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Case of the month

Smoke signals

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This male child had been well until the age of 8 years when he developed episodes of weakness and jerking of his right arm. He was felt to have partial seizures and treated with anticonvulsants. 4 years later he presented to hospital with a 5 day history of headaches and confusion associated with dysphasia. These symptoms gradually improved, but he was re-admitted the following month with left hemiplegia and a left homonymous hemianopia. CT (Figure 1) and cerebral arteriography (Figure 2) were performed. An anteroposterior view of the right common carotid injection is illustrated.

What is the diagnosis?

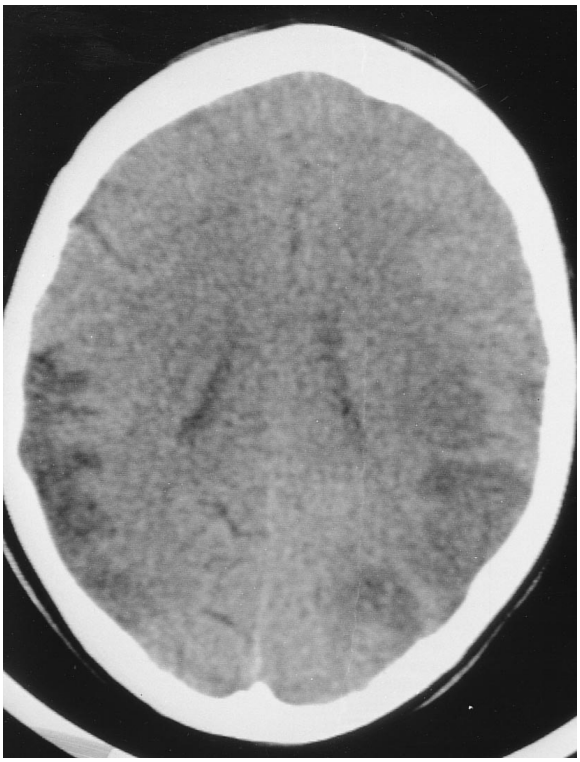

Figure 1. CT scan.

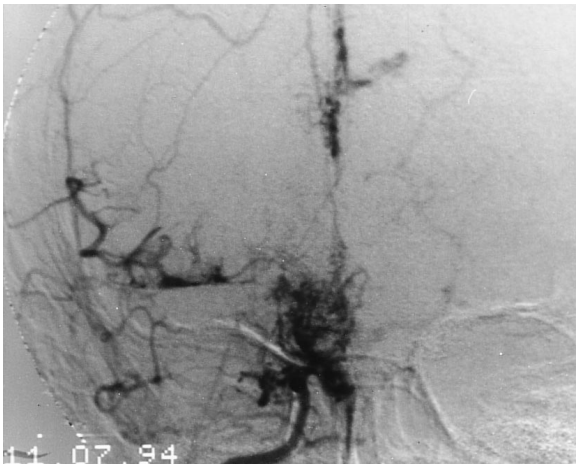

Figure 2. Cerebral arteriogram.

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The CT scan shows widening of the cortical sulci in the left parietal region, consistent with focal atrophy due to an old cortical infarction. In the right parietal region an extensive zone of ill defined low density extending out to the cerebral cortex, associated with a slight mass effect, is consistent with a more recent evolving infarct.

The right carotid arteriogram shows occlusion of the internal carotid artery in the region of the carotid syphon. The artery terminates in a leash of tiny collaterals, probably meningohypophyseal vessels. The distal portions of the anterior and middle cerebral arteries are seen to fill via these collaterals.

The diagnosis is moyamoya disease.

Discussion

Moyamoya disease was first described in Japan in 1957 by Takeuchi and Shimizu [1]. It is characterized by bilateral progressive occlusion of the intracerebral vessels, initially affecting the terminal parts of the internal carotid arteries and progressing to involve the anterior, middle and, in some cases, the posterior cerebral arteries. Simultaneously, a dense network of abnormal collateral vessels is formed at the base of the brain, angiographically resembling a puff of smoke, which is the meaning of the Japanese word "moyamoya".

The cause of moyamoya disease is unknown. It affects all races and has a female preponderance. There is an increased incidence in various unrelated conditions including Down's syndrome, neurofibromatosis, polycystic kidney disease, and following radiotherapy. Postmortem and angiographic studies sometimes show stenoses in extracranial arteries including the renal, pulmonary and external carotid arteries, suggesting that systemic factors may contribute to the pathogenesis. Histology of the affected arteries shows intimal thickening and medial thinning. Increased production of basic fibroblast growth factor (bFGF), a potent mitogen of vascular endothelial and smooth muscle cells, could be responsible. Raised levels of bFGF have been found in the superficial temporal arteries of patients with the disease [2].

The clinical presentation differs between adults and children. Children develop ischaemic events, including TIAs and strokes, as in this case. Seizures are common. The prognosis in this age group is better than in adults, who usually present with cerebral haemorrage from the fragile, abnormal vessels. Additional cerebrovascular risk factors such as diabetes, hypertension and the oral contraceptive pill are likely to modify the natural history in adults.

The diagnosis of moyamoya disease is based primarily on the angiographic findings. Suzuki and Takaku noted six stages [3].

Stage 1. Suprasellar stenosis of carotid arteries.
Stage 2. Collateral "moyamoya" vessels develop at the base of the brain.
Stage 3. Major trunks of anterior circulation become severely stenosed or occluded.
Stage 4. All components of circle of Willis occluded.
Stage 5. "Moyamoya" vessels diminish and collaterals develop from the extracranial circulation.
Stage 6. "Moyamoya" vessels and cerebral arteries disappear. Cerebral hemispheres receive blood entirely from extracranial–intracranial anastamoses.

This case is considered to correspond to stage 3.

Laborde et al noted a good correlation between angiography and transcranial Doppler sonography and recommend this method in diagnosis, surgical planning and post-surgical follow-up [4].

The commonest CT finding is infarction, as in this case. Cerebral atrophy, haemorrhage or an abnormal vascular network may be seen in the region of the basal ganglia on CT and MRI. MR angiography will demonstrate occluded carotid and cerebral arteries as well as the moyamoya vessels [5].

Surgical treatment is aimed at revascularization of the ischaemic brain through extracranial–intracranial anastamosis.

Although moyamoya is a rare disease, it is increasingly recognized outside Japan and should be considered in all cases of childhood cerebral ischaemia and infarction.

References

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