An unusual cause of hemiplegia in a 28-year-old woman

Introduction

This article reports a case of a 28-year-old woman, who had no obvious underlying risk factor, presenting with ischaemic stroke. Further investigation with cerebral angiography found intracranial carotid stenosis and prominent collateral vessels in keeping with Moyamoya disease. ^{99m}Tc-hexamethylpropyleneamine oxime (HMPAO) single photon emission computed tomography with acetazolamide challenge showed severely reduced cerebral vascular reserve, identifying the patient as a candidate for surgery. She underwent superficial temporal artery to middle cerebral artery bypass surgery and has not had any further cerebrovascular events to date.

Discussion

Moyamoya is a rare disorder characterized by progressive intracranial vascular stenosis of the circle of Willis with prominent collateral vessels formation (Scott and Smith, 2009). 'Moyamoya syndrome' is presumed secondary to pre-existing risk factors or associated syndromes (such as radiotherapy to head and neck, Down's syndrome, sickle cell disease and neurofibromatosis type 1). In contrast, 'Moyamoya disease' refers to the idiopathic condition, which is almost always bilateral and predominantly affects the Asian population, although increasingly cases have been observed in the western population (Scott and Smith, 2009).

Patients present with neurovascular insufficiency, i.e. stroke, transient ischaemic attack, seizure, or symptoms related to abnormal collateral flow, e.g. haemorrhage or headache.

Diagnosis is made using multiple imaging modalities. Computed tomography may be normal but most commonly shows infarcts at watershed areas that do not conform to any single vascular territory. Computed tomography angiography and magnetic resonance angiography demonstrate stenosis of the intracranial carotid arteries. This can also be found on conventional angiography which may additionally show prominent striatal collateral vessels having 'puffs of smoke' appearance – pathognomonic of Moyamoya.

Besides conservative management and risk factor modification, bypass surgery is the mainstay of treatment for Moyamoya, most commonly by direct anastomosis of the superficial temporal

Figure 1. Single slice of a computed tomography head examination 3 days after the onset of symptoms demonstrates a focal wedge-shaped area of hypoattenuation on stroke windows (width: 80, level: 40) within the right frontal lobe in keeping with an infarct. Further scattered foci of hypoattenuation were demonstrated on other axial slices.

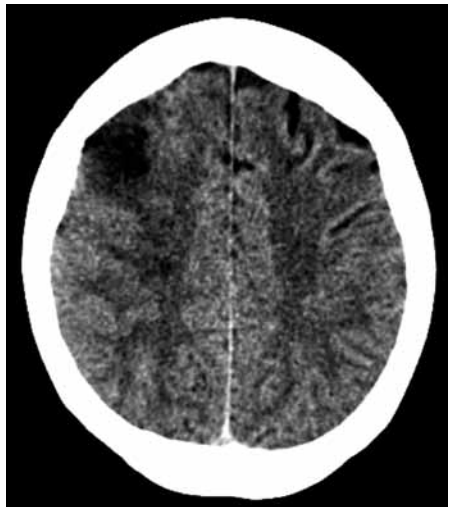
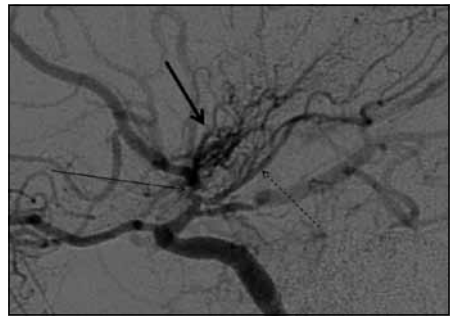


Figure 2. Cerebral angiogram revealed a left supraclinoid internal carotid occlusion (thin arrow), a prominent anterior choroidal artery (dotted arrow) and increased striatal collateral vessels (thick arrow). There was also stenosis of the right supraclinoid internal carotid with increased collateral vessels (not shown).



Case Report

A 28-year-old woman, who had no significant drug, family or past medical history, presented with a 3-day history of sudden onset left-sided weakness. On examination, there was objective loss of power (1/5) in the left upper and lower limbs, paraesthesia, hyperreflexia and an upgoing plantar reflex on the left side. No focal cranial nerve deficit was demonstrated. Lumbar puncture, routine blood tests and vasculitic screen were unremarkable.

Initial computed tomography examination (*Figure 1*) demonstrated a right frontal infarct. A cerebral angiogram (*Figure 2*) showed left internal carotid intracranial occlusion and right internal carotid intracranial stenosis with collateral vessel formation, and a diagnosis of Moyamoya disease was made.

A ^{99m}Tc-hexamethylpropyleneamine oxime (HMPAO) single photon emission computed tomography study before and after acetazolamide infusion was performed to assess reserve capacity of cerebral blood flow. This demonstrated poor reserve (*Figures 3a* and *b*), putting the patient in a high risk category. She subsequent underwent bilateral superficial temporal artery to middle cerebral artery bypass surgery and has not had another cerebrovascular event to date. ^{99m}Tc-HMPAO single photon emission computed tomography was not repeated post procedure.

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artery to the middle cerebral artery (Matsushima et al, 2012). Patients who have evolving symptoms, indicating ongoing infarct, and poor vascular reserve are considered surgical candidates (Matsushima et al, 2012).

