Ischemic Cerebrovascular Disease in the Young
Two Common Causes in India

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SUMMARY Mandurai City is a composite of many villages with a total population not exceeding one million. Undoubtedly more than 95% of the hospital patients come from the rural areas. Our Government Hospital is the largest and reflects the medical assistance provided for people of average and low income.

In our experience, 15% of cerebrovascular strokes occur in those below 40 years of age. Meningovascular neurosyphilis in men (10-15%) and puerperal cerebral venous thrombosis in women (20%) were the two commonly proved causes. Haemorrhagic stroke was uncommon. Embolic strokes from rheumatic heart disease formed 10%. In over 50%, the cause of stroke was not clear. Among 150 patients with neurosyphilis, mostly men, 25 had cerebral arterial thrombosis with stroke syndrome.

This study reports on 138 patients with cerebral venous thrombosis and 7 patients with arterial thrombosis in puerperium. Mortality was 20% and quality of survival was good. The incidence, clinical picture, and diagnostic investigations are discussed with a review of literature.

CEREBROVASCULAR DISEASE (CVD) occurs less commonly in the younger than 40 years age group. The incidence has been reported from Europe and Japan as varying between 17.4% and 33%. Indian reports estimate the incidence as between 15 and 30% of strokes in all ages. It has been pointed out that the low life expectancy in India (52 years) biases the Indian sample.

Over the past ten years in our teaching hospital (1800 in-patients), 300-350 patients were referred with vascular stroke each year and 15% of them were below the age of 40 years. Neurosyphilis in men and puerperal cerebral venous or arterial thrombosis in women were the commonly proved causes and accounted for 25% of stroke cases in the young. The causative factors in 40 patients in one year (1982) were as follows: puerperal venous thrombosis (CVT), 8 patients, neurosyphilis 5, rheumatic mitral stenosis with atrial fibrillation 5, and other causes in 22 patients including isolated examples of haemorrhagic strokes (due to aneurysm, A.V.M., malignant hypertension, coarctation of aorta, bacterial endocarditis, leukemia) and ischaemic strokes (due to aorto-arteritis, tuberculous meningitis, lupus erythematosus, fibromuscular dysplasia of cervical carotid artery). With increasing use of echocardiography, more cases of cardiac sources of embolism including mitral valve prolapse will be detected. Nevertheless, in over 50% of cases, the cause of stroke in the young is undetected. Higher incidence of haemorrhagic stroke has been reported in India from Kerala, West Bengal, and Chandigarh. However, in most centres, the incidence of haemorrhagic strokes in the young is around 13%. We are aware of the high incidence of cerebral and subarachnoid haemorrhage in Japan and the decreasing incidence of stroke with control of hypertension.

Puerperal Cerebral Venous and Arterial Thrombosis

Incidence

Vascular stroke has been reported as a rare complication by many Western workers, however, it has been reported as a common cause of stroke in puerperal women. Recent publications considered the frequency as from 1 in 2500 to 1 in 10,000 deliveries. The incidence in our centre is around 4.5 per 1000 obstetric admissions compared to the western incidence. Reports from other Indian workers also indicate the higher incidence and that it is 10-12 times more frequent than in other western countries.

In an epidemiological field study reported from Vellore in 1972, 175 stroke victims were women out of a total of 850 patients. Forty-eight percent of patients were below the age of 40 years. Twenty-two patients had puerperal vascular stroke. Mortality was 17%.

A report from Haryana stated that vascular stroke might account for 26% of young strokes and identified obvious venous (angiographic) abnormalities in superior sagittal sinus or cortical veins in 16 patients.

A report from Bombay identified this cause in 6 out of 93 young stroke victims. Work in Delhi reported 3 necropsies: superior sagittal sinus thrombosis alone in one, associated haemorrhagic infarct in MCA territory in one, and MCA infarct alone in one patient.

Work in Chandigarh reported the incidence of young strokes as 16%. The autopsy findings in 8 patients out of a total of 24 vascular stroke cases in two years, were reported. There was occlusion of superior longitudinal sinus alone or with one transverse sinus in 6 patients, and occlusion of vein of Galen and infarction in 2 patients.
Materials and Methods

In-patient statistics for the past ten years at our hospital recorded 8000 deliveries every year. Among these about 400 had toxemia of pregnancy. After excluding gestational epilepsy and toxemia of pregnancy, all patients with impairment of consciousness, seizures, or focal neurological deficit were transferred to the neurology unit. Postpartum eclampsia was excluded by the occurrence of illness beyond 48 hours after delivery, absence of hypertension or albuminuria, and presence of focal seizures and signs. Electroencephalography, lumbar puncture, or plasma fibrinogen levels were not helpful in this differential diagnosis. Blood counts, routine haematological and biochemical tests, EEG, lumbar puncture, and angiography were done to exclude metabolic causes, infections, and other space occupying lesions. The diagnosis was confirmed by high plasma fibrinogen, positive findings in angiography, sinogram, and in a few cases by explorative craniotomy or autopsy.

A yearly average of 50 patients are referred to us with symptoms of vascular stroke. Of these, about 30 present with stupor or coma with seizures and are transferred to the neurology unit for further study.

Results

Puerperal vascular stroke accounted for 15–20% of strokes in the young in our centre and we have studied in detail over 145 such patients. Most of the reports about this illness are only based on hospital study and include only the more ill patients. The actual incidence is bound to be more frequent if all the municipal hospitals and home deliveries are considered.

