A rare presentation of stroke in young age: moyamoya disease

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A 37-year-old hypertensive housewife presented with a sudden onset of left-sided hemiplegia, hemianaesthesia, dysarthria and urinary incontinence. The condition was preceded by recurrent attacks of motor neurological deficits over a 3-year duration. She reported a history of a fall from a height at the age of 10, which was followed by a hearing deficit and a history of two caesarean sections after eclampsia. The blood pressure was 170/100 mmHg. Laboratory investigations revealed hyperglycaemia (fasting glucose 306 mg/dl) and normal kidney function tests. The computed tomography scans revealed old multiple bilateral cerebral infarcts with recent intracranial haemorrhage in the right parietal region. The inflammatory markers (ESR and CRP) and immune profile (ANA, anti-ds DNA and ANCA) were found to be normal. Cerebral angiography revealed a complete occlusion of the intracranial parts of both internal carotid arteries at their supraclinoid segments along with the proximal parts of the anterior cerebral artery and middle cerebral artery, with collaterals from the posterior circulation. Consequently, the diagnosis of moyamoya disease with the collaterals was confirmed. Antihypertensive medications and insulin were administered. Cerebral dehydration measures were undertaken with partial improvement. A superficial temporal artery–middle cerebral artery bypass operation was performed with some postoperative improvement. One month later, she suffered a new stroke with severe impairment of the level of consciousness; the computed tomography scans revealed a large recent cerebral infarct, her condition deteriorated rapidly and she died shortly thereafter.

Keywords:
angiography, CT scan, moyamoya, stroke

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Case report

A 37-year-old housewife known to be hypertensive for 8 years on irregular treatment and not known to be diabetic presented to the casualty department with a sudden onset of left-sided paralysis and sensory loss associated with dysarthria and urinary incontinence but no history of trauma, fever or manifestations of increased intracranial tension. The condition was preceded by recurrent attacks of motor neurological deficits over a 3-year duration with minimal residue. She reported a history of a fall from a height at the age of 10, which was followed by a hearing deficit and a history of two caesarean sections after eclampsia. Examination revealed a noncooperative, depressed and fully conscious patient. Blood pressure measures 170/100 mmHg; pulse 80/min, regular, of average volume and carotids are equally felt bilaterally. The heart and chest were normal. The abdomen had a scar of the caesarean section. Neurological examination revealed left-sided hypotonia and hyporeflexia with Babinski's sign, slurred speech and mild weakness of the right upper limb with grade 3–4 power. Laboratory investigations revealed an accidentally discovered hyperglycaemia (fasting glucose 306 mg/dl) and normal kidney function tests. The computed tomography (CT) scans revealed old multiple bilateral cerebral infarcts with recent intracranial haemorrhage in the right parietal region (Fig. 1). The aetiology of the neurological deficits was suggested to be different from atherosclerosis on the basis of age, recurrence of stroke and concomitant findings of cerebral infarcts and haemorrhage. The heart and carotids were proven not to be responsible, and disseminated sclerosis was ruled out as it does not present with cerebral haemorrhage; consequently, we were convinced that this lady had a primary disease of the cerebral vessels such as vasculitis with aneurysm, arteriovenous malformation or rare vascular anomaly that can present in adults by both infarction and stroke and are characteristic of moyamoya. The inflammatory markers (ESR and CRP) and immune profile (ANA, anti-ds DNA and ANCA) were found to be normal. Cerebral angiography was performed and revealed a complete occlusion of the intracranial parts of both internal carotid arteries at their supraclinoid segments along with the proximal parts of the anterior cerebral artery (ACA) and middle cerebral artery (MCA) (Fig. 2). Multiple collaterals were seen arising from the posterior circulation through multiple perforators and cortical branches, with refilling of the distal part of ACA and MCA (Fig. 3); consequently, the diagnosis of moyamoya disease with collaterals was confirmed. A small saccular aneurysm (2.8×2.2 mm) of the supraclinoid segment of the right internal carotid artery was also detected.

