pecker, LT., Simn. J. and Gyls-Herry, J.F. Nishimoto’s disease: Significance of its angiographic appearance. Neuro-radiology. 1973;5 :223-230.
5. Takahashi, M., Miyauch, T. and Masayoshi Konada. Computed tomography of Moya Moya disease.

Demonstration of occluded arteries and collateralvessels as important diagnostic signs. Radiology, 1980;134:671-676.
6. Ichiro Yamada, I., Yoshihare Mat, S. and Sushima. Moya Moya disease comparison of assessment with MR. Angiography verses conventional angiography. Radiology, 1995;196:211-218.
7. Takahashi, M Magnification angiography in Moya Moya disease. Radiology, 1980;136:379-386.