Moyamoya desease, a woolly question…

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BACKGROUND

Moyamoya disease (MMD) is a chronic, occlusive cerebrovascular disease characterized by progressive stenosis at the terminal portion of the internal carotid artery (ICA) and an abnormal vascular network at the base of the brain. Although well known for its ischemic complications, these basal arterial collaterals are prone to rupture making MMD a possible and overlooked cause of deep intracerebral haemorrhage (ICH) in young adults.

CASE REPORT

A 46-years-old Caucasian female was admitted to the Emergency Department for acute onset of severe headache associated with retching and vomiting, followed by seizure. The patient was affected by Neurofibromatosis type 1 (NF1), dyslipidaemia and epilepsy. She had been completely seizure free for twenty years after the beginning of Phenobarbital therapy. Her sister was affected by NF1 too. None of her relatives had a history of cerebrovascular diseases or epilepsy. Admission brain CT scan showed ICH of the left fronto-parietal lobe associated with intraventricular haemorrhage and midline shift. Brain CT angiography (CTA) revealed hypoplasia and occlusion of the right internal carotid artery (ICA), left ICA stenosis with narrowing of the left anterior and middle cerebral arteries and compensatory dilated collateral circles of the deep cerebral perforating arteries raising the suspicion of MMD. The patient deteriorated quickly (GCS 1 + 3 + 2) and surgical evacuation of the hematoma was performed. Digital subtraction cerebral angiography confirmed MMD showing severe bilateral stenosis of the distal ICA and the typical collateral hazy network of cerebral perforating arteries. The patient was discharged with right sided hemiparesis (GCS 4 + 4 + 1).

DISCUSSION

MMD should be considered in young patients with acute neurological symptoms referable to ischemic or haemorrhagic stroke and epilepsy. The aetiology still remains largely unknown. The typical vessel occlusion results from hyperplasia of smooth-muscle cells and luminal thrombosis without atherosclerotic or inflammatory signs. Moreover, the fibrocellular intimal thickening may also affect extracranial arteries, including cervical carotid, renal, pulmonary, and coronary vessels. MMD is strongly associated with radiotherapy to head or neck, Down's syndrome, thyroid disease and, as in our patient, with NF1. Disease progression may be slow, relapsing, progressive or fulminant. Young patients presenting with ICH, especially in the absence of hypertension, probably have an underlying secondary cause of ICH, such as vascular abnormalities. Vascular imaging is therefore mandatory to detect the typical changes of MMD in this particular clinical setting.

