Clinical Reports

Cerebral infarction due to moyamoya disease in an 18 year old female

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Summary: Moyamoya disease was diagnosed as the cause of cerebral infarction in an 18 year old white female. The clinical features, pathology and treatment of this occlusive cerebrovascular disorder are discussed.

Introduction

Moyamoya disease was originally described in Japan, and it remains more prevalent there than in western countries.^{1–3} The main features are stenosis of the internal carotid artery and its branches, with compensatory telangiectasias at the base of the brain. The name moyamoya is derived from the angiographic appearance of the filling of the basal telangiectasias which produce a cloudy image resembling a puff of smoke – *moyamoya* in Japanese.^{1–3} Although predominantly a disease of children and females (female to male ratio, 3:2),⁴ moyamoya disease can occur at any age and 7% of cases are thought to be familial.⁵ In children the initial symptoms are due to ischaemia whereas adults more often have haemorrhagic manifestations.^{1–3}

There are very few reported cases of moyamoya disease presenting in adulthood in non-Japanese individuals; furthermore, whilst ascertainment bias cannot be entirely excluded, the majority of such patients have had haemorrhagic events. It has been suggested that the oral contraceptive pill may increase the likelihood of cerebral infarction in adults with moyamoya disease.⁶ We report a case of cerebral infarction in a white 18 year old female with moyamoya disease supporting this suggestion.

Case report

An 18 year old white female was investigated following a 2 week history of progressive confusion, emotional lability, mutism and left-sided weakness. Examination showed an expressive

aphasia with a left-sided hemiparesis more markedly affecting her arm. The right-sided limbs were hypertonic. There were no abnormal findings on general examination. Fourteen months earlier she had experienced temporary weakness of the right arm with slurred speech but she did not come to neurological attention. She was a non-smoker but had been taking the oral contraceptive pill for several months prior to hospital admission. There was no family history of early onset vascular disease or of neurological disease.

A haematological profile, routine biochemistry and clotting screen comprising platelets, prothrombin and activated partial thromboplastin times were normal, the anti-cardiolipin antibody was negative. Autoimmune studies including anti-neutrophil cytoplasmic antibodies, carbon-reactive protein, syphilis serology, angiotensin converting enzyme and human immunodeficiency screen were normal or negative. An echocardiogram and abdominal ultrasound examination were normal. A cerebrospinal fluid examination revealed no evidence of infection or intrathecal immunoglobulin synthesis. An electroencephalogram showed an excess of slow wave activity in the frontal areas but no other abnormality.

A computerized tomographic (CT) head scan demonstrated areas of low attenuation involving both grey and white matter within both frontal areas. There was patchy enhancement of the cortex of both frontal lobes. One week later a repeat CT scan showed further additional small foci of low attenuation in the white matter of the frontal lobes with some diffuse swelling in the right hemisphere: clinically the patient's condition was unchanged. A stereotactic biopsy was undertaken and she was started on oral steroids pending histology. The histology was consistent with infarction and showed no evidence of a vasculitic process. To exclude

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any vascular anomaly an arteriogram was undertaken; this showed bilateral internal carotid artery stenosis with collateralization arising from the circle of Willis consistent with the diagnosis of moyamoya disease (Figure 1). As there were widespread changes on the CT head scan, a brain biopsy was undertaken before angiography. In retrospect, angiography would have provided the diagnosis at an earlier stage. Cerebral blood flow (CBF) was determined using the xenon-133 inhalation method. Global CBF was lower than normal for age (right hemisphere 45 ml/minute/100 g, left hemisphere

47 ml/minute/100 g). Reactivity to hypercapnea was only present in the posterior cortical regions (between 11.3 and 4.83%/mmHg).

The patient's condition gradually improved over the ensuing month with partial resolution of her hemiparesis. However, she remains aphasic and emotionally labile. Steroid treatment has been stopped. She has been commenced on aspirin and is at present awaiting cerebral revascularization by means of omental grafting.

Discussion

The primary lesion in moyamoya disease is stenosis of the intracranial arterial trunk which is thought to be the result of intimal fibrocellular thickening. This leads to the development of the extensive collateral circulation. Occasionally these abnormalities have been found in association with a number of unrelated conditions: neurofibromatosis, Down's syndrome, tuberose sclerosis, fibromuscular dysplasia, radiation-induced vasculitis, sickle cell anaemia, Fanconi's anaemia and polycystic kidney disease.^{2,3,7-10} The frequency of certain HLA markers and autoantibodies has been reported to be of higher frequency in patients with moyamoya disease but there is no unique association.¹¹ Furthermore, there is little to suggest an immune basis for this disease from histological studies.¹² The natural history of moyamoya disease is not fully established but a better prognosis appears to be associated with presentation in adult life. Whilst no predictive factors have been identified, it seems quite likely that in adult cases the presence of additional risk factors for cerebrovascular disease may precipitate infarction. In the study by Bruno and co-workers⁶ all seven female cases of moyamoya disease aged between 17 and 40 presenting with cerebral infarction were taking the oral contraceptive pill; six also had additional risk factors (diabetes mellitus and/or hypertension).

The optimal management of moyamoya disease is unknown. Although reports are largely based on non-randomized treatments, several direct and indirect cerebral revascularization procedures appear to be of benefit.^{13,14} Treatment with steroids and vasodilatation has proved ineffective.^{15,16} However, long-term aspirin therapy may be appropriate for adult cases.⁶

The aetiology of moyamoya disease is unknown but is not confined to Japanese individuals and may account for 4% of cerebral infarctions in young individuals.⁶ In reporting this case, we feel that moyamoya should be considered when investigating young adults with cerebral infarction. It seems likely that the presence of additional risk factors for cerebrovascular disease may alter the natural history of the condition in adults.

Figure 1 (a) Right internal carotid angiogram showing stenosis of the terminal internal carotid and proximal anterior and middle cerebral arteries (arrow) with hypertrophied ganglionic perforating vessels. (b) Left common carotid angiogram showing more severe proximal intracranial vessel stenosis and a more florid collateral supply (arrow) including cortical collaterals from the posterior cerebral artery.

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Community-acquired bacteraemic *Acinetobacter* pneumonia with survival

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Summary: A 65 year old man was admitted with segmental consolidation of the left upper lobe after having stayed in a hotel for 2 days. He deteriorated rapidly on conventional antibiotic therapy and required ventilatory support. *Acinetobacter calcoaceticus* var. *anitratus* was grown from the sputum and blood cultures, which was treated with a combination of anti-pseudomonal agent, aminoglycoside and cotrimoxazole. He made a slow but remarkable recovery from the pneumonia. *Acinetobacter* is a rare potentially fatal cause of community-acquired pneumonia.

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Introduction

Acinetobacter calcoaceticus is an aerobic pleomorphic, encapsulated Gram-negative predominantly coccobacillus or diplococcus, which may be confused with *Neisseria* or *Haemophilus*.¹ It is widely distributed in water and soil and may be frequently