Antihypertensive medications were administered to control the blood pressure and insulin to control blood glucose;

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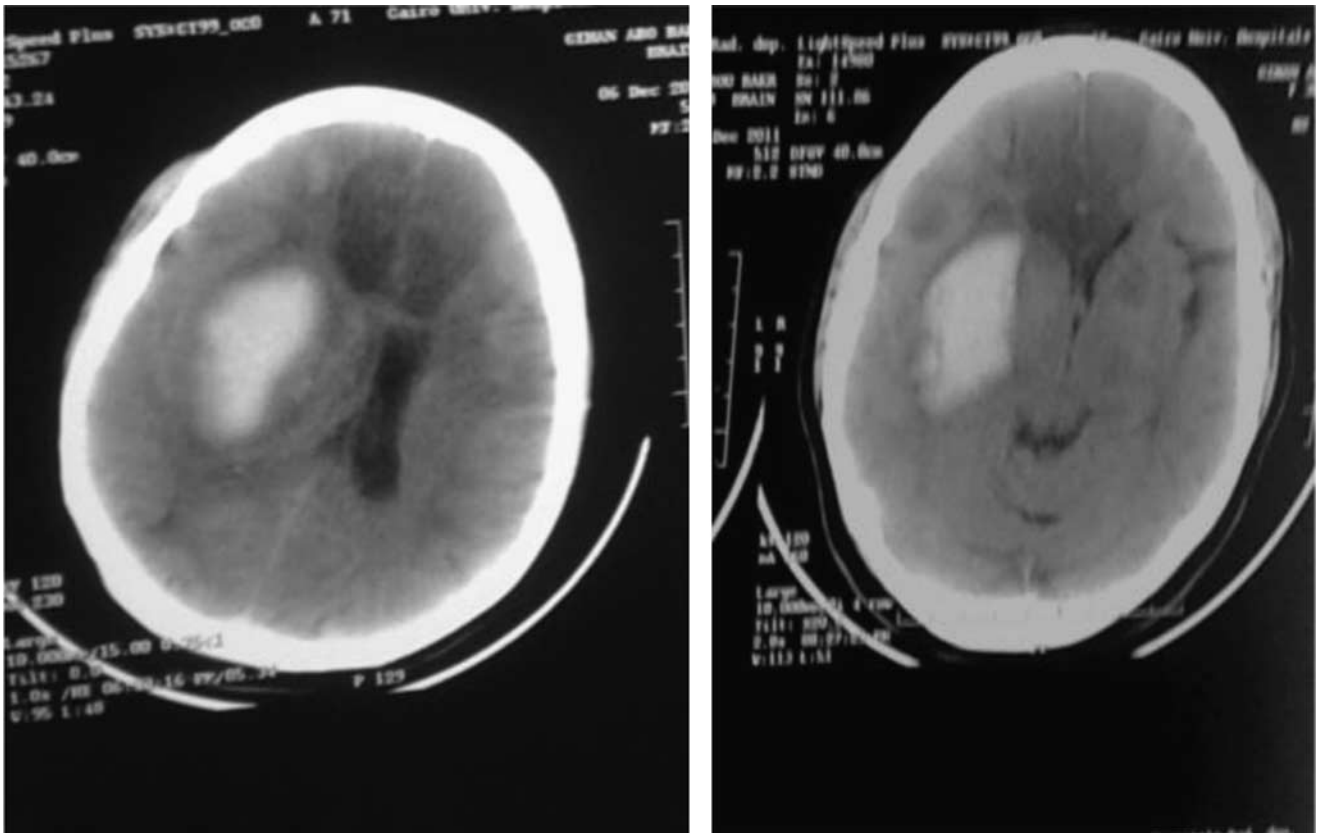
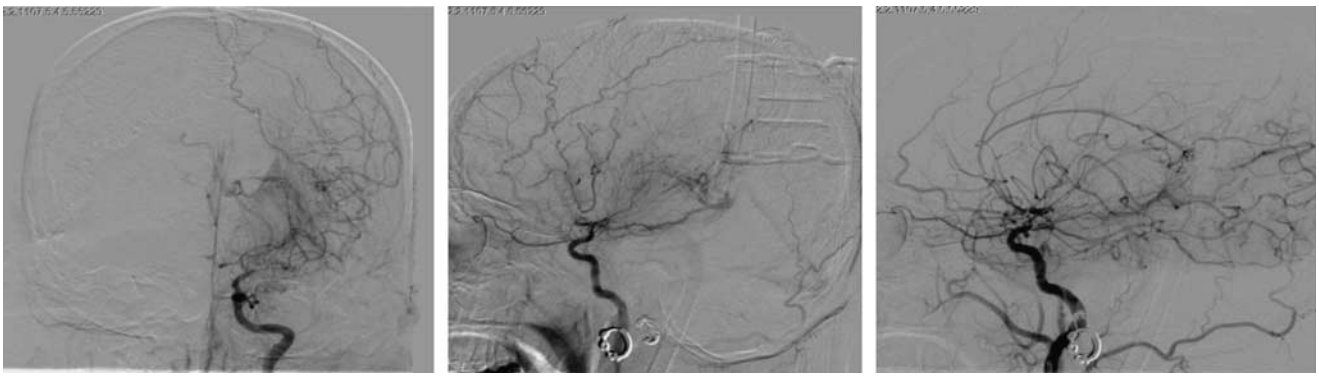
Figure 1



Multiple cerebral infarcts and haemorrhage.