Cerebral vascular reserve can be assessed with ^{99m}Tc-HMPAO single photon emission computed tomography before and after acetazolamide (Knop et al, 1992; Kim et al, 2000; Garai et al, 2002; Ma et al, 2007). HMPAO is a lipophilic compound which crosses the blood–brain barrier but is converted to a hydrophilic derivative in the brain, thus trapped inside the cells (Suess et al, 1991). Being a carbonic anhydrase inhibitor, acetazolamide provokes an increase in cerebral blood flow via a transient carbonic acidosis (Sullivan et al, 1987).

In normal individuals, the physiological response to acetazolamide challenge is increased HMPAO uptake indicating increased cerebral blood flow. However, in patients with carotid stenosis with inadequate collateral vasculature, there is a paradoxical reduction of cerebral uptake of HMPAO post acetazolamide challenge (Rogg et al, 1989), as demonstrated in this case.

Conclusions

This case illustrates the classical features of Moyamoya disease, namely a presentation of vascular insufficiency and the absence of associated risk factors, and bilateral stenosis of the intracranial carotids and the associated collateral vessels formation with the altered cerebral blood flow dynamics demonstrated on ^{99m}Tc-HMPAO single photon emission computed tomography following acetazolamide challenge. **BJHM**

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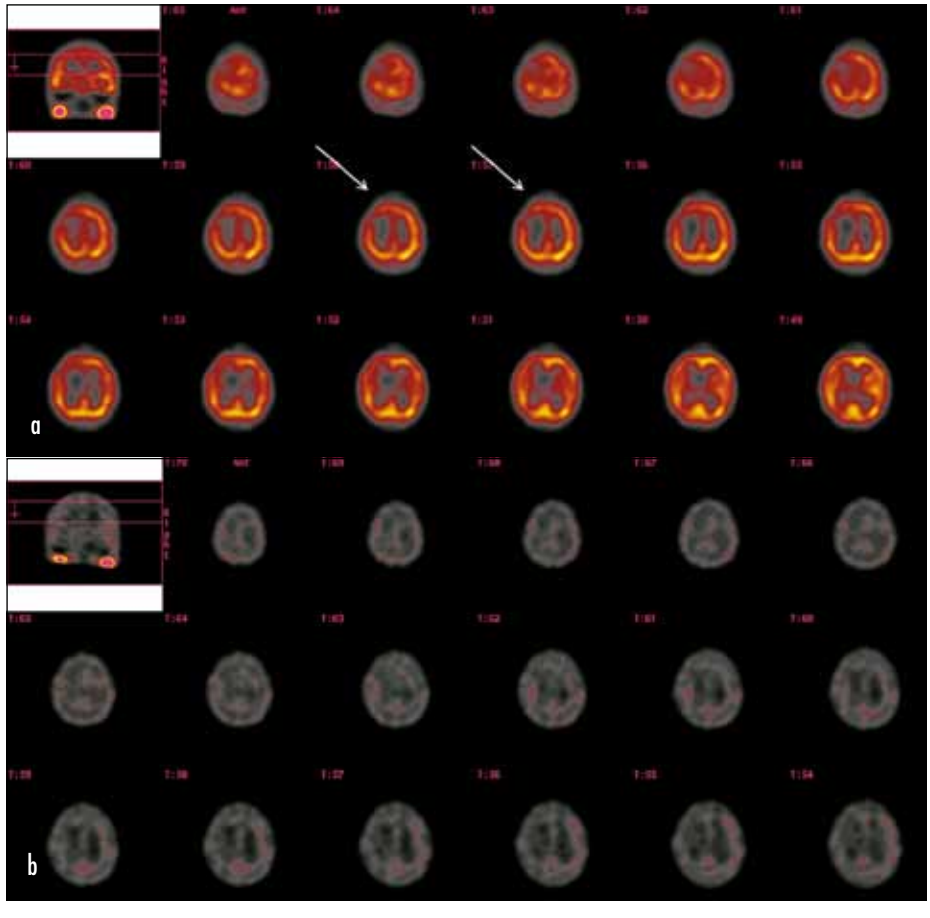
Figure 3. a. The initial ^{99m}Tc-hexamethylpropyleneamine oxime single photon emission computed tomography study (500 MBq) displayed on a red-orange colour scale where red-to-orange indicates areas of relatively high tracer uptake, grey colour indicates relative paucity of tracer uptake (photopenia). Axial images demonstrate global uptake of tracer in the cerebral cortex indicating normal perfusion in a resting state. A wedge-shaped photopenic area in the right frontal lobe (arrows) corresponds to the known cerebral infarct. Central areas of photopenia correspond to the ventricles. b. The procedure was repeated 2 days later immediately following slow intravenous injection of 1000 mg acetazolamide. There is little cortical uptake of tracer indicating poor perfusion and hence poor cerebral vascular reserve.



LEARNING POINTS

- In young patients presenting with a cerebrovascular event, particularly those with associated risk factors, an underlying intracranial arterial stenosis should be considered.
- Diagnosis of Moyamoya requires multiple complementary imaging studies.
- ^{99m}Tc-hexamethylpropyleneamine oxime single photon emission computed tomography with acetazolamide can help to identify suitable candidates for bypass surgery.