The clinical picture is typical. The illness occurs in the first three weeks after a normal child birth at full term, often in a multiparous woman. Only 22 were primiparous. The general ratio of multi to primi is 2:1 in our hospital records. None of these multiparous women had a similar illness in earlier pregnancy. Only 7 of our patients were readmitted with recurrence of vascular stroke in subsequent pregnancies.

The typical patient has focal or multifocal or generalized seizures (80%), stupor or coma (50%), fleeting or regressing neurological deficits like limb weakness (60%), or cranial nerve palsies and signs of raised intracranial tension (18%) (evidenced by papilloedema or increased CSF pressure or midline shift in angiography), and deep leg vein thrombosis in 12%. Cortical deficits like agnosia, apraxia, aphasia, or visual field defects are rare. Pelvic sepsis was seen only in 5 patients.

In our study, plasma fibrinogen was raised well over control values in 104 and normal in 16 patients. Other findings in routine haemogram were normal. The CSF was abnormal in only a few patients. It was haemorrhagic in 3/56 and showed elevated proteins in 12/56. EEG done in 50 patients showed focal or lateralisating abnormalities only in 10. Carotid angiogram was done in 72 patients. The arteriogram appeared normal in 40, showed obvious venous abnormalities in 14 (like occlusion of major sinus or distortion and displacement of cortical veins around avascular areas), and cerebral infarction with pseudo-tumour pictured in 11 patients. Sinogram showed blocked sagittal sinus in 3 patients and normal in one. Craniotomy revealed haemorrhagic infarct in 2 patients while two more were confirmed on autopsy.

Apart from routine treatment of cerebral infarction, we find Heparin very useful in the more ill and stuporous cases without haemorrhagic CSF. The mortality has been reduced to 20% from the earlier reported mortality of 28 to 33%. Fatal pulmonary embolism occurred in a few patients after the first week. Ninety percent of those who survived, recovered fully and resumed their domestic or agricultural work.

Cerebral Arterial Thrombosis in Pregnancy and Puerperium

Western reports attributed the symptoms to arterial ischaemia in many of their cases. Arteriograms done in 27 patients showed arterial occlusion in 55%. It was pointed out that absence of demonstrable occlusion may be due to clot lysis.

In India, arterial occlusion has been reported variously. In our centre, MCA occlusion was seen in 5 and internal carotid artery occlusion in 2 patients only, though carotid angiogram was done in 72 women. There was no evidence to suggest vasculitis, embolism, L.E., or blood dyscrasia as a cause of stroke in our patients, and serology (VDRL) was negative. Echocardiogram assessment was not available in most of these cases to exclude mitral valve lesions apart from clinical signs. Puerperal cardiomyopathy, amniotic fluid embolics, or choriocarcinoma could be rare causes of embolic strokes and were not detected in any of our patients.

In contrast to CVT, women with arterial occlusion were often conscious, with dense persisting focal motor deficit, and rarely had seizures or papilloedema.

Neurosyphilis

Before the penicillin era, the incidence of neurosyphilis was reported as 29%,16 but this illness has rapidly declined in recent times.

Meningovascular type is the commonest type in India, whereas more cases of parenchymatous neurosyphilis are reported from the West. The upgraded Institute of Venereology at Madras is the National reference centre for serology in syphilis, and our laboratory results are counterchecked and approved periodically. We have recorded the highest incidence of neurosyphilis in our country. Consistently over the past ten years it is around 4% whereas it is around 1–1.5% in other teaching institutions of our country. At Madras, the total annual average number of cases with positive serology is 2500 and neurosyphilis was confirmed in 30 patients. Corresponding figures for our centre is 30 out of 750. Meningovascular syphilis represents 60% of cases of neurosyphilis, primary optic atrophy 20%, and rest includes GPI and Tabes.

Strokes due to meningovascular syphilis account for 10–15% of young strokes. Patients are usually con-
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conscious because of the small size and pale infarcts, but may have seizures. MCA thrombosis is commonly demonstrated and vertebrobasilar ischemic strokes are equally common. While angiogram demonstrates the vascular occlusion, aetiology is established by CSF biochemical changes, positive serology (VDRL) in blood and CSF, and/or specific tests in blood and CSF (like FTA, FTA ABS in most of the cases even when VDRL is negative and more recently immunoglobulin study in the CSF). Recovery is usual, and quality of survival is good in 50% with specific therapy.

We have studied 150 patients with neurosyphilis. Twenty-five men had presented with stroke syndrome as a part of meningo-vascular syphilis in the age group 20–40, all with positive blood and CSF serology and (in many cases) positive CSF and FTA tests. None of these patients had any other obvious cause for stroke including cardiac lesions.

Quality of Survival

We followed up 46 consecutive young stroke victims (age below 40) with occlusive (non-haemorrhagic stroke) in the carotid territory. At the end of 2 years, 26 were fully independent and working, 12 partially disabled and ambulant, and 8 dependent. This group included all causes, but most of the patients who resumed their work belonged to the group of vascular stroke or neurosyphilis.

References

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Stroke. 1984;15:733-735
doi: 10.1161/01.STR.15.4.733
Stroke is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0039-2499. Online ISSN: 1524-4628

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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