Figure 2



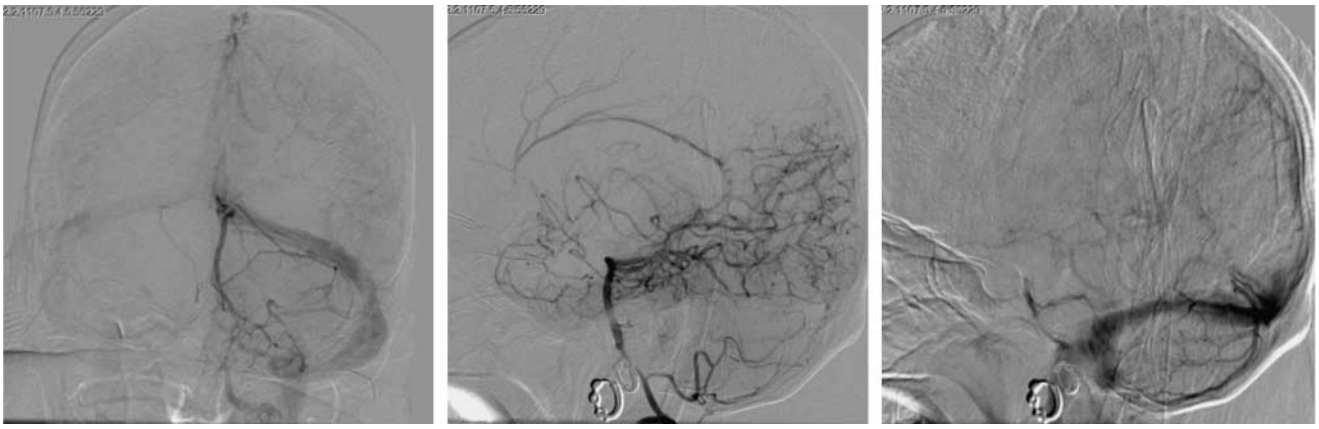
Complete internal carotid artery occlusion at the supracleinoid segment along with the proximal segments of the anterior cerebral artery and middle cerebral artery.

however, her conscious level deteriorated rapidly, aspiration pneumonitis developed with high fever, and the CT scans revealed a frontal lobe infarction. She was admitted to the ICU, administered antibiotics and antipyretics and cerebral dehydration measures were undertaken with partial improvement. The decision to operate was taken after neurosurgical consultation. The procedure chosen was superficial temporal artery–MCA bypass with some post-operative improvement. One month later, she was readmitted as a result of a new stroke with severe impairment of the level of consciousness; the CT scans revealed a large recent cerebral infarct. She was admitted to the ICU, but unfortunately her condition deteriorated rapidly and she died shortly thereafter.

Conclusion

This was a case of definite moyamoya disease that is quite rare, with a characteristic bilateral occlusion of terminations

Figure 3



Smoke puff-like appearance of the collaterals from the posterior cerebral circulation.

of the internal carotid arteries and proximal segments of the ACA and MCA, with refilling of the ACA and MCA branches through the collaterals with the posterior cerebral artery. The disease was first reported in Japan and then to a lower extent in East Asia and North America, followed by other countries. The name moyamoya means puff of smoke in Japanese, as the collaterals in this disease are weak, small and scattered.

The aetiology is not yet known but genetic and acquired factors have been reported. The acquired factors are brain irradiation, Down syndrome, neurofibromatosis and sickle disease, whereas chromosomal abnormalities, especially chromosome number 17, were detected in Japan with variable genetic penetrance [1]. The pathological changes observed in the arteries affected are smooth muscle atrophy and overgrowth of the inner vascular wall with subsequent stenosis or occlusion. Increased levels of fibroblast growth factor, vascular endothelial growth factor and adhesion molecules were recorded [2]. The presentation ranges from asymptomatic to transient ischaemic attacks, stroke, headache, seizures, cerebral haemorrhage and visual and cognitive deficits [3]. Diagnosis is based on CT scans of the brain and MRIs that can detect acute and chronic brain infracts and cerebral haemorrhage. However, cerebral angiography is still the gold standard for a definitive diagnosis [4]. The treatment is bypass surgery, usually superficial temporal artery–MCA bypass, with a reduction in the incidence of infraction in the treated side by 89% compared with the nonoperated side and a 5-year infraction free period in 94% of patients compared with 36% patients in the untreated group [5]. In general, 4% of patients are reported to develop postoperative strokes; unfortunately, our patient belonged to this category as she died 30 days after the operation following the development of a new large cerebral infarct [3].

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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