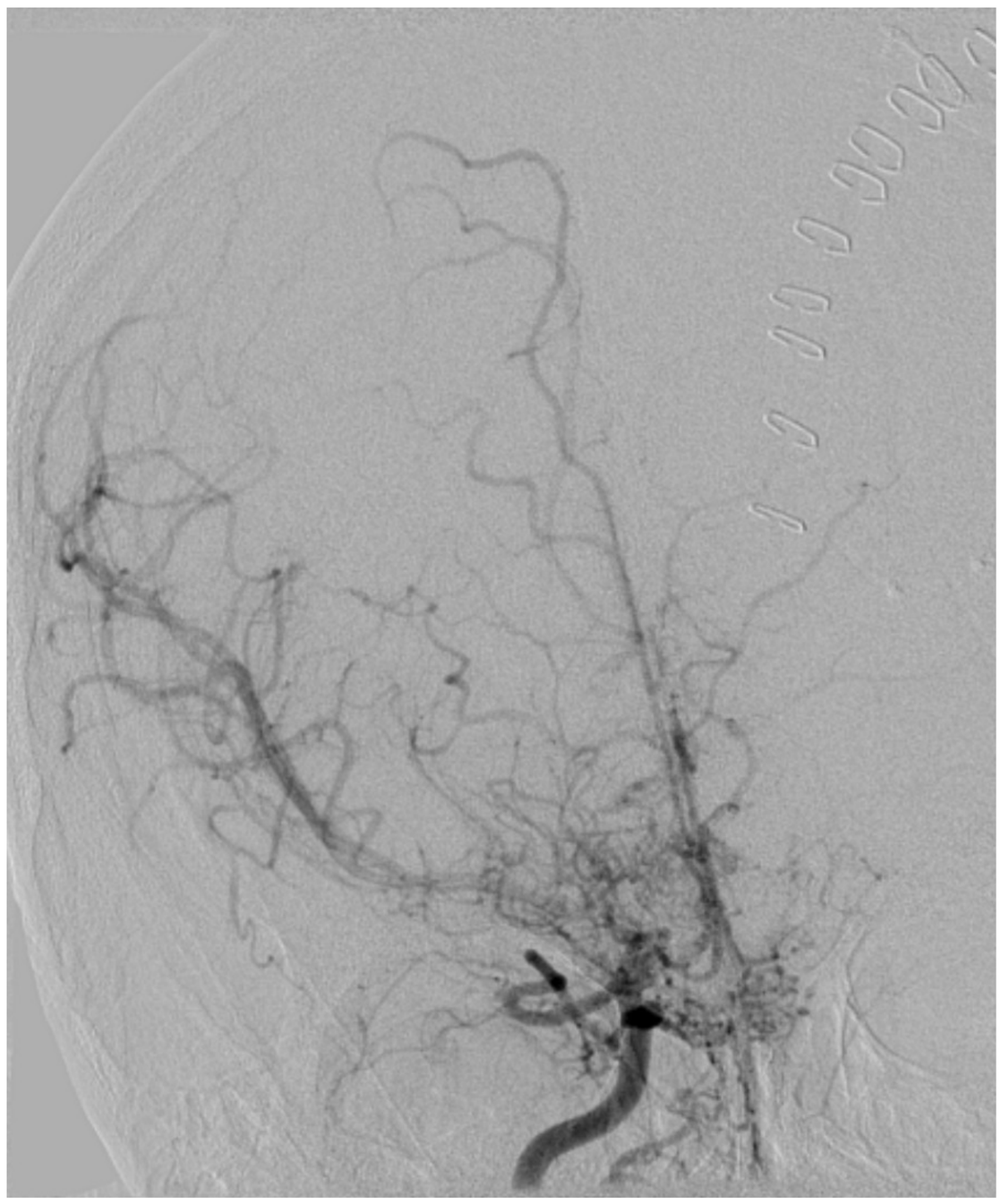

Figure 1. Angiographic study showing severe stenosis of the right internal carotid artery and the collateral network of cerebral perforating arteries.

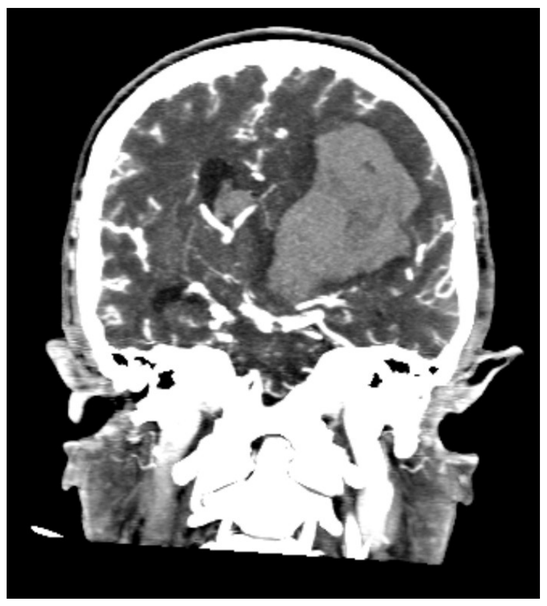

Figure 2.
Brain CT showing massive intraparenchymal hemorrhage in the left fronto-parietal lobe with partial lateral ventricular engorgement, with 2 cm midline shift.
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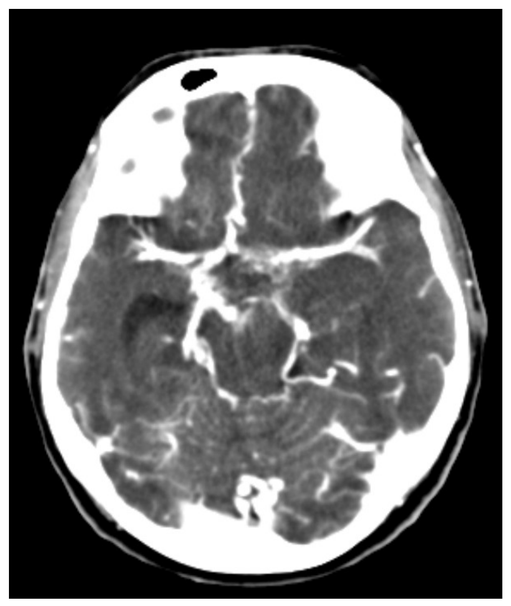

Figure 3.
Brain CT angiography revealing stenosis of the left internal carotid artery with narrowing of the left anterior and middle cerebral arteries.

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